CHAPTER 30

Neoplasms of the Joint

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- The most common primary neoplasms of the joint are pigmented villonodular synovitis and synovial chondromatosis and a diagnosis is best made using magnetic resonance imaging (MRI).
- Other primary lesions are rare and include lipoma arborescens, synovial hemangiomas, intracapsular chondromas, and synovial chondrosarcomas.
- Secondary neoplasms of the joint are synovial sarcoma and giant cell tumors.
- The malignancies that metastasize to bone also may invade the articular space.

Although some neoplasms originate in the joint, others penetrate or metastasize to it. Pigmented villonodular synovitis and synovial chondromatosis are the most common proliferative disorders arising from within the joint. Other primary lesions are rare and include lipoma arborescens, synovial hemangiomas, intracapsular chondromas, and synovial chondrosarcomas. Synovial sarcoma and giant cell tumors are neoplasms that tend to extend into the joint. The malignancies that metastasize to bone also may invade the articular space.

PRIMARY NEOPLASMS OF THE JOINT

Pigmented Villonodular Synovitis

Pigmented villonodular synovitis (PVNS) is a rare proliferative disorder of unknown etiology that affects the synovial lining. PVNS does not exhibit cellular atypia, but there is recent evidence of cytogenetic abnormalities. Yet the presence of synovitis suggests an inflammatory process. The etiology of PVNS remains unresolved. Regardless, it is characterized by inflammation and deposition of hemosiderin in the synovium (1). It occurs in three forms: an isolated lesion involving the tendon sheaths (giant cell tumor of the tendon sheath); a solitary intra-articular nodule (localized PVNS); and a diffuse villous and pigmented lesion involving synovial tissue (diffuse PVNS) (2,3). This section focuses on the latter two forms.

The typical presentation is a 20- to 40-year-old patient who complaints of a traumatic swelling of a single joint (4–9). The knee is involved 80% of the time. Some patients may experience pain, warmth, and stiffness in the joint (7,8,10). Mechanical symptoms, such as locking and instability, may develop, particularly if the joint contains a large pedunculated nodule (11). The symptoms typically are episodic or slowly progressive (7). Results of laboratory studies, such as a complete blood count and erythrocyte sedimentation rate, are within normal limits and can help exclude infection and rheumatoid arthritis. Aspiration of the joint reveals a brown, red, or yellow fluid (7,9,12).

During the initial stages, plain radiographs reveal periarticular synovial swelling, absence of synovial calcification, normal bone density, and preservation of the cartilage space (13). Bone changes develop in the later stages. Recent evidence suggests that tissue expression of matrix metalloproteinases in PVNS contributes to the destruction of bone and cartilage often seen in PVNS (14). In joints with small synovial volumes (e.g., the hip), the synovial villi may abut the bone and cause subtle erosions. As the villi grow, pressure within the joint capsule increases. The villi then invade the bone and juxta-articular cysts appear (15,16). If the disorder is not diagnosed and treated, joint destruction can ensue.

Due to deposition of hemosiderin, a magnetic resonance image (MRI) typically will show nodular foci of decreased signal on both T1 and T2 images (Figures 30-1, 30-2) (12). Additionally, low signal on fast field

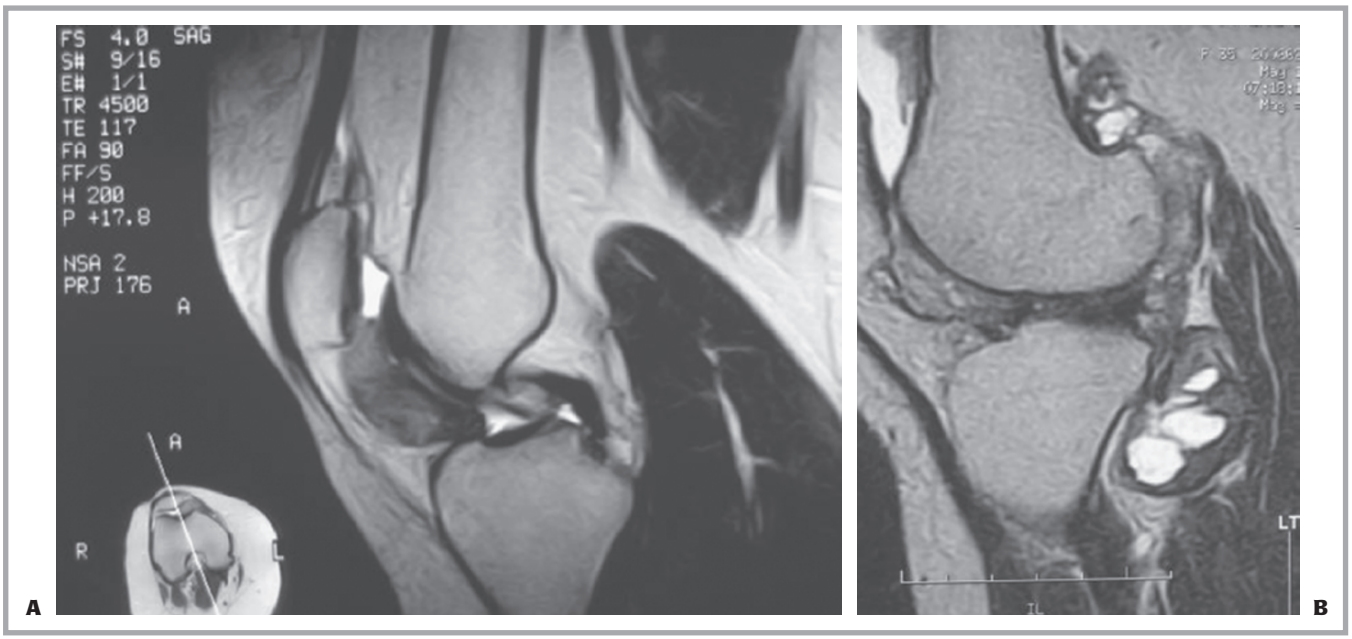

FIGURE 30-1

(A) T2 magnetic resonance imaging (MRI) sequence of heterogeneous mass anteriorly in the knee consistent with nodular PVNS. (B) T1 MRI of pigmented villonodular synovitis (PVNS) showing extensive involvement posteriorly with hypointense areas consistent with hemosiderin.

echo (FFE) sequences are helpful in the diagnosis of PVNS. Joint effusions will be present, and commonly bone erosions will be identified in PVNS of the hip, ankle, elbow, and wrist (17). In cases of localized PVNS, the MRI will show the single nodular mass (6). It also may show the extent of the disease, which helps the surgeon plan an appropriate treatment.

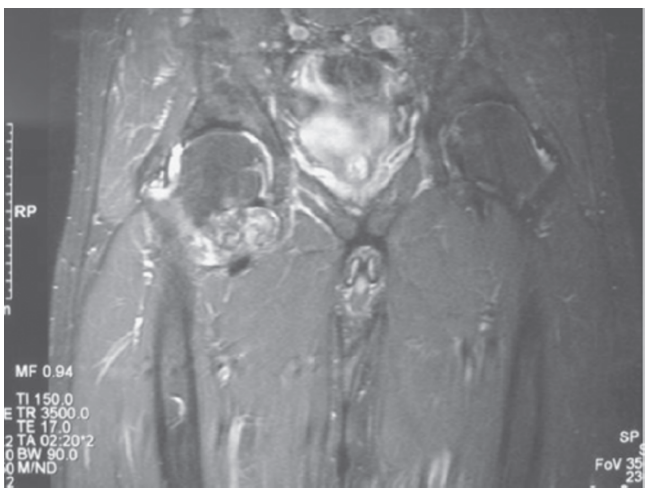

FIGURE 30-2

Magnetic resonance image showing extensive synovium and nests of cartilaginous loose bodies.

If the diagnosis remains in question, an arthroscopic exam can show the gross appearance of the lesion. In its localized form, PVNS appears as a solitary yellow, pedunculated nodule. The surface often is lobulated and cuts with a "buttery" feel. It often occurs on the anterior aspect of the knee and is similar in appearance to giant cell tumor of the tendon sheath (6,8). In the diffuse form, the thickened synovium contains folds of villi and sessile or pedunculated nodules. The entire joint appears to be covered with brown and orange seaweed. The nodules have been described as grape-like masses protruding in the joint cavity. They typically are friable and bleed with minimal trauma. Some of the villi contain bulbous ends and give the appearance of a straggly beard. Other villi possess fine points and look like ferns. The villi may invade bone or, less commonly, extend beyond the joint capsule and into the extra-articular soft tissue (4–6,18). A biopsy can confirm the diagnosis of PVNS. All three forms of PVNS share a similar histology, characterized by hypercellular subsynovial connective tissue (19). The synovial lining is one to three layers thick and outlines the nodules and villi. In some areas, the ends of the villi fuse to form clefts. Histologically, the subsynovial stroma contains collagen-producing fibroblast and phagocytic histiocytes. These cells are polyhedral and contain pale nuclei and abundant cytoplasm. They tend to proliferate and may be visualized in their mitotic

stage. Some histiocytes will phagocytize hemosiderin; some will fuse to form multinucleated cells; and others will form foam cells. The hemosiderin-laden macrophages give PVNS its rusty brown color and are more common in the diffuse form. The lipid-filled foam cells account for the yellow color that dominates the localized form. The foam cells and hemosiderin-laden macrophages tend to localize to the periphery, and giant cells tend to be scattered throughout the areolar tissue (Figure 30-1) (19).

Good clinical outcomes can be obtained with local excision of a solitary nodule (6). However, once diffuse pigmented villonodular synovitis is diagnosed in a young patient, a total synovectomy is recommended (9,12). If the MRI shows the lesion to be accessible by arthroscopy, arthroscopic resection may be worthwhile, as it is associated with relatively low morbidity (12). However, an open synovectomy may be necessary because lesions typically extend beyond the reach of the arthroscope (2,3,9). In the past, synovectomy has been associated with recurrence rates as high as 40%. An incomplete synovectomy has been cited consistently as the main cause (2,7,9,16). Similar conclusions are found when arthroscopic synovectomy is performed. The level of synovectomy for diffuse PVNS is critical in reducing the recurrence (20). Following open synovectomies via an anterior and posterior approach in knees with PVNS, an 8% recurrence rate and minimal morbidity was noted (9). If diffuse PVNS is diagnosed in an older patient with degenerative joint disease, an arthroplasty can give excellent results. In a young patient, arthrodesis may be considered as a savage procedure.

Radiation therapy has been advocated as an adjuvant to surgery to control this disease process. In a prospective study, external-beam radiation combined with partial (anterior) arthroscopic synovectomy approached success rates of total synovectomy. The authors recommend the use of adjuvant external-beam radiation at anti-inflammatory doses of 2600 cGy for treatment of diffuse PVNS of the knee when subtotal synovectomy is performed (21). Intra-articular injection of yttrium 90 has been used to treat diffuse PVNS. Studies regarding its effectiveness are inconclusive, and its use remains experimental. It may prove useful as an adjunct to subtotal excision in extensive lesions in which complete excision would result in unacceptable morbidity. Prior to its use, patients ought to be informed that it can impair tissue healing, exacerbate stiffness, and possibly cause sarcomatous degeneration (7,9).

Synovial Chondromatosis

Synovial chondromatosis is a benign metaplastic disorder that occurs when subsynovial mesenchymal cells mature into chondroblasts instead of fibroblasts. Rather than producing collagen, these cells form nodules of cartilage. These nodules initially expand within the loose areolar tissue, and then protrude into the joint cavity that is covered only by the synovial lining. Eventually, the cartilaginous nests are extruded and form loose bodies (Figure 30-2). Nourished by the synovial fluid, the chondroblasts continue to multiply. As the loose body enlarges, its central portion loses contact with the nutritional source and dies. The necrotic area then calcifies (22).

On gross exam, the synovial lining appears swollen because it contains multiple nodules of hyaline cartilages (Figure 30-3). These masses are of various sizes and are of a translucent whitish-gray color. The microscope shows the cartilage nests to be in different stages of maturity. Occasionally, a capillary may invade some of these areas and allow endochondral ossification to occur (22).

Synovial chondromatosis occurs in three phases (23). During the initial phase, there are no loose bodies, but metaplastic activity occurs within the synovium. Loose bodies appear during the intermediate phase. During the final phase, metaplastic activity ceases, but multiple loose bodies persist.

The disease most often afflicts those in their third or fourth decades and occurs twice as often in men. It is almost always monarticular, and only rarely is it isolated to a bursa or tendon sheath. It affects the knee joint more than 50% of the time, although the hip, elbow, shoulder, ankles, and other joints may be involved (22,24). Swelling, discomfort, and decreased range of motion are the most common symptoms. As the disease progresses, such mechanical symptoms as locking and giving way may develop (24,25). Eventually, the pedunculated cartilaginous masses and loose bodies can destroy the joint surfaces and lead to more severe symptoms.

During the early stages, plain radiographs may show a nonspecific soft tissue mass, due to the presence of

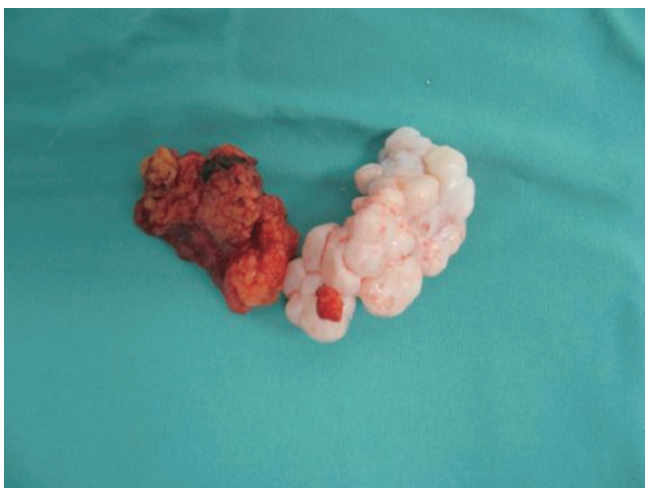

FIGURE 30-3

Gross photograph of cartilaginous loose bodies with surrounding synovium seen in synovial chondromatosis.

nonmineralized cartilage. Bony erosions may be seen, secondary to focal pressure. During the second and third stages, multiple juxta-articular calcifications or loose bodies are seen (Figure 30-4). These nodules typically are of a similar size and uniformly scattered. Joint space narrowing, osteophytes, sclerosis, and abundant calcified loose bodies represent end-stage disease (26).

If osteonecrosis, rheumatoid arthritis, post-traumatic arthritis, or degenerative arthritis is noted, a diagnosis of secondary synovial chondromatosis must be considered. This diagnosis is especially likely if the presence of the disorder preceded the diagnosis of chondromatosis. In contrast to the primary form, secondary synovial chondromatosis shows fewer ostechondral bodies, which vary more in size, and does not recur or show histologic atypia (11,26,27).

Magnetic resonance imaging can help define the location of the cartilaginous nodules and is the best noninvasive study to confirm a diagnosis of synovial chondromatosis. An intermediate density on T1 sequences and an intermediate-to-high density on T2 sequences characterize the immature nodules (Figure 30-5). Calcified and ossified areas appear hypointense on both T1 and T2 images. The exception occurs when a loose body contains marrow fat, which appears as a hyperintense area on T1 (26–29).

Synovial chondromatosis is treated by removal of the loose bodies and excision of all abnormal synovium. Stiffness may occur, and recurrence rates have been reported as high as 11% after open treatment. In rare cases, the lesion may transform into a chondrosarcoma (25–27). Synovial chondromatosis of the hip can be treated arthroscopically assisted. The use of the arthroscope to address the intra-articular component of the disease avoids dislocation of the hip and is less invasive. Arthroscopically assisted synovectomy and removal of loose bodies has been shown effective by some surgeons (30). Similarly, arthroscopy for chondromatosis of the shoulder has been used with anticipated advantages of more complete removal of loose bodies, decreased postoperative pain, and enhanced rehabilitation (31). Dysregulation of the hedgehog signaling pathway has been implicated in the many benign cartilaginous tumors. Increased expression of the hedgehog transcription factor in mice recreates synovial chondromatosis. Medication-induced blockage of the hedgehog signaling pathway may be a future treatment option (28).

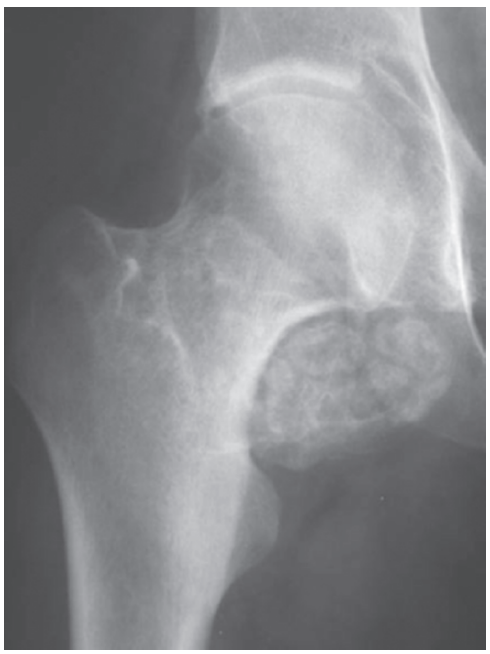

FIGURE 30-4

Plain radiograph of synovial chondromatosis showing calcified cartilage in the hip joint.

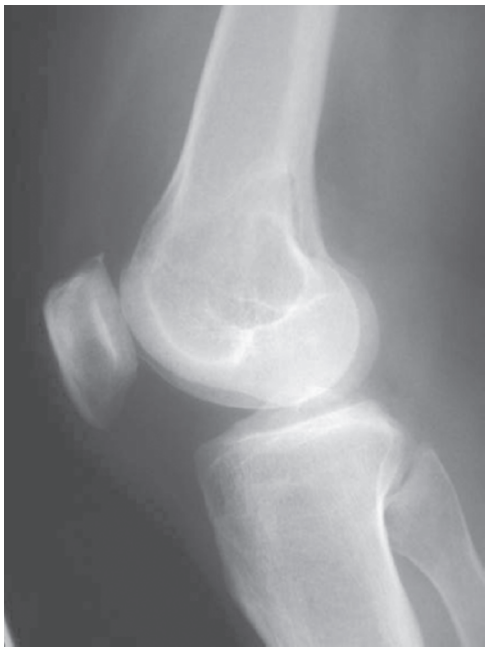

FIGURE 30-5

A lateral radiograph demonstrating a radiolucency involving the distal femoral epiphysis. Biopsy demonstrated tissue consistent with a giant cell tumor of bone.

Other Primary Joint Tumors

A solitary intra-articular lipoma may occur but is extremely rare. More commonly, excessive intra-articular adipose tissue is due to lipoma arborescens. This entity involves fatty synovial villi and often is associated with osteoarthritis, rheumatoid arthritis, and trauma. It usually occurs in the knee and causes joint swelling and pain (29). Synovectomy often is curative.

Synovial hemangiomas usually occur in children and young adults and almost exclusively involve the knee. Plain films often show the pathognomonic phleboliths. Histologically, it is identical to the soft tissue hemangiomas. Both the localized and diffuse forms can cause pain and hemarthrosis. This benign vascular neoplasm is treated by surgical excision (29,32).

Intracapsular solitary chondromas, like extra-articular chondromas, are benign cartilaginous neoplasms that may calcify. They may present as firm intra-articular mass.

Synovial chondrosarcomas are exceptionally rare and may be primary or secondary to synovial chondromatosis. Treatment is wide surgical resection (27).

SECONDARY JOINT NEOPLASMS

Synovial Sarcoma

Synovial sarcoma is an uncommon, highly malignant tumor involving mesenchymal cells. It typically occurs near tendon and fascial planes, although, on rare occasions, it may arise within or adjacent to a joint (22,33). The lower extremities are affected most frequently and the incidence is highest among those between the ages of 15 and 40 years (32). Although synovial sarcoma suggests a relationship to normal synovium, the disease is rarely found intra-articularly. However, there have been case reports of solely intra-articular involvement of synovial sarcoma (34).

Patients typically present complaining of a slowly growing soft tissue mass. Approximately 50% of the time, the lesion is described as painful. Plain radiographs often reveal a large, lobulated, juxta-articular mass. Calcification is seen in up to one third of cases and often has a diffuse speckled appearance. MRI shows nonspecific characteristics, but can narrow the diagnosis and define the lesion's anatomic location (29).

A biopsy often is required to confirm the diagnosis and will show the sarcoma to be one of the three types. The biphasic form is the most common and involves obvious epithelial and mesenchymal differentiation. The plump cuboidal or tall columnar epithelial cells line mucin-filled clefts and cystlike spaces. The round and oval epithelial cells form nests and cords. The fibroblasts are spindle-shaped and may be arranged in a manner similar to that seen in fibrosarcoma. Sometimes the field is dominated largely by either the epithelial cell (rarely) or the fibroblast (more commonly). The lesion then is categorized as monophasic. The monophasic form can be confused with other neoplasms of fibrous or epithelial origin and is thought by some to carry a worse prognosis. A rare, poorly differentiated type, represented histologically by numerous mitotic round cells, also has been described. Rapid growth and a very poor prognosis characterize this form (22,32,35).

SYT-SSX2 is the fusion product of translocation (X,18) found in a vast majority of synovial cell sarcomas. The fusion product has now been identified to regulate beta-catenin recruitment to the nucleus which subsequently regulates cell adhesion. Identification of the fusion product by molecular diagnostics in synovial sarcoma is becoming a standard (36).

Once synovial sarcoma is diagnosed, wide surgical resection with removal of any affected lymph nodes is indicated (32). Although adjuvant radiation and chemotherapy have improved the overall prognosis, the risk of regional and pulmonary metastasis remains high. Reports have shown the 5- and 10-year survival rates to be 55% and 40%, respectively (37). Increased age, tumor size greater than 5 cm, and 10 or more mitotic figures per 10 high-powered fields are thought to increase the risk of metastasis and/or death (33).

Giant Cell Tumor

Giant cell tumor is a benign, but locally aggressive, tumor of unknown origin that most commonly affects 20- to 40-year-olds. This lesion involves the knee (distal femur and proximal tibia) 50% of the time (32), and the distal radius and proximal humerus are the next most common sites. Plain radiographs show a purely lytic lesion that begins in the epiphysis and abuts the articular surface (22). It frequently extends into the joint (Figure 30-6) (36), tends to recur, and, 1% to 2% of the time, it will become malignant and metastasize to the lungs. The addition of phenol, bone graft, and methylmethacrylate to marginal resection can decrease the recurrence rate and allow the joint to be preserved. The use of a high speed burr also can decrease the rate of

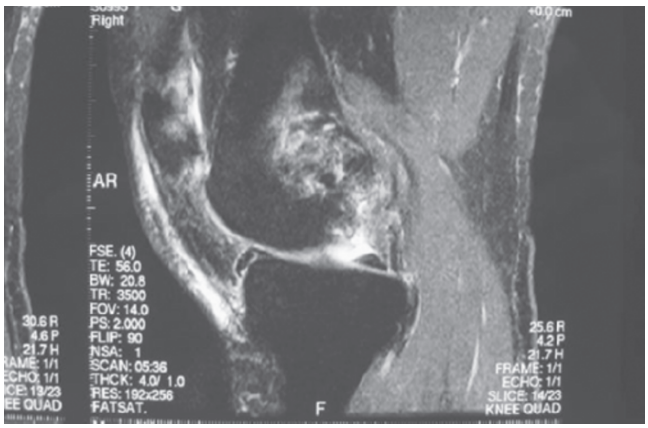

FIGURE 30-6

A sagittal fast spin echo (FSE) image of the distal femur demonstrating a distal femoral giant cell tumor of bone extending into the articular space along the cruciate ligaments.

recurrence. Radiation therapy should be reserved for inoperable tumors, as it is associated with malignant transformation (32).

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