A Congenital Scoliosis Case Characterized with Contralateral Hemivertebrae

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22.1 Introduction

Congenital scoliosis is a complex spinal problem that may end up with a disaster if not treated properly. Pathology contains many challenges in every step starting by the patient's admission.

Aim of this case presentation is to discuss a complicated hemivertebra case with its diagnosis, natural history and treatment options.

22.2 Case Description

A 2-year old boy who complains of back deformity admitted to outpatient clinic. There was no other system abnormality in his history. However, elder brother was operated because of congenital scoliosis and hemivertebra excision was done 2 years ago. Physical examination revealed right sided loin and left sided rib humps (Fig. 22.1) and no neurology. P-A long cassette X-Ray demonstrated left T10 and right L2 hemivertebrae that were fully segmented according to CT scan (Fig. 22.2). Magnetic resonance imaging of whole spine showed no intraspinal anomaly (including syrinx, Chiari malformation, diastematomyelia) and spinal cord was ended at the level of L1.

Posterior hemivertebra excision, short segment pedicle segment fixation and fusion was done for the L2 hemivertebra at first session. Postoperative period was uneventful and same procedure was repeated for the T10 hemivertebra 3-month later (Fig. 22.3).

Surgical Technique Resection of hemivertebra is performed by a single stage posterior approach as mentioned, with fusion of the adjacent levels only by using pedicle screw fixation. The posterior elements of the spine and, in the thoracic spine, rib head of hemivertebra on convex side are exposed subperiosteally at the affected levels. One level above and one level below 4 mm diameter pedicle screws are inserted. Then, transverse process of hemivertebra at the convex side is excised (together with rib head at thoracic spine) and body of the hemivertebra is dissected retroperitoneally (or via extrapleural approach) with the finger or Harrington's elevator. This allows us better orientation and less blood loss. Lamina and pedicle of index level is removed with Kerrison rongeur or high-speed burr. Exciting nerve root should be protected at lumbar level, but it can be sacrificed at thoracic level. After control of epidural bleeding body of hemivertebra is resected with adjacent discs and cartilaginous end plates. Compression of the pedicle screws at the convex

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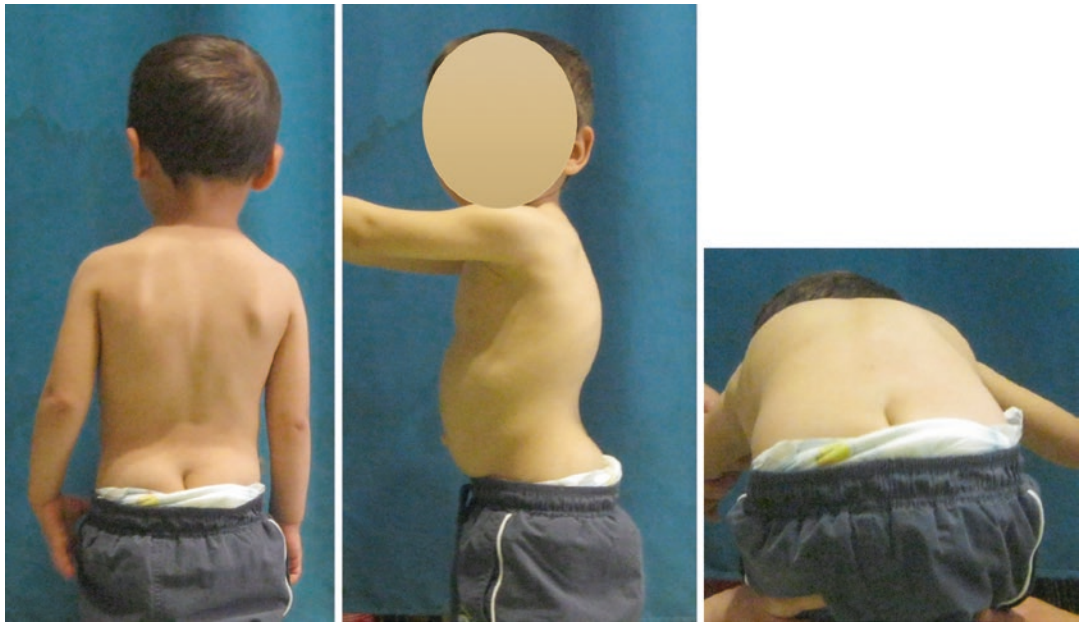

Fig. 22.1 Clinical pictures of 2-year-old boy. Forward bending test shows a right sided loin and left sided rib hump

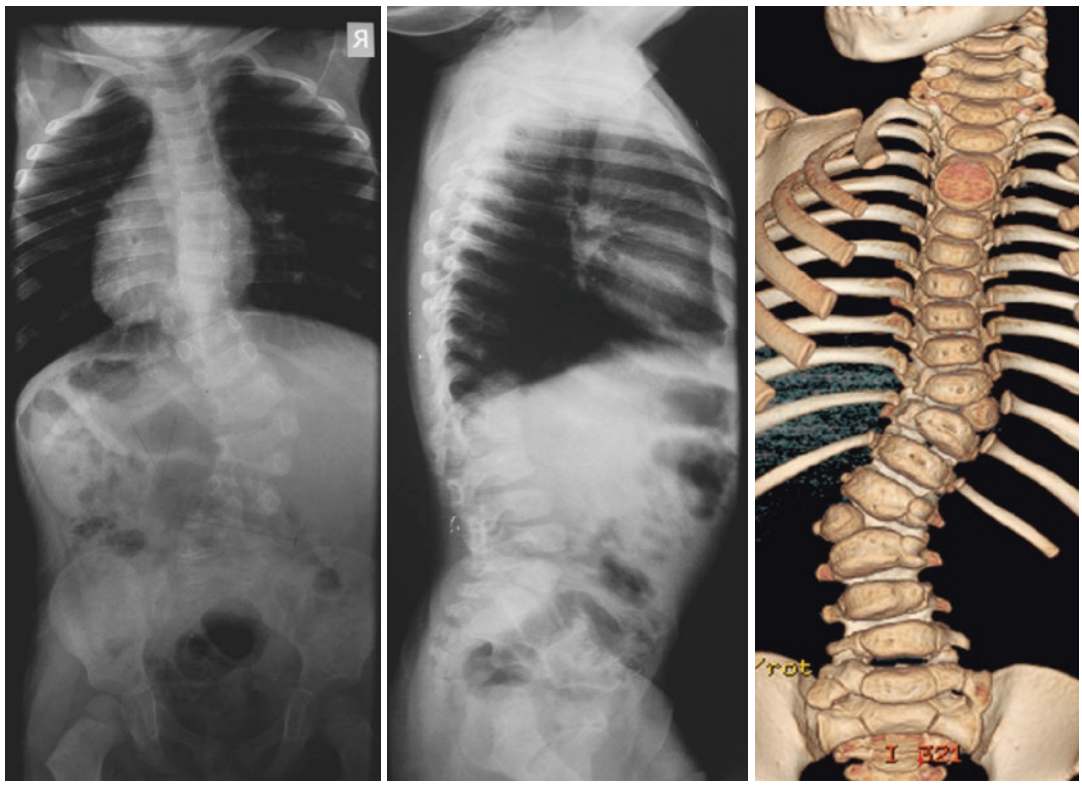

Fig. 22.2 P-A and Lateral long cassette X-ray and 3D reformatted CT scan (c) of the patient show contralateral hemivertebrae at T10 and L2 levels

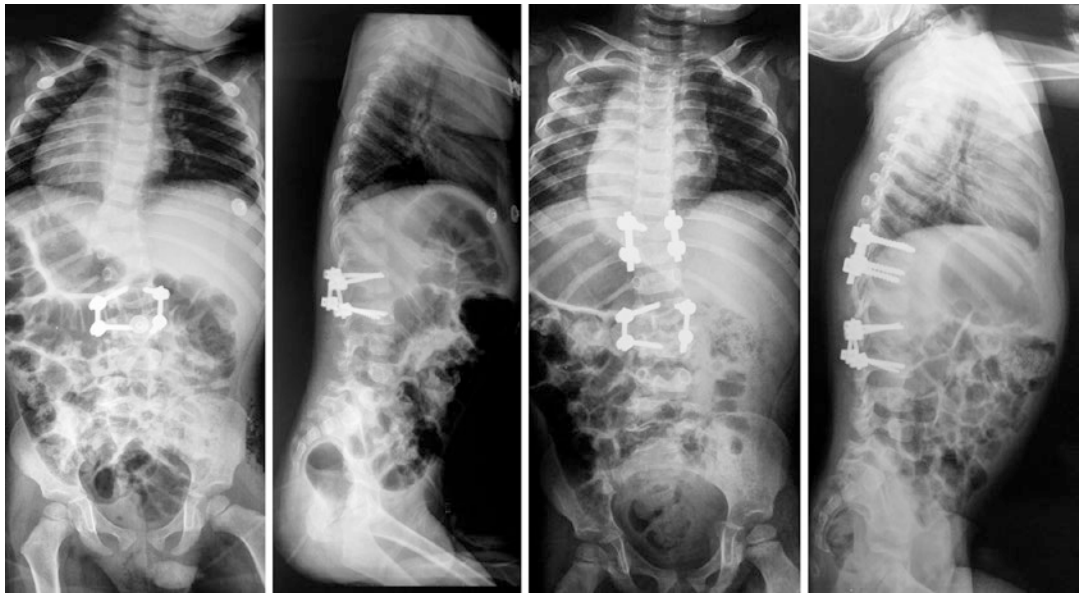

Fig. 22.3 Postoperative P-A and Lateral X-ray examinations of both sessions. There was an apparent shoulder imbalance in the control X-ray of first session which was corrected by the contralateral hemivertebra resection with second operation

side is done by the assistance of application three-point bending to the trunk of the patient. This prevents screw loosening due to direct excessive stress. Local bone is used for one level fusion (Fig. 22.3). Patients can be mobilized freely with a TLSO. TLSO can be weaning of 3 months, postoperatively.

There was no any postoperative complication. Patient has been following for 6 years and now he is 8-year-old with a balanced spine in all three planes (Fig. 22.4).

22.3 Discussion of the Case

Congenital scoliosis is the most frequent congenital deformity of the spinal column. It can be classified as defect of formation or defect of segmentation, yet most malformations have combined features of these deformities. Hemivertebra should be defined under the failure of formation group [1]. Natural history which is important for decision making differs according to anatomic location of hemivertebra. It has been well defined that locations at lumbar and thoracolumbar junction have bad prognosis and, presumably, progress. However, hemivertebra located in middle and upper thoracic spine is compensated well and may not need operation [2]. Morphology of the deformity also an important factor for prognosis. Worst scenario for the prognosis is fully segmented hemivertebra (two growth plates) and contralateral unsegmented bar. Estimated progression rate as fast as 10° a year according to Mc Master. Table 22.1 shows the hierarchic ranking from bad prognosis to good prognosis [2–4].

There is a consensus in the literature that hemivertebra should be treated as soon as possible after the diagnosis if it is located certain anatomic regions. Early treatment prevents extension and structural differentiation of deformity and also compensatory curves. Posterior resection, short level instrumentation and fusion is accepted a gold standard by the current literature [5, 6].

Preoperative whole spine MRI is crucial for the patients who planned the surgical intervention since the intraspinal anomalies are frequently possible

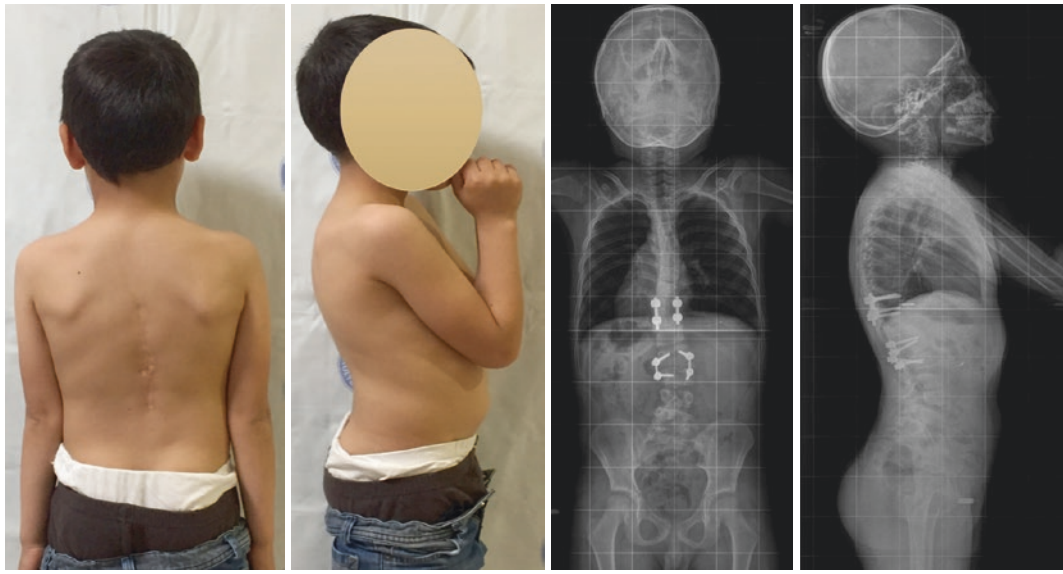

Fig. 22.4 Acceptable correction is maintaining after 6-year

Table 22.1 Grading of morphologies from the worst to bad prognosis

Fully segmented hemivertebra with a contralateral unsegmented bar
Unilateral unsegmented bar
Two consecutive fully segmented hemivertebrae
Fully segmented hemivertebra
Semisegmented hemivertebra
Wedge vertebra
Incarcerated hemivertebra
Block vertebra

Adapted from McMaster and Arlet et al. [2–4]

with congenital deformities [7]. We also performed whole spine MRI in order the check spinal cord anomalies. CT scan also was taken for detailed evaluation of pathoanatomy and found that there were only two contralateral hemivertebra without segmentation defect (Fig. 22.2). Hemivertebra excision was performed to L2 level. The first decision was a possible follow-up for the contralateral hemivertebra located at T10 level. However, clinical and x-ray examination revealed an uneven shoulder balance after the index operation (Fig. 22.3). T10 hemivertebra was also excised for preventing further coronal decompensation. Mid-term follow-up showed a balanced spine (Fig. 22.4).

22.4 Conclusions and Take-Home Message

In conclusion, congenital scoliosis is a complex spinal problem that needs detailed preoperative work out. Immediate surgical intervention should be done in special circumstances. Parents should be informed well for possible need of remedial interventions.

Pearls and Pitfalls

- Advanced imaging studies should be done for possible intraspinal anomaly and detailed anatomic interpretation
- Early surgical intervention should be done hemivertebrae located lumbar and thoracolumbar junction
- Surgeon's competency is crucial for this intervention
- Patient should be followed up closely until the end of adolescent period

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