Congenital costo-vertebral fibrous band and congenital kyphoscoliosis: a previously unreported combination

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Abstract Congenital kyphoscoliosis (CKS) results from abnormal vertebral chondrification. Congenital fibrous bands occur in several locations with variable impact on vertebral development. We report a previously unreported case of a female infant with CKS presenting with an L2 hypoplastic vertebra and a costo-vertebral fibrous band extending to the skin in the form of a dimple. We also describe the therapeutic approach, consisting of surgical excision of the fibrous band and postoperative fulltime bracing, with a 7-year follow-up. We recommend a high index of suspicion in any unusual presentation of CKS and insist on case by case management in such cases.

Keywords Congenital · Kyphoscoliosis · Fibrous · Bands

Introduction

Congenital kyphoscoliosis (CKS) results from abnormalities in vertebral chondrification centers [1]. Genetic causes [2, 3] have been identified and CKS may be seen in hereditary and polymalformative syndromes. CKS was also found in isolated idiopathic vertebral malformations [4]. Congenital fibrous bands have been widely reported in all body areas, specifically near the spine in congenital torticollis [5–7] and thoracic outlet syndrome. To our knowledge, no etiological association between congenital fibrous

bands and CKS has been reported. We report a previously unreported case of CKS caused by congenital fibrous bands, with description of the tailored surgical treatment and 7-year follow-up of this patient.

A 10-month-old female infant presented to our clinic with a limb length inequality and trunk deformity. She is the product of a full-term pregnancy and C section delivery without perinatal complications. There was no history of consanguinity or spinal or limb deformity in the family. She was diagnosed at birth with a left posterior supra iliac skin dimple in the concavity of a trunk deformity, as well as a limb inequality. Indication for convex hemiepiphysiodesis of her congenital scoliosis was made in another institution and the child was referred to our facility for a second opinion (Fig. 1).

On physical examination, the skin and subcutaneous tissue surrounding the dimple were hard, producing an irreducible tethering of the last ribs to the iliac crest. Neurological examination was unremarkable.

Radiographic measurement of lower extremities shows a slightly taller left limb (4 mm). Spinal radiographs showed anterior and lateral hypoplasia of L2 with a 28° scoliosis and 36° kyphosis (Fig. 2).

Lumbar MRI showed a fibrous band extending from the skin dimple to the concavity of a vertebral deformity, with an apical trapezoidal L2. The MRI report mentioned a possible associated fusion of the posterior lamina and left pedicles of L1 and L2 which we had difficulty to confirm or identify properly. No medullar abnormality is seen (Figs. 3a–d, 4a, b).

At the age of 11 months, surgical excision of the fibrous band was carried out with no intervention on the spine, with satisfactory postoperative results. Exposure was done with a left lateral transverse approach. The subcutaneous band was palpated and removed en bloc along with the skin

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Fig. 1 Left lumbar skin dimple

dimple (pathology showed fibrous tissue only) with the discovery of a tissue compaction anterior to the body of L2. This compaction was removed with preservation of the spinal nerve (tested by a nerve stimulator).

Treatment with an anti-kypnosis Boston type brace was decided with clinical and radiographic periodical evaluation. 7 years following surgery, lumbar kyphosis is fully corrected with a rectangular shaped L2 vertebra on lateral radiographs and no evidence of fusion between L1 and L2 vertebrae. A 20° right lumbar and 20° contralateral thoracolumbar scoliosis persist. The brace is still worn full time with bi-annual follow-up and brace adjustment. The child has normal psychological and motor development.

Discussion

Skin dimples in the lumbar region are often markers of neurological anomalies such as tethered cord syndrome and may hide dermal sinuses, all associated with neurological manifestations [8–10]. This association has embryological implications, as the skin and neurological tissues have the same ectodermal origins [11] and defects in separation between these tissues yield skin anomalies accompanying the deep spinal cord anomalies. In this case, the skin dimple hid no tracts, and did not reach the spinal cord. In addition, the spinal cord did not present any anomaly with the MRI clearly showing a fibrous magma condensing lateral to the body of the L2 lumbar vertebra, away from the spinal cord.

The L2 vertebra in this case is articulated with L1 and L3 with a missing part in the anterior lateral aspect of the vertebral body. This raises the question as to whether L2 in our patient is a hemivertebra. Although the MRI report mentioned a possible fusion of the posterior lamina and left pedicles of L1 and L2, there was no evidence that it existed

Fig. 2 Preoperative scoliosis series showing 28° scoliosis and 36° kyphosis

Fig. 3 **a, b** Coronal MRI preoperative series. The *arrow* indicates the fibrous packing. **c** Sagittal MRI preoperative series. The *arrow* indicates the fibrous packing at the kyphoscoliosis concavity apex.

d Axial preoperative MRI series, the *arrows* indicate the fibrous band extending from the skin dimple to the lateral aspect of L2 vertebra

all along the patient's follow-up, since gradual improvement of the curve was noted.

Some hypotheses on hemivertebrae claim that they are caused by an absence of formation of the hemivertebra after the intersegmentary mesenchymatous condensations have formed, which prefigure the future vertebra, or by a vascular related atrophy and disappearing of the hemibody in the mesenchymatous state [12–14]. The remaining part of the vertebra will cause a mainly scoliotic or kyphoscoliotic deformation of the spine. In the kyphoscoliotic form, only a posterior part of the vertebra is formed, contrary to the hypoplastic vertebra in our patient, in which the whole L2 vertebra is present, except for a slice of the anterior lateral vertebral body. These findings along with the presence of the fibrous costo-iliac band forming a magma in the missing part of the L2 vertebra made us challenge the hemivertebra diagnosis in our patient, hence, the therapeutic approach.

Orthopedic treatment yielded various improvement rates in congenital kyphosis and kyphoscoliosis due to hemivertebrae [15], and is considered in cases with compensatory curves and long flexible curves with few congenital anomalies [1].

The stronghold of hemivertebrae treatment is surgical, with three described procedures. Hemivertebrectomy [1, 16] convex epiphysiodesis with or without instrumentation [17, 18] and transpedicular epiphysiodesis with posterior unilateral instrumentation [19]. Based on the rationale that the fibrous band contributes to the spinal deformity, we chose to excise it before intervening on the spine. The good response to band excision and brace treatment confirmed our preoperative hypothesis. However, it is difficult from this case to know if surgical excision alone without bracing would have led to the same correction, but we deliberately decided to use it to increase the chances of curve improvement.

Fig. 4 **a** Current total spine AP radiograph, out of brace, showing a left 20° thoracolumbar and 20° right lumbar curvature. **b** Current total spine lateral radiograph, out of brace

Conclusion

Congenital kyphoscoliosis may also be caused by extra-vertebral anomalies such as a tethering fibrous band. Surgical excision of the fibrous band along with bracing may be enough to treat the condition, in opposition to conventional CKS where bracing is ineffective and surgery is often required.

Conflict of interest None of the authors has any potential conflict of interest.

References

1. Jaskwicz D, Ali RM, Patel TC, Green DW (2000) Congenital scoliosis. *Curr Opin Pediatr* 12(1):61–66
2. Horan GS, Kovács EN, Behringer RR, Featherstone MS (1995) *Dev Biol* 169(1):359–372
3. Mark M, Rijli FM, Chambon P (1997) Homeobox genes in embryogenesis and pathogenesis. *Pediatr Res* 42(4):421–429
4. Bergoin M, Giraud F, Philipp N, Ayme S, Bollini G, Gennari JM, Turcat Y (1988) Congenital scoliosis in homozygote twins. Apropos of a case report. *Rev Chir Orthop Reparatrice Appar Mot* 74(Suppl 2):115–118
5. Lee JJ, Lim SY, Song HS, Park MC (2010) Complete tight fibrous band release and resection in congenital muscular torticollis. *J Plast Reconstr Aesthet Surg* 63(6):947–953
6. Tatli B, Aydinli N, Caliskan M, Ozmen M, Bilir F, Acar G (2006) Congenital muscular torticollis: evaluation and classification. *Pediatr Neurol* 34(1):41–44
7. Porter SB, Blount BW (1995) Pseudotumor of infancy and congenital muscular torticollis. *Am Fam Physician* 52(6):1731–1736
8. Ackerman LL, Menezes AH (2003) Spinal congenital dermal sinuses: a 30-year experience. *Pediatrics* 112(3 Pt 1):641–647
9. Ponger P, Ben-Sira L, Beni-Adani L, Steinbok P, Constantini S (2006) International survey on the management of skin stigmata and suspected tethered cord. *Childs Nerv Syst* 26(12):1719–1725
10. van Aalst J, Beuls EA, Cornips EM, van Straaten HW, Boselie AF, Rijkers K, Weber JW, Vles JS (2009) The spinal dermal-sinus-like stalk. *Childs Nerv Syst* 25(2):191–197

11. Bollini G, Jouve JL (2000) Malformations congénitales du rachis. Encyclopedie médico-chirurgicale. Elsevier Masson SAS, France
12. Kaplan KM, Spivak JM, Bendo JA (2005) Embryology of the spine and associated congenital abnormalities. *Spine* 30(5):564–576
13. Nakajima A, Kawakami N, Imagama S, Tsuji T, Goto M, Ohara T (2007) Three-dimensional analysis of formation failure in congenital scoliosis. *Spine* 32(5):562–567
14. Marks DS, Qaimkhani SA (2009) The natural history of congenital scoliosis and kyphosis. *Spine* 34(17):1751–1755
15. Dubousset J, Zeller R, Miladi L (1992) Le traitement orthopédique dans la pathologie vertébrale du petit enfant. Encyclopedie médico-chirurgicale. Elsevier Masson SAS, France
16. Aydogan M, Ozturk C, Tezer M, Mirzanli C, Karatoprak O, Hamzaoglu A (2008) Posterior vertebrectomy in kyphosis, scoliosis and kyphoscoliosis due to hemivertebra. *J Pediatr Orthop* 28(1):33–37
17. Wild A, Jäger M, Koch H, Webb JK (2002) Treatment of congenital scoliosis in an 8-month-old child. *Arch Orthop Traum Surg* 122(7):418–420
18. Noordeen MH, Garrido E, Tucker SK, Elsebaie HB (2009) The surgical treatment of congenital kyphosis. *Spine* 34(17):1808–1814
19. Ginsburg G, Mulconrey DS, Browdy J (2007) Transpedicular hemiepiphysiodesis and posterior instrumentation as a treatment for congenital scoliosis. *J Pediatr Orthop* 27(4):387–391