CHAPTER 8
A Survey of Diseases of Peripheral Nerve, and Nerve Root

INTRODUCTION:

In chapter 2, we have already considered, the differential features of disease of muscle compared to disease of peripheral nerve compared to disease of the nerve root compared to central nervous system disease. Diseases of peripheral nerve and nerve root both represent, lower motor neuron disorders. In contrast to disorders of muscle, both involve, motor and sensory features. As we will discuss in greater degree, peripheral nerve disorders, may involve specific nerves in a local manner (mononeuropathies) or peripheral nerve in a more generalized manner (polyneuropathies). Mononeuropathies must be distinguished from radiculopathies. This distinction can be made based on the pattern of motor deficit, the pattern of deep tendon stretch reflex deficit, the pattern of sensory deficit and the distribution of pain. Figure 8-1 compares radicular (dermatomal or segmental) sensory innervations to the superficial sensory innervations of peripheral nerve.

PERIPHERAL NERVE DISEASE.

Diseases involving the peripheral nerves have a combination of motor and sensory symptoms and signs. Lower motor neuron findings are present with a flaccid type weakness and atrophy. Sensory symptoms and findings (involving to a variable degree all modalities of sensation) are present within the same distribution as the motor findings. These patients, however, do not show evidence of damage to the long fiber systems involved in transmitting sensory and motor information within the central nervous system.

In general, mental status is intact. Deep tendon reflexes and superficial reflexes (response to plantar stimulation) are absent within the distribution of the involved peripheral nerves. Fasciculations may be present within the distribution of the involved peripheral nerves. Damage to the sympathetic fibers, traversing the peripheral nerves, may result in alterations in sweating and skin temperatures. Nerve conduction studies demonstrate reduction in velocity (if the basic process involves loss of myelin, demyelination) or in the amplitude of the motor or sensory action potential (if the basic process involves predominately a loss of axons). In mononeuropathies the specific site of damage (conduction block) may be demonstrated. The EMG will demonstrate abnormal spontaneous activity: fibrillations (onset 10-25 days after the axonal damage) positive sharp waves and fasciculations.

As will be discussed later in greater detail, diseases of peripheral nerves are essentially of two types: (a) localized mononeuropathies involving a single peripheral nerve, often due to trauma or compression or less often, occlusion of blood supply. (b) Symmetrical polyneuropathies - usually distal and usually due to a metabolic disturbance involving many nerves. The polyneuropathies reflect many of the systemic, toxic and metabolic disease considered in pathology. As a general rule any patient with diabetes mellitus or with chronic alcoholism or receiving chemotherapy is likely to present symptoms or signs (often subclinical) of a polyneuropathy. In many cases we are not able to establish a specific cause for the polyneuropathy. We may not have sufficient information about environmental or industrial exposure. In some cases we have insufficient information regarding the family history.

In a mononeuropathy, the weakness and sensory signs and symptoms are clearly within the distribution of a specific plexus peripheral nerve, e.g., sciatic, radial, median, or ulnar. *(Fig. 8-1)*. Common sites of compression include the radial nerve at the radial (or spiral) groove of the humerus, the ulnar nerve at the

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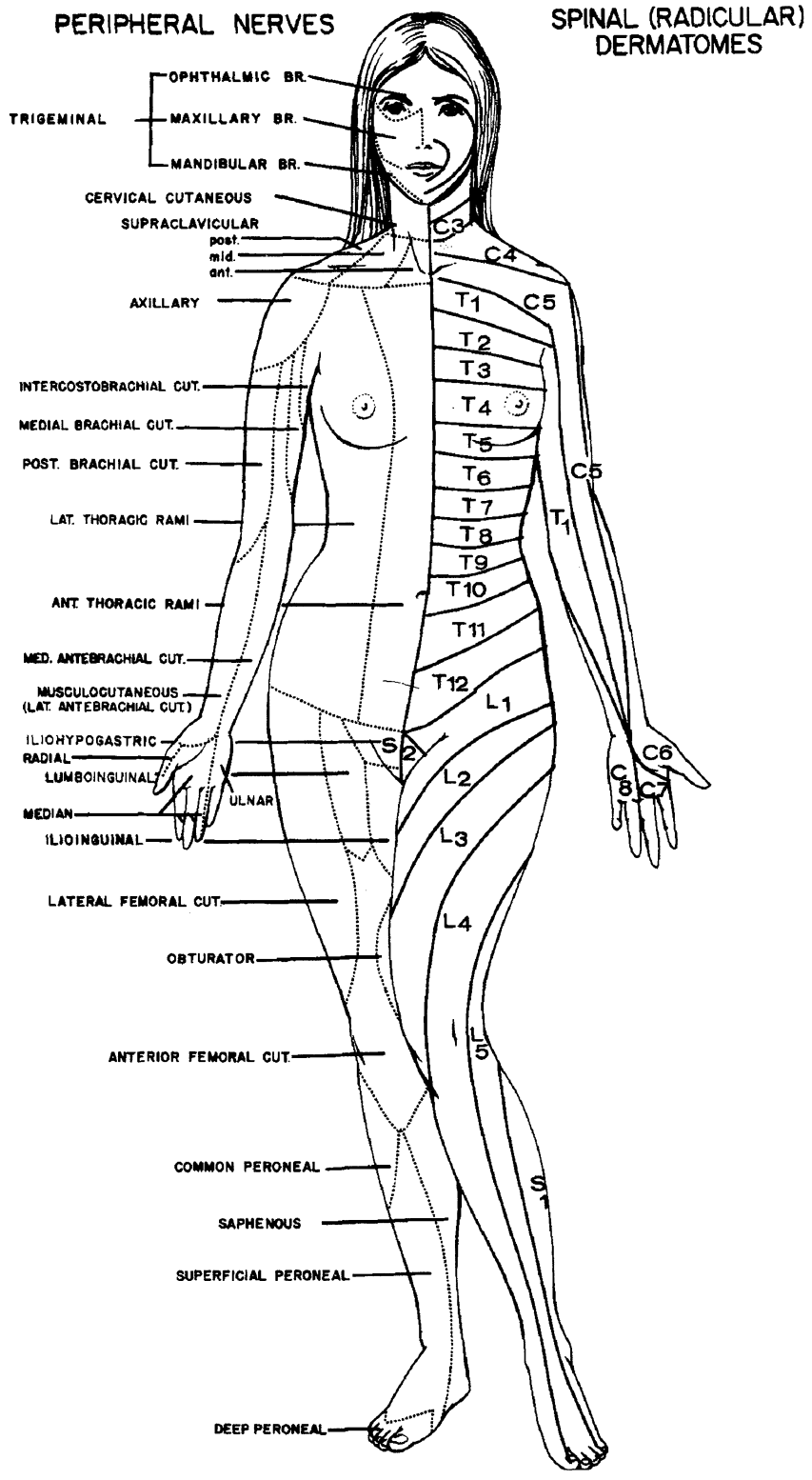
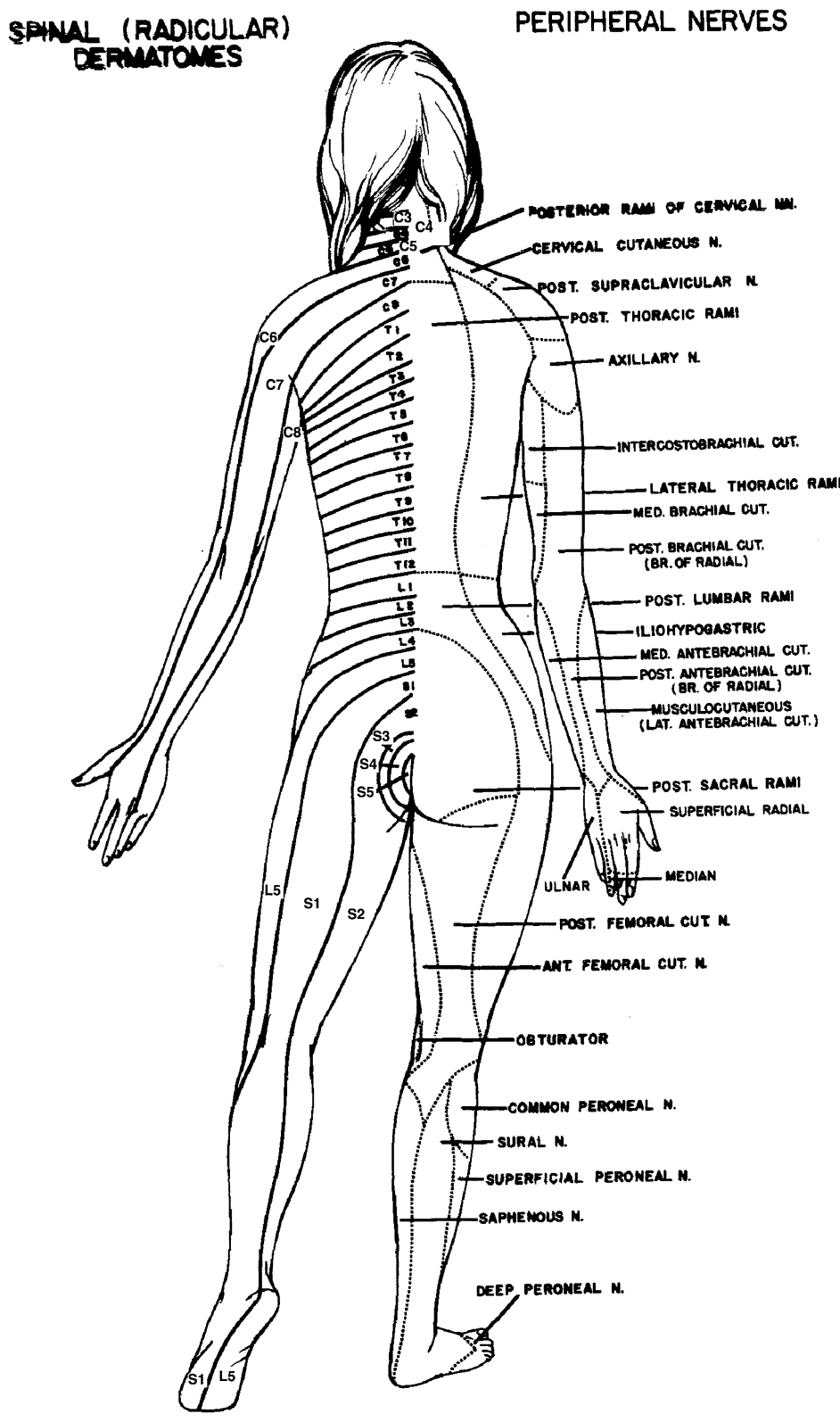


Figure 8-1. Comparison of radicular (dermatome or segmental) and peripheral nerve innervation.


SPINAL (RADICULAR) DERMATOMES
PERIPHERAL NERVES
C3
C4
C5
C6
C7
C8
T1
T2
T3
T4
T5
T6
T7
T8
T9
T10
T11
T12
L1
L2
L3
L4
L5
S1
S2
S3
S4
S5
POSTERIOR RAMI OF CERVICAL NN.
CERVICAL CUTANEOUS N.
POST. SUPRACLAVICULAR N.
POST. THORACIC RAMI
AXILLARY N.
INTERCOSTOBRACHIAL CUT.
LATERAL THORACIC RAMI
MED. BRACHIAL CUT.
POST. BRACHIAL CUT. (BR. OF RADIAL)
POST. LUMBAR RAMI
ILIOHYPOGASTRIC
MED. ANTEBRACHIAL CUT.
POST. ANTEBRACHIAL CUT. (BR. OF RADIAL)
MUSCULOCUTANEOUS (LAT. ANTEBRACHIAL CUT.)
POST. SACRAL RAMI
SUPERFICIAL RADIAL
ULNAR
MEDIAN
POST. FEMORAL CUT N.
ANT. FEMORAL CUT. N.
OBTURATOR
COMMON PERONEAL N.
SURAL N.
SUPERFICIAL PERONEAL N.
SAPHENOUS N.
DEEP PERONEAL N.

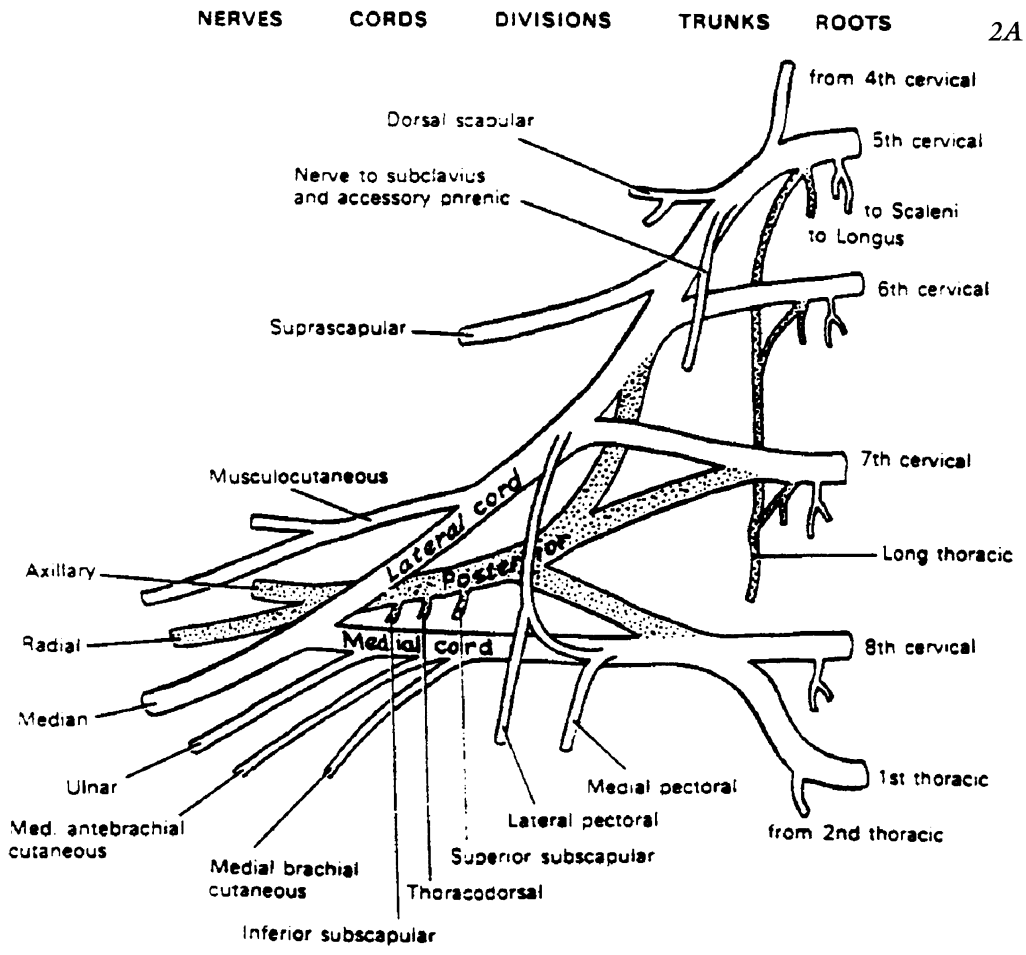


Figure 8-2. A) Plan of the Brachial Plexus: Note origin of median nerve from both medial and lateral cord, of ulnar nerve only from medial cord with radicular origin, from C8, T1, of radial nerve from posterior cord only. (From Clemente, C. Gray's Anatomy. Philadelphia. Lea and Febiger. 30th Ed. 1985, p. 1205).

olecranon process of the elbow, the median nerve in the carpal tunnel, the brachial plexus at the thoracic outlet, and the peroneal nerve as it leaves the popliteal fossa and curves around the head of the fibula.

A specialized form of mononeuropathy: mononeuropathy multiplex reflects multifocal involvement of peripheral nerve for example in inflammatory diseases of blood vessels.

In addition, in neurofibromatosis (Von Recklinghausen's disease), multiple peripheral nerves and nerve roots may be involved by tumors arising from the Schwann cell and mesodermal components. This disorder will be discussed in relation to the spinal cord.

MONONEUROPATHIES: UPPER EXTREMITY

BRACHIAL PLEXUS –[C5, 6, 7, 8 T1- *(Fig.8-2, 8-3)]*

Patients frequently present with a complaint of transient distal sensory symptoms and weakness involving the upper extremity occurring in relationship to sleeping posture or position of the arm. The symptoms are often bilateral. Rarely the symptoms are persistent rather than intermittent. These symptoms are referred to as the **thoracic outlet syndrome or the neurovascular syndrome of the thoracic outlet.** Both the brachial plexus and the subclavian artery pass through a relatively narrow

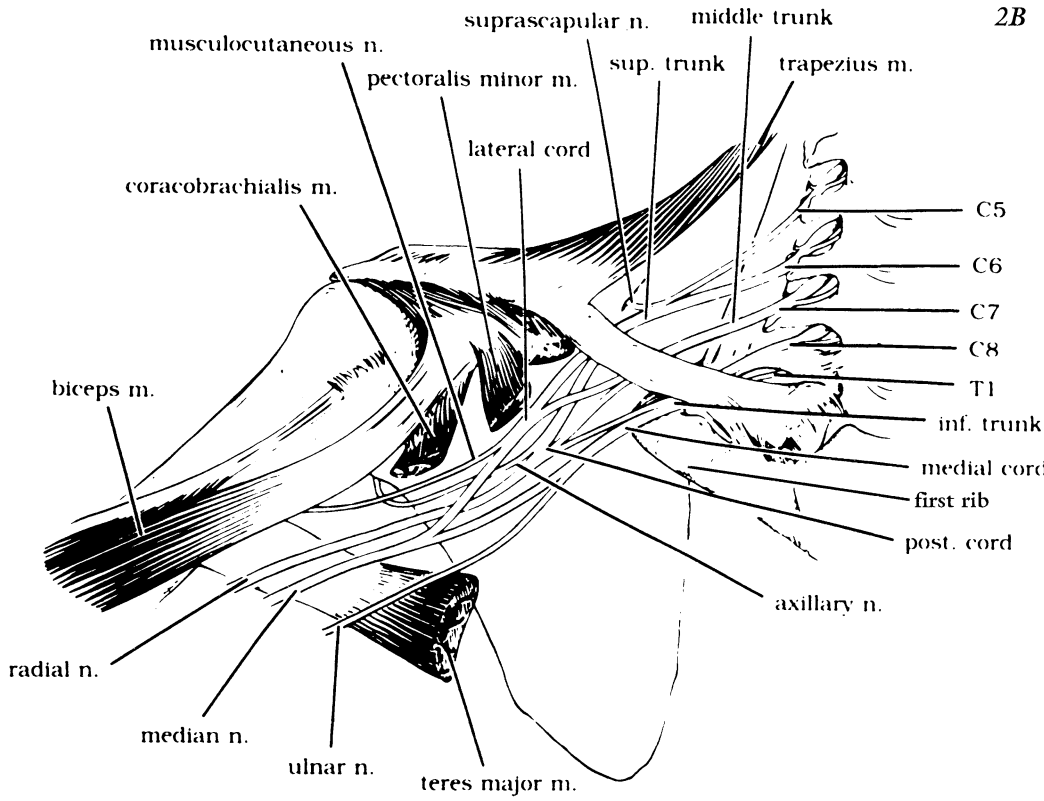


Figure 8-2. B) Brachial plexus. Relationship to the neural foramina the first rib and the clavicle. (From Zimmerman J. and Jacobson S. Anatomy. Boston. Little Brown 1989 p.176.

area with clavicle anteriorly and the first rib posteriorly *(Fig.8-2)*. **Alterations in posture may produce compression of the brachial artery - producing a tingling pins and needles sensations ("paresthesias") involving all fingers and a distal weakness.** Often specific maneuvers will demonstrate the intermittent compression of the brachial artery. **Neural syndromes of the outlet tend to involve primarily the ulnar nerve, or the lower roots (C8-T1).** At times a cervical rib or tight band may further compress the narrow outlet.

Upper Plexus: In addition to the relatively benign thoracic outlet syndrome, often trauma, traction downwards as in birth injuries (Erb's) or brachial neuritis may involve the upper plexus roots C5-6 *(Fig.8-2)* resulting in weakness of muscles about shoulder and flexors at elbow. Brachial neuritis presumably has a post infectious or immunologic or familial etiology. The specifics are often unclear. Brachial neuritis is often associated with considerable pain in the arm. Radiotherapy to the axilla for breast carcinoma may also involve the upper plexus in a relatively nonpainful syndrome.

Lower Plexus: Malignant infiltration of the plexus from tumors at the apex or upper lobe of the lung ("Pancoast tumor") often involves the lower (C7, 8,T1) half of the plexus producing severe pain. The related sympathetic plexus is often involved producing a Horner's syndrome. Upward traction on the plexus may occur at birth (Klumpke's) or in children who suddenly are pulled up by the arm producing damage to the lower half of the plexus.

Case 8-1 provides an example of a brachial plexopathy and Horner's syndrome in a patient subsequently found to have an apical carcinoma of the lung.

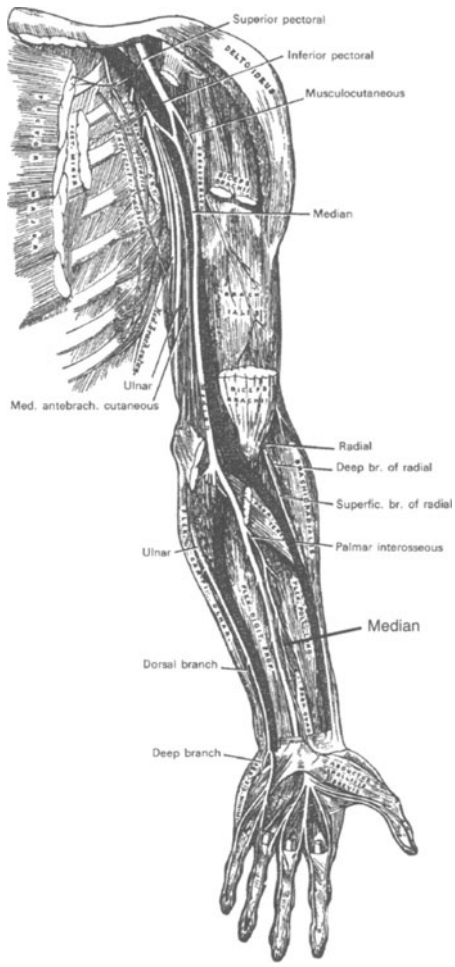


Figure 8-3. Dissection of Nerves of the Left Upper Extremity from anteriorly. Note exposed position of the ulnar nerve in the olecranon groove at the elbow, position of median nerve at the carpal tunnel at the wrist and relationship of the brachial artery to the brachial plexus and clavicle. See also fig 8-4. (From Clemente, C. Gray's Anatomy 30th Ed. Philadelphia. Lea and Febiger 1985, p. 1214).

Case 8-1 This 57 yr. old right-handed white male with a 30-year history of heavy cigarette smoking (2 packs per day) had a 2-year history of progressive pain in the left upper extremity beginning with mild pain at the left elbow, then a lack of sensation in the left ulnar distribution. Eight months prior to admission, the left hand had become swollen and painful. Severe pain on motion at the shoulder also developed. Six months prior to evaluation drooping of the left eyelid was first noted followed by a decrease in sweating on the left side of face and body. Three months prior to admission the patient had the onset of weakness of the left arm. The patient soon noted.

Physical examination demonstrated swelling, redness, change in temperature and sweating and limitation of motion of the left arm, plus distension and firmness of the left supraclavicular space. Orthostatic hypotension was present

Neurological examination: *Symphabetic System:* Seen as a full Horner's syndrome on the left. *Motor System:* atrophy in the median (thenar eminence) and ulnar (first interosseus) distribution, weakness in left arm most marked in the median and radial distribution. *Reflexes:* Decreased deep tendon reflexes left arm *Sensory System:* Decreased pain in the left C7, C 8 dermatomes.

Clinical diagnosis: Brachial plexopathy and Horner's syndrome due to apical lung tumor (Pancoast tumor).

Laboratory data*: Computerized tomography of the lower neck and upper chest* after venous injection of the contrast material into the left arm indicated: near occlusion of the left subclavian vein at the level of the first rib and a necrotic mass in the left lower neck posterior to the thymus and carotid sheath and invading the muscular structures of the prevertebral region and arising from or extending to the left apex of the lung. *Nerve conduction and EMG studies* indicated a severe axonal lesion of the lower half of the brachial plexus with a marked involvement of median and ulnar nerves and less of the radial nerve. *Biopsy of the left supraclavicular mass* revealed adenocarcinoma presumably of pulmonary origin.

Subsequent course: The patient received radiotherapy (3000 rads) to the left lung apex and the left supraclavicular area with a decrease of pain and of swelling in the arm. The patient expired 1.5 years after radiotherapy and 3.5 years after onset of symptoms.

Often a brachial neuritis involves only a limited portion of the brachial plexus. Limited involvement of the long thoracic nerve (C5, 6, 7) is relatively frequent.

Case 8-2 presented on CD ROM provides an example of a neuropathy of the long thoracic nerve occurring in a woman 1 day after delivery.

Ulnar nerve (C8, T1): This nerve is often exposed to trauma as it passes through the groove behind the medial epicondyle at the elbow *(Fig.8-3)*. Each of us has struck our "funny bone" at some time and we are all acquainted with the ulnar distribution of positive sensory symptoms, ring and 5th finger. Repeated trauma to the nerve or fractures at the elbow may produce more persistent symptoms including atrophy of the hypothenar and interossei muscle and weakness of digiti quinti abductor, ring and fifth finger flexors, the interossei, and the thumb adductor.

Median nerve (C5, C6, C7, C8, T1): The major syndrome of the median nerve relates to compression at the carpal tunnel by the overlying transverse carpal connective tissue. *(Fig. 8-3, 8-4)*. Pain and paresthesia extend into the median nerve supplied fingers predominantly index and middle, with less invovement of thumb and median side ring finger when severe. There is significant weakness and atrophy involving the median supplied thenar muscles of the hand:-abductor pollices brevis, opponens and the finger flexors. A positive Tinel's sign is present on percussion over the carpal tunnel. Carpal tunnel syndrome may complicate any process where edema or swelling occurs at wrist or hand: rheumatoid arthritis (as in case 8-3), trauma, myxedema (hypothyroid state) or acromegaly (a state associated with excessive production of pituitary growth hormone-see Chapter 16).

Case 8-3 This 83 yr. old right-handed white widow and retired shoe factory worker with a 20-30 year history of rheumatoid arthritis, in relationship to an exacerbation of her joint symptoms, had the onset of tingling paresthesia in the left median distribution of hand and fingers most prominent in the middle finger, less in index and ring and minor in thumb. There was pain in the hand particularly or making a fist.

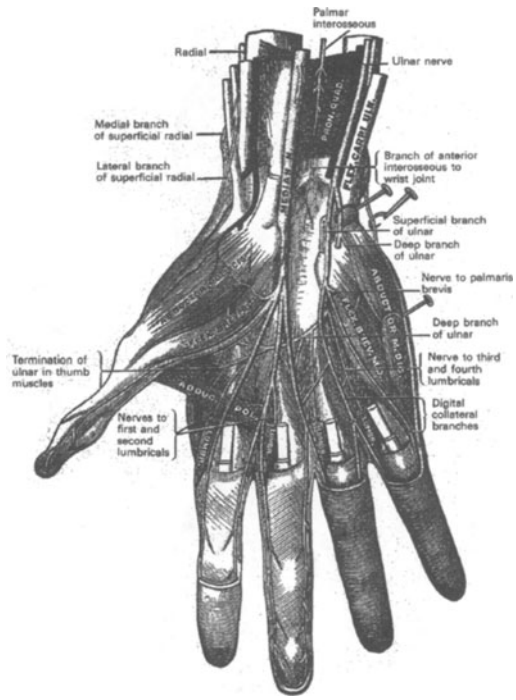


Figure 8-4 Dissection of Nerves and Muscles of the palm of the right hand.(From Clemente, C: Gray's Anatomy 30th Ed. 1985, p. 1219 after Testut).

Physical examination demonstrated mild inflammatory synovitis in the hands, with mild soft tissue swelling in the wrists, ankles, proximal interphalangeal joints, and metacarpal phalangeal joints.

Neurological examination demonstrated a minor weakness in left hand grip and left thumb opponens and a marked tenderness over the left carpal tunnel area with a positive Tinel's sign on palpation or percussion of the median nerve in the carpal tunnel (tingling paresthesias extending from the carpal tunnel into the median nerve supplied fingers).

Clinical diagnosis: Compression of median nerve at carpal tunnel.

Laboratory data:Nerve conduction studies indicated severe delays in both sensory and motor conduction for median nerve at carpal tunnel on the left with mild findings on the right.

Subsequent course: Despite the use of nonsteroidal anti-inflammatory agents, re-evaluation 2 weeks later indicated additional pro-

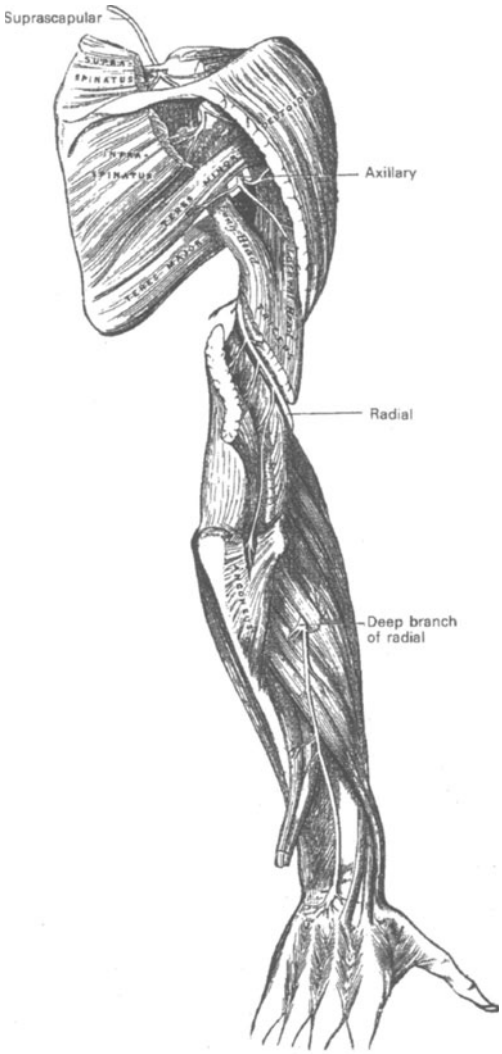


Figure 8-5 Dissection of the nerves of the right upper extremity - from posteriorly. Note the radial course of the radial nerve providing innervation to the triceps muscle, and to the extensors of wrist and finger (From Clemente, C.: Gray's Anatomy. 30th Ed. 1986, p. 1220.)

gression: greater weakness in grip and pain and touch sensation now decreased bilaterally in the median nerve distribution. Despite the use of local steroid injections, reevaluation one month later indicated persistence of symptoms, with atrophy present in the thenar eminence on the left. Carpal tunnel surgery on the left hand was performed with improvement of symptoms. When symptoms developed in the right hand, similar surgery was performed with relief.

Radial nerve (C6, 7, 8): The major syndrome of this nerve relates to injury at the spiral groove of the humerus (Fig.8-5) related to fractures or to so called "Saturday Night Palsy" (pressure effects on arm draped over a hard surface are noted due to excessive alcohol intake so that the paresthesias fail to awaken the patient and the compression therefore continues). The characteristic weakness involves the wrist and finger extensor as well as the long abductor of the thumb.

The brachioradialis muscle is usually involved. Lesions at this level however usually spare the triceps muscle also supplied by radial nerve. A wrist drop is characteristic. A cock up wrist splint is utilized to maintain the hand in a physiological position. With time recovery will occur.

MONONEUROPATHIES : LOWER EXTREMITY

LUMBAR SACRAL PLEXUS [L1-S3 (FIG.8-6, 8-7)].

The lumbar plexus is composed of nerve roots L1 - L4, and the sacral plexus of nerve roots L4-S3. Both may be involved by malignancies within the pelvis. Both may also be commonly involved in the painful diabetic mononeuropathy that occurs on a vascular basis.

Neuropathies may commonly involve the following branches of the lumbar plexus.

1. Lateral femoral cutaneous nerve of the thigh (L2, L3). This nerve supplies the anterior-lateral aspect of the thigh. It enters the thigh by passing between the two points of attachment of the lateral aspect of the inguinal ligament to the anterior superior iliac spine (Fig.8-8). There is sensitivity of the supplied cutaneous area to contact from any clothing or repetitive tactile stimulation. Tingling paresthetica are present. At times burning painful paresthesia is present; thus, the name "meralgia paresthesia". The common causes are weight gain or weight loss. The following underlying diseases are frequent: obesity, pregnancy, dia-

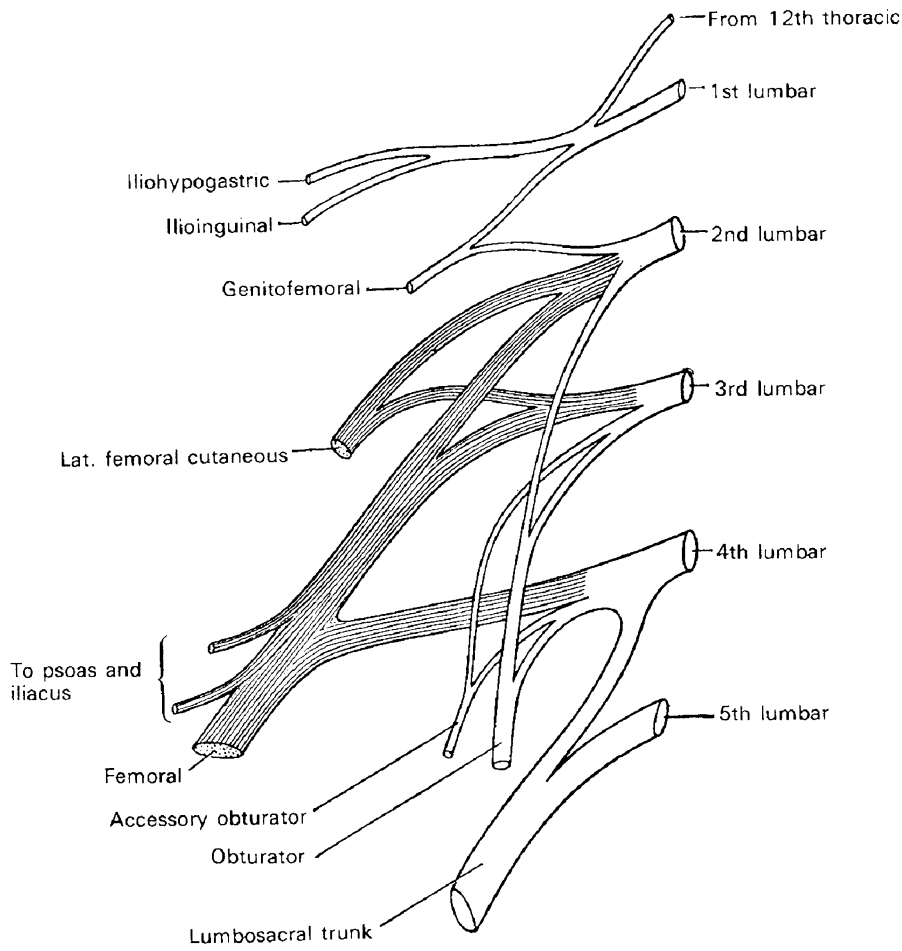


Figure 8-6. Plan of the Lumbar Plexus. The anterior divisions of L2, L3, L4 unite to form the obturator nerve. The posterior divisions of L2, L3, L4 (shaded) unite to form the femoral nerve. Twigs from L2, and L3 forms the lateral femoral cutaneous nerve. (From Clemente, C.: Gray's Anatomy, 30th Edit. 1985 p. 1226).

betes and infrequently pelvic malignancy.

2. Obturator nerve (L3, L4), *(Fig.8-8, 8-9,8-10)* **this nerve supplies the adductors of the thigh.** The major causes of injury relate to difficult deliveries: pressure from fetal head or forceps.

3. Femoral nerve (L2, 3,4), *(Fig-8-8, 8-10).* This nerve supplies the quadriceps muscle necessary for extension at the knee and the iliacus and psoas muscles necessary for hip flexion. Diabetic mononeuropathy on a vascular basis is the most common cause of this neuropathy. Hematomas into the iliac muscle may compress the nerve. Pelvic malignancies and pelvic surgery may injure the nerve.

SCIATIC NERVE AND DIVISIONS

Neuropathies may commonly involve the sciatic nerve, (the major trunk originating from the sacral plexus) or the two major divisions of the sciatic nerve: the common peroneal and the posterior tibial nerves. The two divisions are bound together from the plexus to just above the popliteal fossa. They are separate nerves below that point. The other branches of the sacral plexus are a) the gluteal nerves to the gluteus medius and gluteus maximus; b) the pudendal nerve (S2, 3,4) to the perineal muscle of the anal sphincter.

Sciatic nerve: (L4, L5, S1, S2), *(Fig.8-11).* This nerve supplies the hamstring muscles and

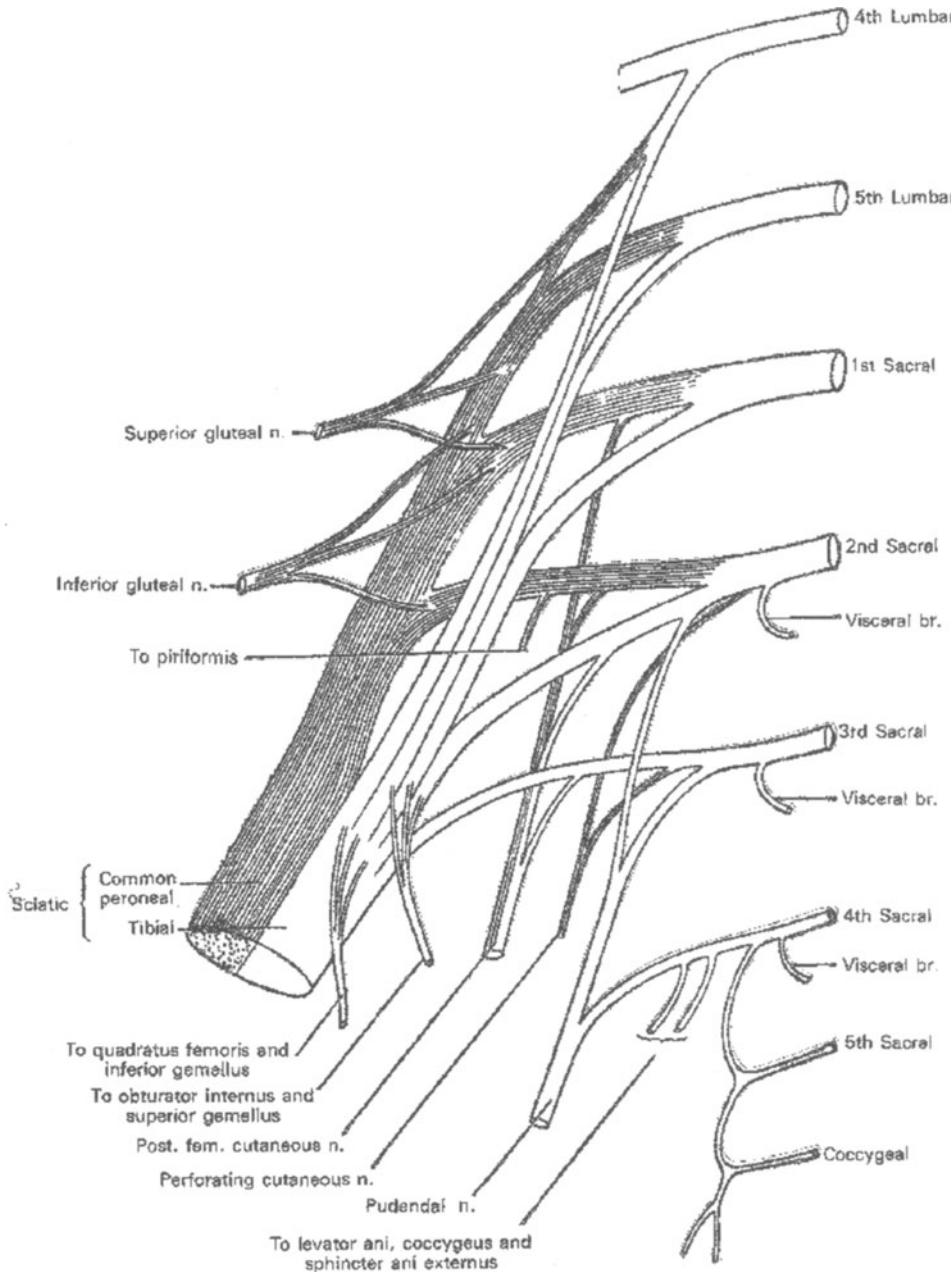


Figure 8-7 Plan of the Sacral Plexus and Coccygeal Plexus: Posterior divisions are depicted as striated. Anterior divisions are unshaded. Note origin of common peroneal nerve from posterior divisions L4-S2 and of tibial nerve from anterior divisions (L4-S3). The two nerves are joined together to form the sciatic nerve (From Clemente, C: Gray's Anatomy, 30th Ed. 1985, p. 1235).

all muscles below the knee. Common causes of injury are a) fractures of pelvis or femur, b) gunshot wounds of the buttock; c) Injections of medications into the buttock - e.g. penicillin. d) Diabetes mellitus mononeuropathy.

Peroneal Nerve: (L4, L5, S1), *(Fig.8-10, 8-11)*: This nerve has a) superficial branch: cutaneous and muscular supply to the everter of the foot (peroneal muscles) and b) deep peroneal or anterior tibial branch supplying the

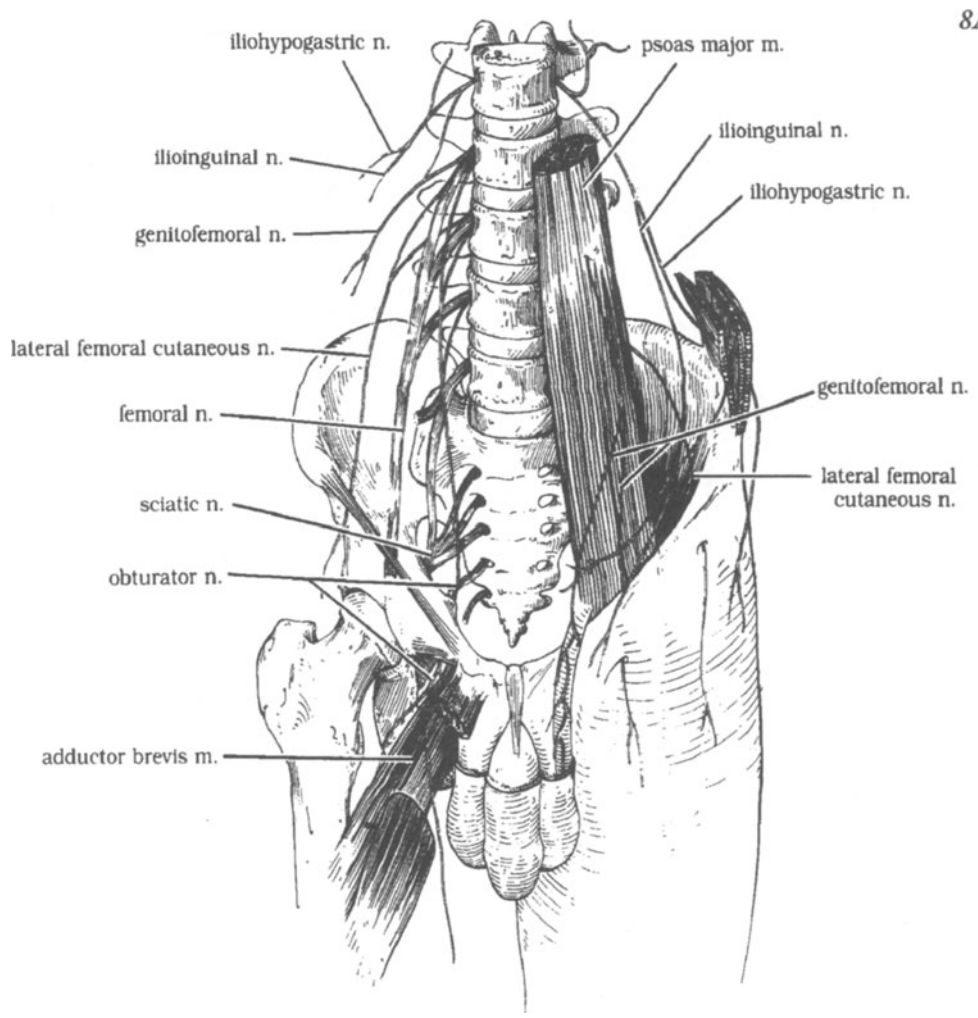


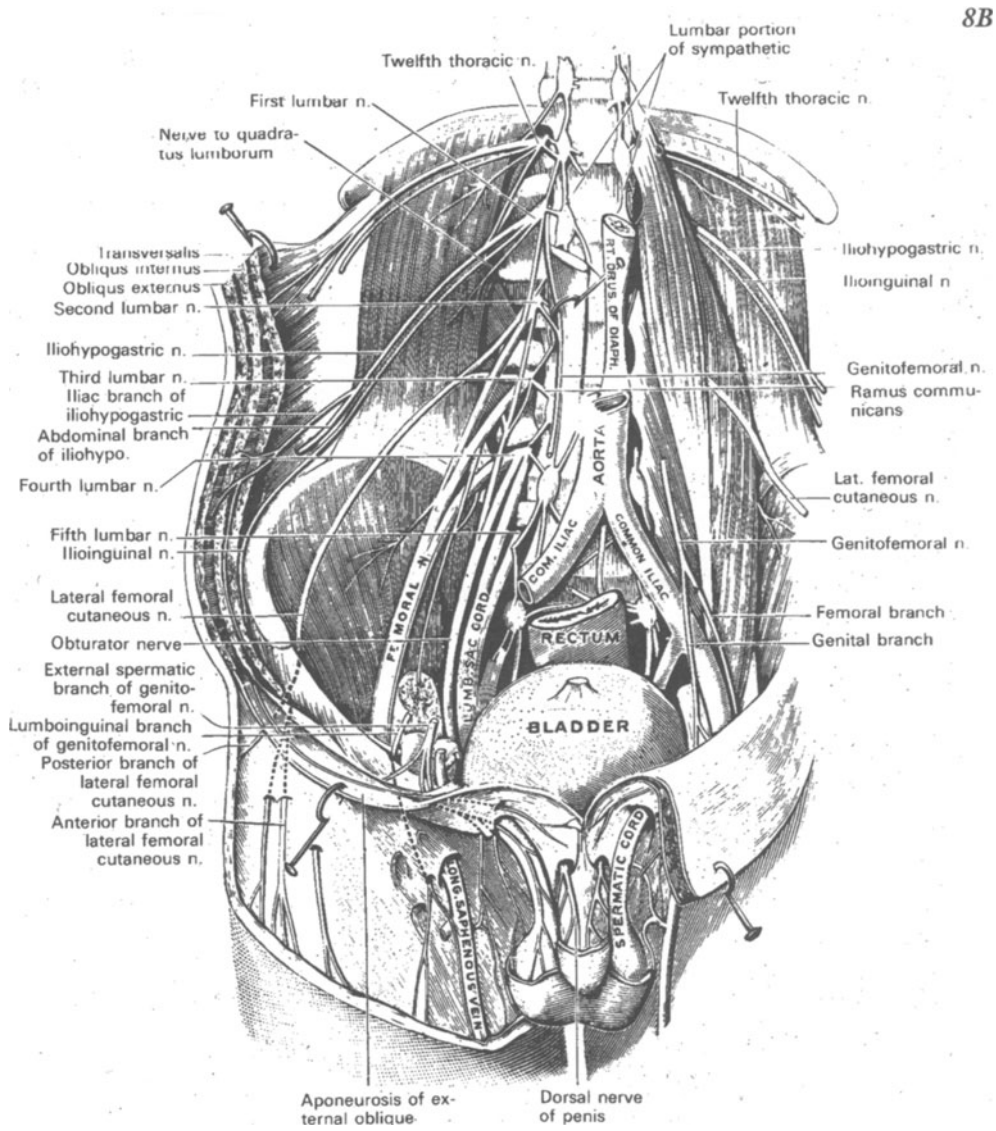
Figure 8-8. A) Dissection of the lumbar and sacral plexuses. demonstrating their relationship to the vertebral foramina and the inguinal ligament and the sciatic notch.(From Zimmerman, J.and Jacobson, S. Anatomy 1989 p.108.

ankle and toe dorsi flexor muscles and sensation to the dorsum of foot and lateral aspect of the calf. This nerve passes around the head of the tibula from the popliteal fossa to the anterior aspect of the leg. In this location, the nerve is relatively superficial and very subject to pressure from leg crossing (when seated in tight coats or tight boots) or operating or obstetrical room stirrups. The major manifestation of compression is a foot drop or unstable ankle (due to weakness of ankle dorsiflexors and everter).

Tibial Nerve: (L4, L5, S1, S2): *(Fig.8-11)*

This nerve supplies the posterior calf muscles. The gastrocnemius and soleus muscles, (the plantar flexor and then continues as the posterior tibial nerve to supply intrinsic foot muscles. It also supplies sensation to the plantar surface. The posterior tibial nerve passes behind the medial malleolus in a relatively superficial location and enters the tarsal tunnel. In this location, the nerve may be palpated and is subject to compression. Entrapment also occurs due to disease in the tendon sheaths that accompany the nerve in the tunnel.

The following case 8-4 provides an example of a mononeuropathy involving the lumbar -



Deep and superficial dissection of the lumbar plexus. (Testut.)

Figure 8-8. B) Dissection of the Lumbar Plexus. Note relationship of the plexus to the psoas muscle (dissected away on right side of abdomen) iliacus muscle and pelvic structures. Note the course of the lateral femoral cutaneous, femoral and obturator nerves and possible sites of compression. (From Clemente, C: Gray's Anatomy 30th Ed. 1985 p. 1228).

sacral plexus in a diabetic patient who also had a peripheral neuropathy.

Case 8-4: This 75 yr. old right-handed white widow with a one-year history of non-insulin dependent diabetes mellitus had the sudden onset of weakness in the right leg, most prominent at hip and knee, three weeks prior to evaluation. Shortly thereafter, she had the onset of a severe toothache like pain in the lateral and anterior surface of the right leg from hip to knee. No other symptoms were present.

Neurological examination: *Motor System:* Weakness was present in the proximal muscles of the right lower extremity. *Reflexes:* Deep tendon stretch reflex was absent at the right Achilles and patellar compared to left. *Sensory*

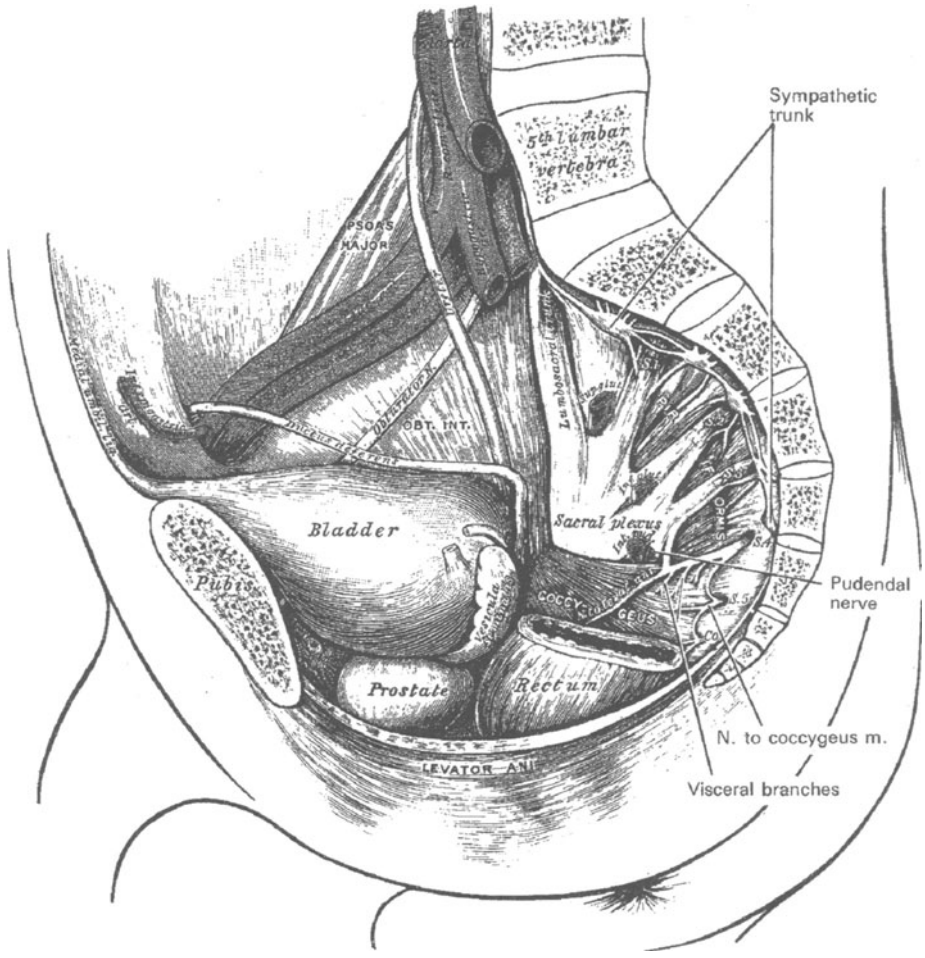


Figure 8-9. Dissection of the Sacral and Pudendal Plexuses. Sagittal View. Note the relationship of the plexus to the visceral structures. Note also the course of the obturator nerve (from the lumbar plexus) as it passes forward to enter the obturator canal and foramen. See also Fig. 8-6B (From Clemente, C.: Gray's Anatomy, 30th Ed. 1985, p. 1233).

System: Pain sensation was decreased in a symmetrical manner over the toes and feet and two thirds of the distance up the calves. However there was also a focal deficit in pain sensation over the right third lumbar dermatome. Vibration was decreased at toes compared to ankles compared to knees in a bilateral manner.. Significant tenderness was present over both the sciatic nerve at the sciatic notch and the femoral nerve in the femoral canal of the anterior thigh.

Clinical diagnosis: 1. Acute diabetic mononeuropathy: lumbar and to a lesser degree sacral plexopathy; 2. Diabetic distal peripheral neuropathy.

Laboratory data: *nerve conduction studies* indicated slowed conduction over the left sural nerve, absence of conduction right sural nerve and both peroneal nerves - leg to ankle. The *EMG studies* demonstrated acute denervation in sampled muscles supplied by the right femoral nerve: rectus femoris, vastus medial and iliopsoas plus muscles supplied by major components of the sciatic nerve gastrocnemius and peroneus longus. *Myelogram and CT scan* of the pelvis were normal.

Subsequent Course: The patient improved but then 9 months later has an acute exacerbation in the right leg and one year later had a similar process involving the left leg.

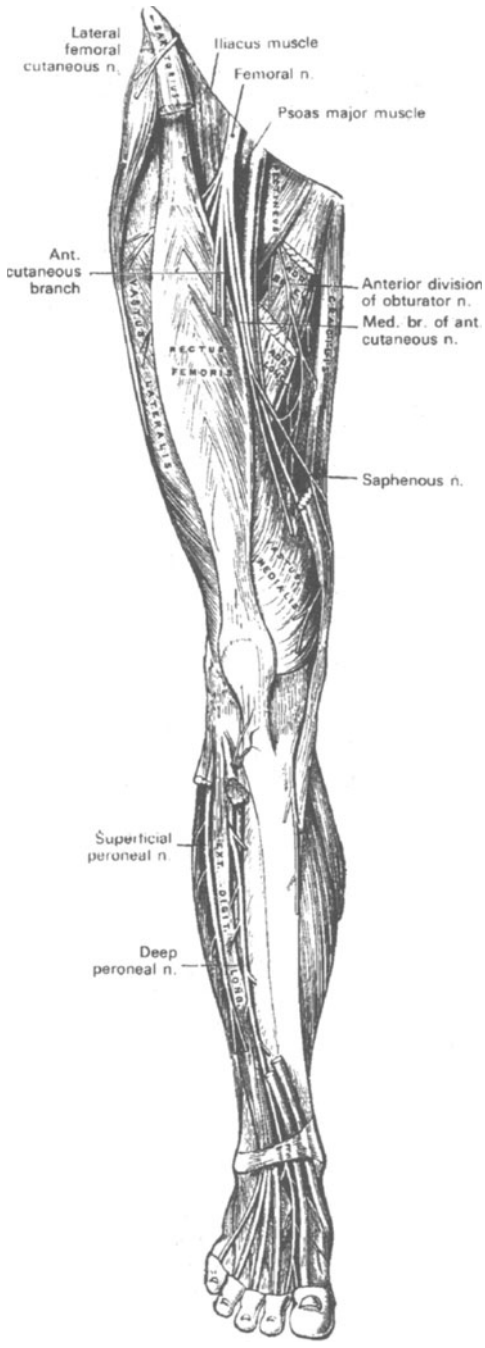


Figure 8-10. Nerves of the Right Lower Limb Anterior View: Note the emergence of the femoral, obturator and lateral femoral cutaneous nerves into the upper anterior thigh. The relatively superficial course of the common peroneal nerve as it passes around the head of the fibula from posterior - (popliteal) location to anterior calf is best seen in Fig. 8-11. (From Clemente, C.: Gray's Anatomy, 30th Ed. 1985, p. 1231).

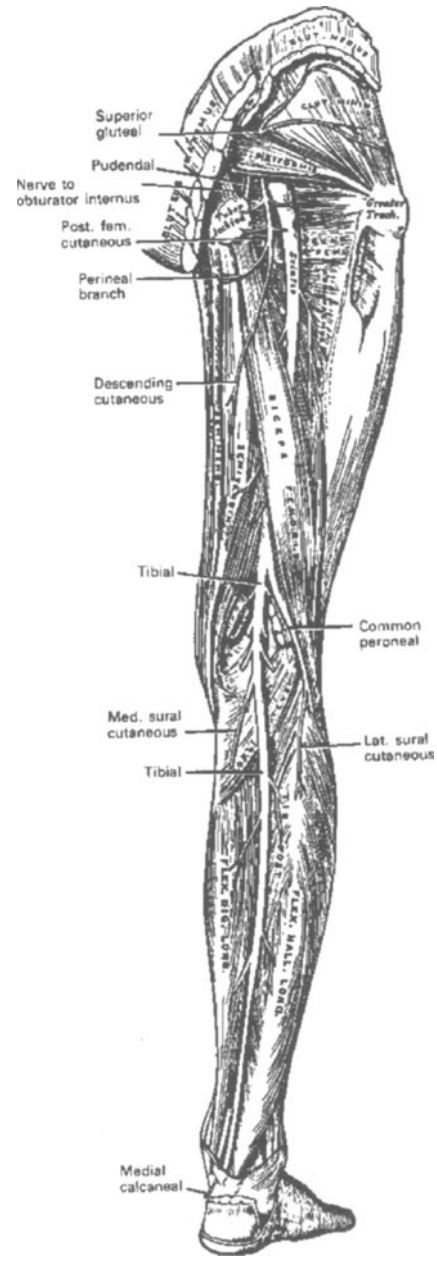


Figure 8-11. Nerves of the Right Lower Extremity Posterior View: The medial and lateral sural cutaneous nerves have been shifted in position by the dissection. Note a) The relationship of the sciatic nerve to possible sites of intramuscular injection into the buttock. b) The relationship of the tibial and peroneal nerves to the popliteal fossa, c) The relatively exposed position of the peroneal nerve in relationship to lateral calf head of fibula (not fully dissected), d) Relatively superficial position of the tibial nerve behind medial malleolus of ankle. (From Clemente, C.: Gray's Anatomy, 30th Ed. 1985, p. 1238)

Over the subsequent 9 months significant improvement occurred so that she was able to ambulate with a cane.

POLYNEUROPATHIES:

Polyneuropathies may be classified from several standpoints:

1. Predominantly motor vs. predominantly sensory vs. predominantly autonomic vs mixed.

2. Acute vs. subacute vs chronic

3. The site of pathology: a) axonal - "dying back phenomena" -distal degeneration as in metabolic diseases or arsenic intoxication. b) Segmental demyelination with the preservation of axons - as in the immunological post infectious polyneuropathy or some connective tissue disorders or chronic lead poisoning.

4. Etiology: Major categories.

a) Infection - leprosy.

b) Inflammation: Vasculitis - periarteritis.

c) Immune disorders.

d) Intoxications: Arsenic, lead, anti neoplastic agents, industrial chemical exposures)

e) Metabolic disorders: nutritional deficiencies of B vitamins, diabetes mellitus, and uremia, critical care unit neuropathies

f) Degenerations: hereditary.

g) Tumors: neurofibrobromas, schwannomas.

From a practical standpoint, the usual approach is to combine the several types of classification.

1. First classify as to acute, subacute or chronic.

2. Then classify as to predominantly motor or sensory or autonomic. The majority are mixed.

3. If possible classify as to axonal, or demyelinating based on EMG /nerve studies and in some cases; on nerve biopsy. (A survey of peripheral nerve histopathology is presented in *Fig.8-12--8-16)*

4. Then classify as to etiology.

Acute progressive motor neuropathy with variable sensory features.

The Guillain Barré Syndrome provides the most common example of an acute rapidly progressive polyneuropathy. (Acute idiopathic polyneuritis, acute inflammatory demyelinating poly infectious polyneuropathy, Landry-Guillain-Barré Disease).

This is a rapidly progressive predominantly motor peripheral neuropathy that evolves over 10-14 days and affects legs, arms and in many cases cranial nerves. In some cases there is progression to involve the muscles of respiration. Sensory symptoms are present but in most cases sensory findings are minor compared to the motor findings. In severe cases, tracheotomy and mechanical respiratory assistance may be required. Prior to the era of modern respiratory assistance, the death rate was as high as 25%.

The essential pathology consists of perivascular lymphocytic infiltrates, perivenous segmental demyelination. Nerve roots as well as peripheral and cranial nerves are involved. In more severe cases, axons as well as myelin are involved with secondary Wallerian degeneration. In very severe cases, with involvement of the proximal nerve or nerve root, the motor neuron may also be damaged and destroyed. The cerebrospinal fluid is usually without a cellular reaction but does shown an increase in protein that peaks several weeks after onset. In most cases, slowing of motor nerve conduction will be demonstrated, consistent with a demyelinating peripheral neuropathy. Acute motor axonal neuropathy is a variant in which motor axonal degeneration occurs with only minimal demyelination and inflammation. In the Miller Fisher variant or syndrome, a gait ataxia, areflexia, and ophthalmoplegia are present but limb weakness is absent and nerve conductions are normal, although CSF protein is increased. There are also sensory and autonomic variants.

The underlying pathogenesis of the syndrome is now apparent. *In 60-70% of patients, a viral respiratory or gastrointestinal infection has occurred 1-2 weeks prior to the onset of the neurological symptoms.* Other patients have experienced infectious mononucleosis, viral hepatitis, other viral illnesses, surgical proce-

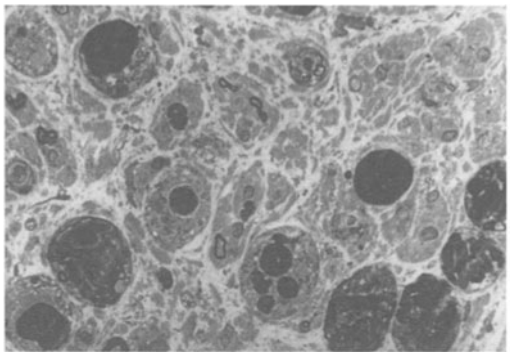


Figure 8-12. Histopathology of Peripheral Nerve I Wallerian Degeneration. Considerable necrosis and degeneration of axons, myelin, and Schwann cells has occurred. The field is dominated by large macrophages containing remnant of axons, myelin, fat globules etc. ("Digestion Chambers"). Toluidine Blue x 100 (approx.). Courtesy of Dr. Tom Smith, Neuropathology, U. Mass. Medical School.

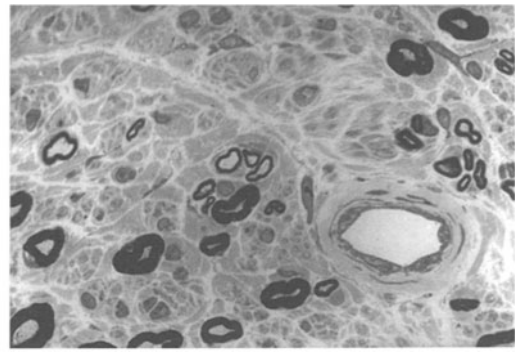


Figure 8-13. Histopathology of Peripheral Nerve II. Chronic Axonal neuropathy. A marked loss of myelinated fibers has occurred. However these are clusters of axons surrounded by a thin myelin sheaths suggesting that regeneration of axons has occurred. Toluidine Blue X 100. Courtesy of Dr. Tom Smith,

dures or immunizations - (old type of serum antibodies vaccine, swine influenza vaccine of 1976). The acute motor axonal neuropathy variant appears to follow infection with the gastrointestinal agent: Campylobacter jejuni.

The pathological and clinical manifestations of the disease are considered to reflect an immunological reaction directed at peripheral nerves. A similar syndrome - experimental allergic neuritis has been produced in rabbits and other laboratory animals 2 weeks after immunization with homogenized peripheral nerve (Waksman and Adams 1955). Sera from patients with the syndrome react with multiple antigens in peripheral nerve. The levels of IgM antibodies against peripheral nerve-myelin correlate with the disease course. Sera from some patients with Guillain Barré Syndrome will produce demyelination in neuronal tissue culture (McFarlin in Asbury and Gibbs 1990; Keski in Dyck 1993).

In more severe cases, plasmapheresis is now employed. In the cases treated by the Guillain Barré Study Group (1985) within two weeks of onset, there was a definite reduction in the length of time for hospitalization, for mechanical respiration and for resumption of walking. More recent studies have suggested that intravenous immune globulin-G maybe even more effective (for a review of current concepts see Ropper 1992). Glucocorticoid steroids usually have no significant effect although massive intravenous therapy (1000mg /day of methylprednisolone) in uncontrolled trials has been reported to have a possible effect.

The following case history 8-5 provides an example of a mild case of this syndrome.

Case 8-5: Approximately two weeks after influenza infection, this 71-year-old right-handed male awoke with tingling of the plantar surfaces of his feet, unsteadiness and weakness at the knees. Over the next 10 days, he experienced a gradual decrease in lower extremity power but no progression of the sensory symptoms. He denied low back pain, bowel or bladder symptoms.

Neurological examination: *Motor Systems:* Weakness was present in the lower extremities both proximal and distal slightly greater on right than left. *Reflexes:* A minimal dysmetria was present in the upper extremities. All deep tendon stretch reflexes were absent in upper and lower limbs except for a trace triceps. Plantars demonstrated no response to stimulation. *Sensory System:* There was a mild decrease in pain and temperature sensation over toes. Perianal sensation was normal. To a greater degree, position and vibratory sensation were decreased in the toes.

Clinical diagnosis: Guillain Barré syndrome: acute post infectious polyneuropathy.

Laboratory data: CBC, glucose, renal functions, liver functions, electrolytes, protein immunoelectrophoresis and ESR (40mm/hr) and MRI of the spine were all normal. CSF: protein was increased to 206 mg%, with 0 white blood cells and a glucose of 76mg%. Serological tests and cultures were negative. EMG/nerve conduction studies demonstrated slowing of motor nerve conduction velocity. - consistent with a demyelinating neuropathy.

Course: the patient's condition stabilized and he was transferred to a rehabilitation facility.

Other causes of the syndrome of acute progressive motor neuropathy:

1) *The exotoxin produced by the bacillus, c. Diphtheria:* pharyngeal and laryngeal muscles are affected 1-2 weeks after the pharyngeal infection followed by a general polyneuropathy: arms then legs after 4-5 weeks.

2) *Acute intermittent porphyria* is inherited as an autosomal dominant trait. A metabolic defect in the liver results in increased production of and high urine levels of porphobilinogen and its precursor delta aminolevulinic acid (involved in hemoglobin metabolism). The severe motor neuropathy may be accompanied by abdominal pain or psychosis or convulsions. Anticonvulsants, barbiturates, sulfa drugs and estrogens may trigger attacks.

3) *Toxic polyneuropathies:*

a) Triorthocresylphosphate :the so-called Jamaican Ginger polyneuropathy seen during the prohibition era reflected contamination of bootleg alcohol by this agent). b) Thallium salts.

All of these acute polyneuropathies as well as many subacute and chronic polyneuropathies are characterized by an acellular cerebrospinal fluid that often has an increased protein content.

Acute polyneuropathies are distinguished from subacute and chronic polyneuropathies. There is no clear-cut distinction, subacute may evolve into chronic over years.

Subacute Polyneuropathies:

The common causes are toxic and metabolic producing a distal symmetrical sensory, motor neuropathy often with painful feet (dysesthesia and hyperesthesia)

1) *Nutritional deficiencies of B vitamins:* thiamine, pyridoxal phosphate, and folic acid, B12, (often multiple) are frequently encountered. a) In association with alcoholism b) in chronic elderly intensive care patients –"critical illness polyneuropathy of Bolton et. al., 1993 c) in the "dry" or neuropathic form of beri beri seen in prisoner of war camps, in famine, or in underdeveloped parts of the world.

2) *Heavy metal intoxication)* Arsenic - distal sensory motor. A more acute form associated with acute arsenic intoxication also occurs .b) Lead - predominantly motor with greater involvement of the upper extremities (wrist drops due to radial nerve involvement).

3) *Hexacarbon industrial solvents* used in glues and plastic production n-hexane and methyl-n-butyl ketone (see Spenser et al 1975).

4) *Other industrial agents:* acrylamide, trichloroethylene.

5) *Medication induced*: a) Isoniazid (INH) for treatment of tuberculosis -produces a pyridoxine deficiency. b) Nitrofuradontins used for the treatment of bladder infection) Anti-neoplastic agents including: cis-platinum and vincristine d) Thalidomide no longer available once marketed as a tranquilizer e) Drugs used to prevent seizures (anticonvulsants)-phenytoin is the most prominent in this group - usually the syndrome is subclinical.

6) *Uremic polyneuropathy* occurs in 60-70% patients with chronic renal failure. The syndrome clears slowly with renal transplantation.

Chronic polyneuropathy: Common causes are as follows:

1) *Diabetes mellitus:* distal symmetrical predominantly sensory - axonal - peripheral neuropathy (see case 4 above) etiology is not clear but may involve accumulation of glucose, fructose and sorbitol in nerves). Autonomic neuropathies or painful distal neuropathies may occur. Successful pancreatic transplantation may halt progression of the disease (Kennedy et al 1993).

2) *Remote effects of malignancy:* carcinoma of lung, ovary, and breast. Mixed sensory motor or motor or selective sensory.

3) *Complications of connective tissue disorder:* periarteritis, rheumatoid arthritis. In some cases this is a complication of HIV infection.

4) *Leprosy:* the most common world wide infectious cause of peripheral neuropathy

5) *Chronic inflammatory demyelinating polyneuropathy (CIDP) Subacute-chronic* "Guillain Barré Syndrome" progressive or relapsing remitting form of the more common acute idiopathic polyneuropathy. These disorders occur on an immunological basis; components of peripheral nerve are the targets of the immune system. These neuropathies are often very responsive to the administration of steroids such as prednisone or drugs that modify the immune system (immunoglobulin G) or to plasmapheresis. (Refer to Dyck, et al 1981, and Dalakas et al. 1981). Some of these cases are associated with HIV infection. (See Cornblatt, 1981).

6) *Abnormalities of plasma proteins usually a monoclonal abnormality of immunoglobulins* (multiple myeloma, macroglobulinemia, cryoglobulinemia and benign monoclonal gammopathy).

7) *Hereditary peripheral neuropathies.*

A. Those disorders in which a specific metabolic defect has been identified. Several may be cited.

1) *Familial amyloid polyneuropathy autosomal dominant (Andrade type).* Amyloid accumulates in blood vessel walls and in the endoneurium. An often-painful syndrome begins in the adult with sensory and autonomic symptoms and slowly progresses to a full polyneuropathy over 10-15 years.

2) *Refsum Disease.* Hereditary ataxic polyneuritis (Autosomal recessive). Phytanic acid (a fatty acid) accumulates in blood and nerves. The symptoms begin in late childhood or adolescence. In addition to a peripheral neuropathy, cerebellar ataxia, a degeneration of the retina (Retinitis pigmentosa) neurogenic deafness and a degeneration of cardiac muscle (cardiomyopathy) are present in most patients.

3) *Bassen Kornsweig Syndrome* - a rare autosomal recessive syndrome which begins in infancy - the metabolic defect is a deficiency of beta lipoprotein and of cholesterol. As above, peripheral nerve, cerebellum, heart, and retina are all affected. In addition - fatty stools, (steatorrhea) retarded growth and an abnormal appearance of red blood cells (acanthocytosis) is present.

4) *Familial dysautonomia (Riley-Day Disease).* A deficiency of serum dopamine B hydroxylase, the enzyme that converts dopamine to norepinephrine results in severe abnormalities of the sympathetic autonomic nervous system. In addition, small myelinated and unmyelinated fibers are involved producing a selective loss of pain and temperature sensibilities.

5) *Porphyria* in contrast to the previous processes produces an acute syndrome (see above).

B. *Mixed sensory motor inherited polyneuropathies without a clearly defined metabolic abnormality.* In the series of 205 patients referred to the Mayo Clinic (Dyck et al 1981) with chronic polyneuropathies of unknown cause, 86 or 42% were found to have an inherited disorder of this type.

The most common disorder in this group is peroneal muscular atrophy -Charcot Marie Tooth Disease (CMT). This disease is inherited as an autosomal dominant. Distal motor involvement is prominent. Sensory fibers are less involved. There are often associated malformations of the feet pes cavus (high arches). Relatives may manifest minimal, minor or partial forms of the diseases which are sometimes asymptomatic - so called "forms fruste". The age of onset is usually in the second or third decade, occasionally later. More recent studies have suggested at least two major forms of the disease referred to as hereditary motor sensory neuropathy I & II, (HMSN I & II) and several minor forms (HMSN III-VII). (Refer to Asbury 1992, and Harding and Thomas 1980).

HMSN Type I is characterized by demyelination with the hypertrophic onion

bulb changes that are seen when demyelination and remyelination has occurred *(Fig. 8-14)*. The nerves may also be grossly enlarged. A marked reduction occurs in nerve conduction velocity. *Three subtypes have been identified: CMT 1A,CMT 1B and CMT 1C.* All of these subtypes are transmitted as an autosomal dominant and have slow conduction implying a demyelinating disorder. Type CMT 1A accounts for 60% of all hereditary peripheral neuropathies and is linked to a point mutation occurring on chromosome 17 at the PMP 22 gene segment. This gene encodes myelin protein and appears to be duplicated (Roa et al 1993). Type CMT 1B has been linked to chromosome 1.

HMSN Type II (CMT 2) has a somewhat later age of onset. Nerve conduction velocity is normal; there is no histologic evidence of demyelination. The underlying pathology is considered neuronal. Most subtypes 2A-2D have autosomal dominant inheritance

HMSN Type III -Dejerine Sotta's neuropathy - (CMT-3) is a less common but more severe recessive or autosomal dominant disorder beginning in the first decade of life with both sensory and motor features. The disorder may map to 1q or 17 p or to other sites. The essential pathologic change is demyelination with subsequent remyelination to produce the hypertrophic onion bulb formations (*Fig 8-15). Nerves are grossly enlarged to palpation;* progression is more rapid and disability greater.

Case 8-6 presented on CD ROM provides an example of a patient with autosomal dominant Charcot Marie Tooth disease and slow nerve conduction consistent with type 1A.

8) *Unknown Cause:* Even when the patient has been fully investigated in a specialized center, a specific etiology may still not be found. In the Mayo Clinic series (Dyck et al 1981) previously cited, 24% of patients remained in the unknown etiology category.

DISORDERS OF THE NERVE ROOT: RADICULOPATHY:

The major problem affecting the nerve root is compression by a ruptured disk or by osteophytes projecting into the neural foramen from the degenerative process affecting the disks (spondylosis).

These processes primarily involve the cervical and lumbar areas. These problems are among the most frequently encountered in the neurological office. In the cervical area the C 5-C 6 and the C6-C7 interspaces are most frequently involved with compression of the C6 or C 7 nerve roots nerve roots Less often the C5 or C 8 nerve roots are involved. The relationship of the disc space to the nerve root and to the spinal cord is demonstrated in *Figure 8-17.*

In the lumbar area the L 4—5 (L5 nerve root) and L5-S1 (S1 or L5 nerve root) interspaces are most frequently involved. Less often the L3-4 or L2-3 disc interspaces are involved with involvement of the L4 or L3 nerve roots.

In the diagnostic analysis of a radiculopathy the following features should be considered:

1) The acute onset of pain in the limb in a radicular distribution (Fig.8-1) with associated pain in the neck or lumbar spine.

2) In general the radicular pain is out of the proportion to the neck or back pain.

3) The radicular pain is described a sharp, shooting, burning, or electric shock or tooth ache like pain. The pain is exacerbated by maneuvers that suddenly increase pressure within the CSF space, for example coughing, sneezing or straining at stool.

4) Radicular sensory symptoms plus or minus radicular sensory findings may be pre-

Figure 8-14. Histopathology of Peripheral Nerve III Segmental demyelination: Teased nerve fiber preparation. This 74 yr. old man had a peripheral neuropathy associated with abnormality of plasma protein (monoclonal cryoglobulinemia). Courtesy of the authors, Lippa, C.F., Chad, D.A., Smith, T.W., Kaplan, M., Hammor, K.: Muscle and Nerve: 626-631, 1986 (Wiley).

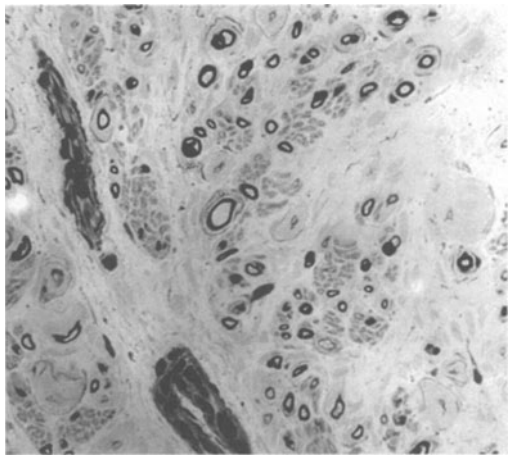


Figure 8-15. Histopathology of Peripheral Nerve IV Hypertrophic "onion bulb" neuropathy. Repeated episodes of segmental demyelination and remyelination may result in finely myelinated axons surrounded by whorls of overlapping intertwined processes of Schwann cells. In addition, this microscopic field demonstrates a significant decrease in total numbers of fibers and a significant increase in connective tissue. Toluidine Blue X63 (Approx.). Courtesy of Dr. Tom Smith.

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5) Selective weakness of muscles supplied by a specific cervical or lumbar nerve root may be present.

6) Selective depression of a deep tendon stretch reflex supplied by a specific cervical or lumbar nerve root may be present.

7) In restricted lateral disk herniations in the cervical area, no long tract sensory or motor or reflex findings should be present that is there is no evidence for an upper motor neuron lesion. In restricted lateral disk herniations in the lumbar area no bladder symptoms should be present.

8) With midline herniations in the cervical area, signs of spinal cord compression may be present and are considered below in chapter 9.

9) With midline herniations in the lumbar area, urinary retention and a disturbance of sexual functions may be present due to involvement of the cauda equina.

The usual distribution of hypalgesia and of radicular pain in the upper extremity is demonstrated in *figure 8-18.* See also figure 2-1 for the lower extremity

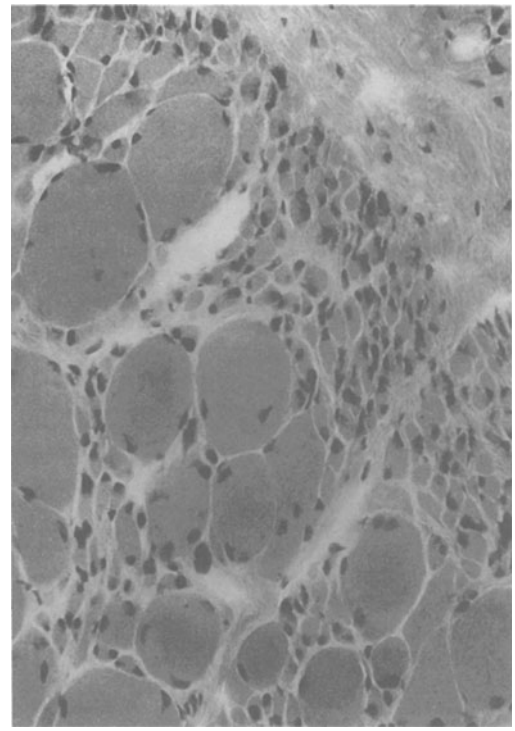


Figure 8-16. Histopathology of Muscle III Neurogenic Atrophy in a patient with Charcot Marie Tooth Disease, Grouped atrophy is evident. H&E X 63 (Approx.) Courtesy of Dr. Tom Smith.

In considering sensory symptoms as compared to sensory findings, when there is involvement of a single nerve root, it is important to note that sensory symptoms (radicular pain and paresthesias) may be present although the examination demonstrates no actual sensory deficit as regard pain or touch sensation. This is a reflection of the fact that there is an overlap of radicular sensory fields. Thus one half of the dermatome of C-7 is also supplied by C-6 and the other one half by C-8 *(Fig.8-19a). The total sensory field then for a single* nerve root is therefore more extensive than demonstrated in the standard sensory diagrams *(Fig.8-19b).*

The following case history (8-7) provides an example of lumbar root compression secondary to a herniated disk.

Case 8-7: This 37 year old right-handed white female day care worker in relationship to heavy lifting had the acute onset of persistent pain in the lumbosacral area shooting as a sharp

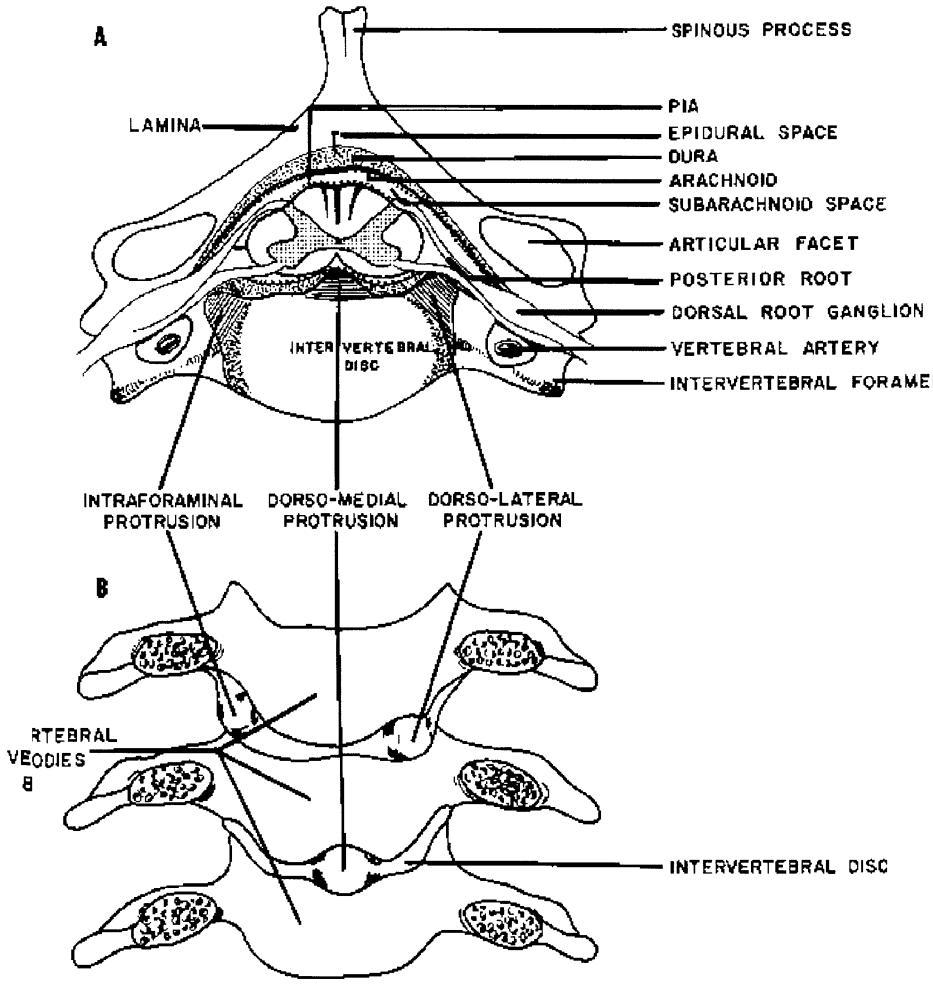


Figure 8-17. Anatomic Relationships of the Cervical Spinal Cord and the Cervical Intervertebral Discs. Lateral and midline disc protrusions are indicated. (Modified after Frykholm, Acta. Chir. Scand. 101:345, 1951).

pain into the left buttock. One month later, she developed numbness left posterior thigh and left posterior calf to the ankle. Three months later, she had the acute onset of severe pain in the lumbar area now extending to the posterior thigh and subsequently to the posterior calf as a shooting pain. Tingling paresthesias now extended into the small toes. The pain in the leg was triggered by coughing or straining at stool. Despite a prolonged period of bed rest, pain medication and non-steroidal anti-inflammatory agents, no improvement occurred.

Neurological Examination (6 months after onset of symptoms): *Motor System:* Significant weakness was present on the left at toe extensors, ankle dorsiflexors, everter and inverters with minor atrophy in the calf. *Reflexes:* The Achilles deep tendon stretch reflex was absent on the left compared to active (3) right Achilles and both patellar reflexes. *Sensory System:* Pain sensation was markedly decreased on the left side over the L5 and S1 dermatomes and to a considerable degree over S2-5. On straight leg rising, pain was present in both leg and back at 45-50 degrees on the left. *Maneuvers:* On palpation, there was marked tenderness over the left sacroiliac area and over the left sciatic nerve at the sciatic notch and over the left posterior tibial nerve behind the medial malleolus of the ankle. The patient was

in severe pain sitting, standing or recumbent.

Clinical Diagnosis: Rupture of disk with involvement of nerve roots L5, S1 and probably cauda equina.

Laboratory data: MRI, *(Fig.8-20)* demonstrated a massive rupture of the disk on the left at the L4-L5 level with inferior extension and marked compression of the cauda equina.

Subsequent course: The patient eventually agreed to removal of the ruptured disk, 5 weeks after the initial neurological evaluation. She had some relief of pain but over the next 3 years she continued to have a foot drop with significant weakness at left ankle and toes, an absent left Achilles reflex, and sensory deficits over the left L 5 distribution.

Most patients (75%) with a more limited lumbar radiculopathy respond to a period of strict bed rest for 3 to 5 days on a firm mattress and bed board, nonsteroidal anti inflammatory agents, heat or cold and possibly the temporary use subsequently of a back support. Those who fail this therapy may require local steroid injection or surgical therapy. Only if surgical therapy is a consideration should MRI studies be obtained.

Benign non-malignant back pain alone, either acute or chronic is an extremely common complaint Usually it is benign in nature. However a complete history and general physical examination is indicated to identify immediate precipitating events. In addition, the history should survey for, any past history of malignancy, or change in bowel habits, urinary pattern or menses etc. A rectal and or pelvic examination is indicated particularly when there is no clear-cut history of trauma. In the middle aged or older male patient, appropriate laboratory studies to rule out prostatic malignancy are indicated (prostatic specific antigen/antibody-PSA). If such a survey is negative and no neurological symptoms or signs are present, then neurological consultation and lumbar MRI scan are not indicated. Plain films of the lumbar spine will usually suffice to rule out orthopedic disease. Such benign back pain does not require a course of bed rest. Non

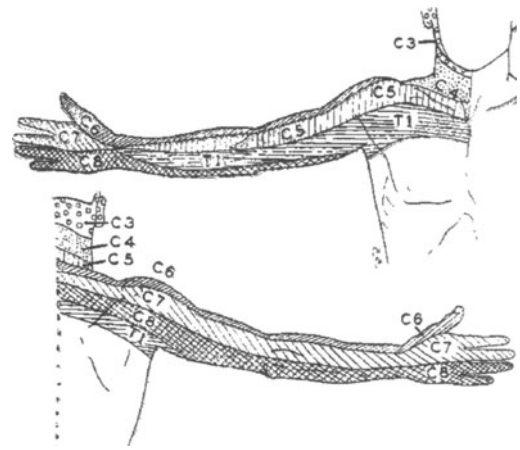


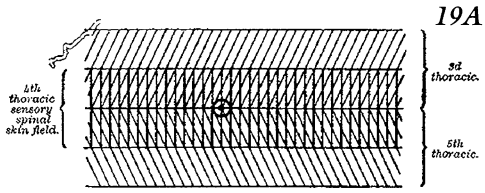
Figure 8-18 Dermatome charts of the upper extremity in man outlined by the pattern of hypalgesia, following rupture of an intervertebral disk. From Keegan, J.J., and Garrett, F.D.: Anat. Rec., 102: 417, 1948 (Wiley).

steroidal anti-inflammatory agents, modification of posture, heat or cold and weight loss if indicated are appropriate measures. Activities at work may have to be temporarily modified, use of a support belt or corset may be indicated.

For cervical radiculopathy, at home cervical traction, a cervical collar, cervical, pillow and, nonsteroidal anti-inflammatory agents are usually effective. Most (75%) patients will usually respond to these measures. When these measures fail and radicular symptoms and findings persist, surgical therapy may be considered. At such a time MRI scans is appropriate.

The following case history 8-8 demonstrates the effect of a ruptured cervical disk producing compression of a cervical nerve root.

Case 8-8: This 45 year-old right-handed married white female educational coordinator 2 weeks prior to evaluation had the acute onset pain in the neck radiating into left arm with pain and tingling in the index and middle finger of the left hand. The pain would shoot into the arm is she coughed or sneezed or strained to move her bowels. 4 months previously the patient had experienced pain in the neck extending to the left shoulder area but that symptom had cleared. In the last several days prior to consultation the patient had



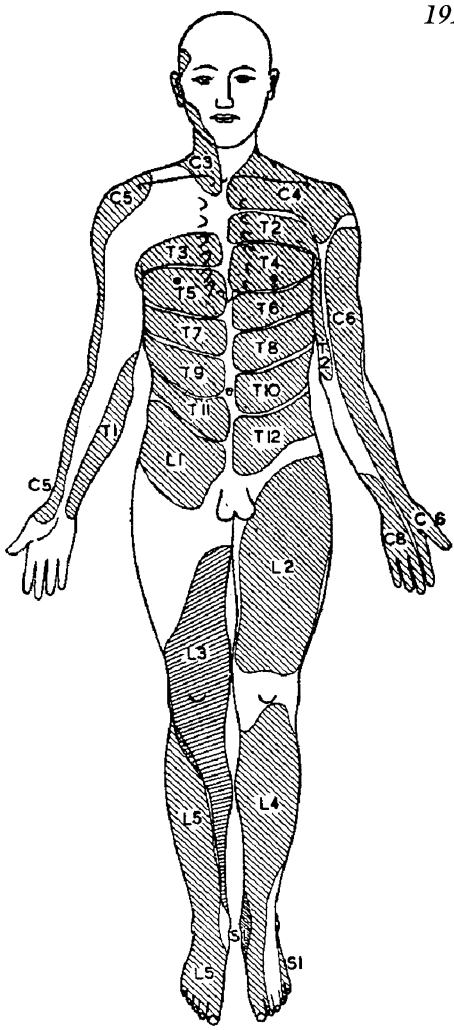


Figure 8-19 A) Method of remaining sensibility to demonstrate the sensory skin field of a nerve root. Sherrington sectioned three roots above and three below the intact root to be studied. In this diagram the overlap of the third and fifth thoracic spinal roots is demonstrated. (From Ransom, S., and Clark, S.: The Anatomy of the Nervous System, 10th Edition. Philadelphia, W. B. Saunders, 1959, p. 129).
B) Dermatomes in man demonstrated by the method of remaining sensibility. (From Lewis, T.: Pain. New York, Macmillan, 1942, p. 20, after Foerster).

developed twitching of biceps and triceps muscles. She had no leg or bladder symptoms.

Neurological examination: The left triceps deep tendon stretch reflex was absent or reversed. Pain sensation was decreased over the left index and middle fingers. Neck motion was limited for extension and rotation and there was tenderness over the spinous processes of C 7 and T1and over the left supraclavicular area.

Clinical diagnoses: Cervical 7 radiculopathy secondary to lateral rupture of disk.

Treatment: The use of cervical traction, cervical collar, nonsteroidal anti inflammatory agents, various pain and anti muscle spasm agents, epidural injection and various measures in physical at therapy for 4 weeks failed to produce any relief. Pain was now predominantly in the arm and her examination now demonstrated additionally significant weakness at the left triceps muscle. MRI *(fig 8- 21)* demonstrated a lateral disk rupture at the C 6 -7 interspace. The patient underwent a left sided laminectomy at that level with removal of ruptured disk material. She had a resolution of symptoms except for a minor residual tingling, a decrease in pain sensation over the index finger, and a mild decrease of the left triceps deep tendon stretch reflex.

Schwannomas arising from the nerve root: These tumors arise from Schwann cells of the peripheral nerve or nerve root. When the nerve root is involved the tumor may be present within the bony canal and/or external to the bony canal. The tumor may enlarge the neural foramen. Depending on the size and location of the tumor the nerve root alone may be involved *(Fig.8-22)* or the spinal cord may also be involved (refer to chapter 9). When the tumor arises from a nerve root within the cauda equina, multiple nerve roots may be involved. *(Fig.8-23).*

Tabes Dorsalis: this late complication of syphilis involving the nervous system is a result of the infectious agent, the spirochete involving the posterior (dorsal) root and to some extent the dorsal root ganglion producing secondary degeneration in the posterior columns.

Disease of the dorsal root ganglion: The

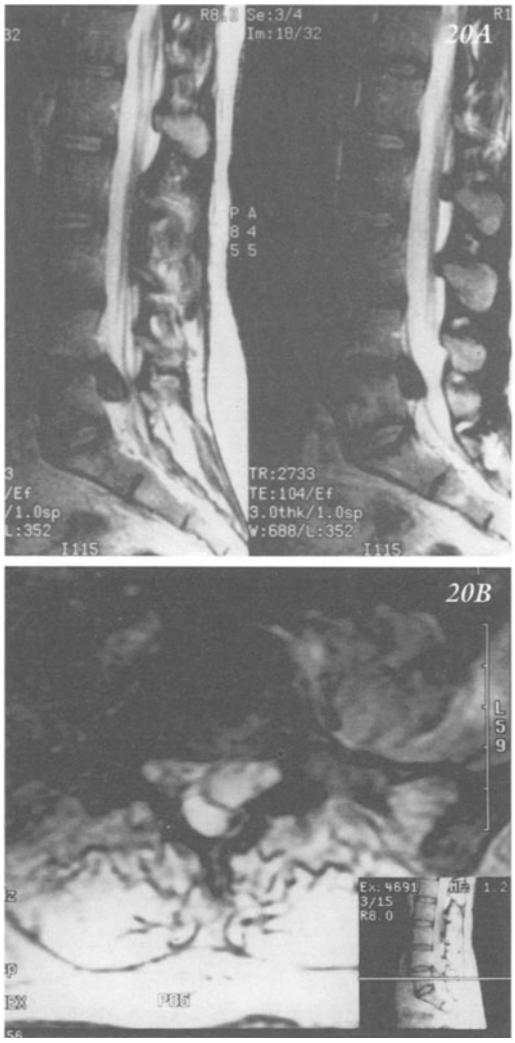
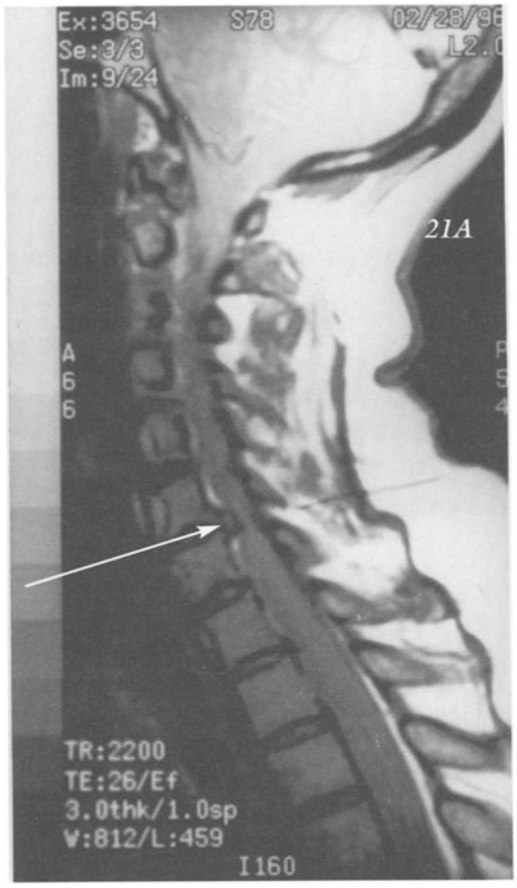


Figure 8-20: Lateral protrusion of a Lumbar disc: Case 8-7: lumbar radiculopathy MRI A) sagittal sections B) transverse section.

major disease involving the dorsal root ganglia is infection by the herpes zoster/varicella virus. In actuality this is a reactivation of a virus remaining dormant in the dorsal root ganglion after previous varicella (chickenpox) infection. Diseases that alter the immune system such as infection, e.g. HIV, or lymphomas may activate the virus and in some cases generalized zoster may occur. Ten-25% of patients with generalized lymphoma will develop H.Zoster. In addition local irritation of a nerve root or a primary neoplastic process arising in organs closely related to the dermatome may activate the virus. The clinical disorder begins with sharp lancinating pain in a radicular distribution. Within 3 or 4 days, the involved dermatome demonstrates a vesicular eruption. The thoracic dermatomes are most frequently involved. In approximately 20% of cases, cranial nerve root sensory ganglion are involved most often cranial nerves 5 (primarily the ophthalmic division) and 7. Although the dorsal root ganglion is primarily involved, occasionally the anterior root or horns are involved producing atrophy and weakness. Rarely, the spinal cord is involved, producing a clinical myelitis. The anti viral agent, acyclovir is effective in reducing the course of disease. *Post herpetic neuralgia* is the term applied to the persistent severe pain occurring in 20% of patients one month after the rash has healed. The occurrence of this chronic pain syndrome is more frequent in elderly patients; 75% of patients with H.Zoster who are over the age of 70 years continue to have severe pain one month after healing of the cutaneous lesions. Treatment with lidocaine skin patches, the tricyclic antidepressant nortriptyline or the anticonvulsant gabapentin or oral opioids may produce a moderate reduction in pain in some of these patients. A recent study has demonstrated the effect of intrathecal methylprednisolone in patients who were still refractory at one year (Kotani et al 2000).

The herpes simplex virus may also remain dormant in the sensory root ganglion and may be activated to produce radicular pain. With the type 2 virus (vaginalis), the sacral segments are often involved, and urinary retention may occur.



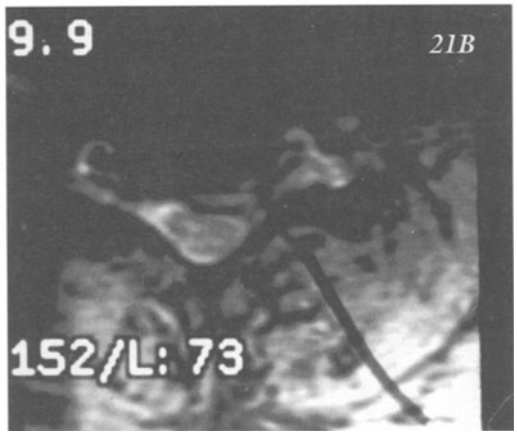


Figure 8-21: Lateral protrusion of a Cervical disc: Case 8-8: Cervical radiculopathy. MRI A) sagittal section B) transverse section.

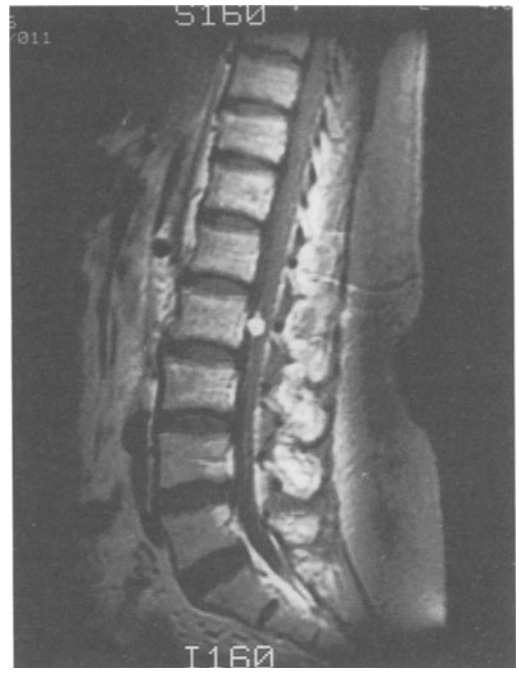


Figure 8-22: Schwannoma of nerve root at L2 level. MRI. This 53-year-old female had an 18-month history of progressive low back pain that began to radiate to the right foot with numbness of the lateral aspect of the right foot and numbness of left thigh. Examination demonstrated only decreased pain sensation of the left anterior thigh and lateral aspect of the right foot. An MRI of the lumbar spine demonstrated a circumscribed Schwannoma at the lumbar-2 level, subsequently removed by Dr. Alex Danylevich.

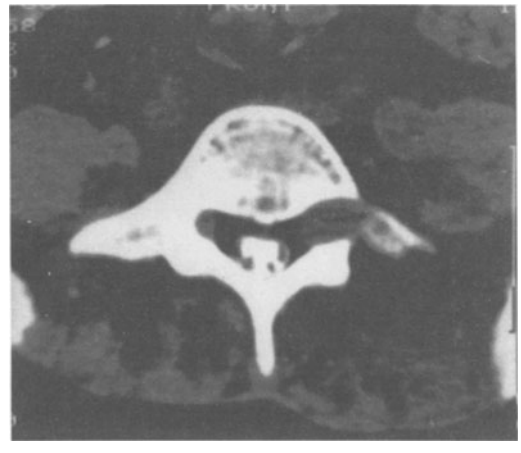


Figure 8-23. This 23-year old female had multiple neurofibromas. In this CT scan a Schwannoma at the lumbar level is demonstrated.