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Case Report

Malformative Scoliosis: About a Case

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Abstract

Congenital scoliosis is an irreducible, three-dimensional spinal deformity, the main component of which is rotational.

They are often associated with other malformations, which must be investigated during management.

Imaging is part of the diagnostic and follow-up work-up, including X-rays of the entire spine, CT scan with reconstruction to better delineate vertebral anomalies, and MRI to visualize nerve structures.

Keywords: *Congenital; Scoliosis; Vertebral puzzle; Malformations*

Introduction

Congenital scoliosis is defined as a spinal deformity secondary to a congenital vertebral malformation that can generate three-dimensional spinal deformities. The discovery of a congenital spinal deformity in a patient raises a number of questions, including its evolutionary potential. Clinical examination and imaging are essential to assess the evolutionary potential of these spinal deformities.

Case Report

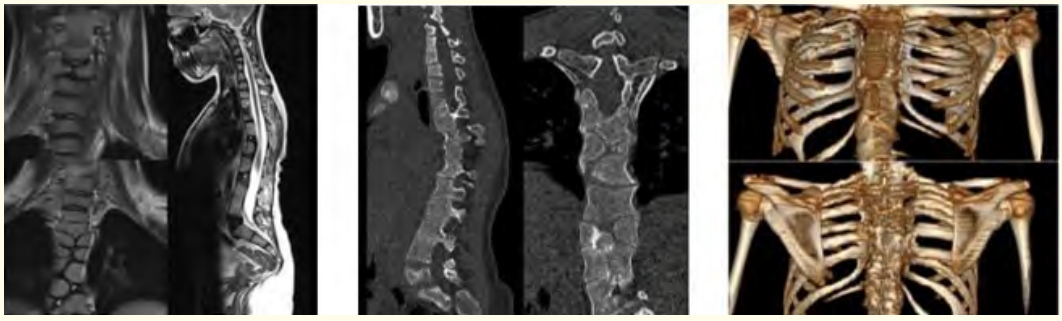

Figure 1

A 15 year old patient, presented with scoliosis and tingling in the right upper limb, with suspected malformative scoliosis and spondylodiscitis. A CT scan of the spine and a spinal MRI were performed, (figure 1) showing the presence of scoliosis with minor lumbar curvature and left convexity, with the vertebra at the eastern vertex: L2, the upper limit: L1 and the lower limit: L5.

In the cervical region, there is a compressed, butterfly-shaped appearance of the vertebral body of C4, agenesis of C7, vertebral blocking of C5-C6 and overlapping of the spinous processes of C5, C6 and D1.

In the dorsal region, there is a vertebral block at the CV level of D2-D3-D4-D5, a butterfly wing aspect of the CV D4 D5, D7 and D9, and a hemi-vertebral aspect of D6, D8.

In the thoracic cavity: synostosis of the posterior arches of right K3 K4 K5 K6 K7, of the posterior arches of right K8 K9, of the posterior arches of left K4 K 5, and of the posterior arches of left K6 K7, agenesis of right K11 and K12, bifid aspect of right K9 and left K7, and S-shaped deformity of left K3.

In the lumbar region, there is vertebral block of L1-L2-L3, compression of L4, sacralization of L5, agenesis of the transverse processes of L1, and failure of the sacral holes of the last left sacral pieces to close.

Together, these signs form a vertebral puzzle, with a combination of anomalies that cannot be classified according to WINTER's classification.

A second classification by Kawakami has been described, according to which the patient presents with malformative scoliosis with compensated longitudinal imbalance.

Spinal cord MRI shows no signs of spondylodiscitis.

Discussion

Congenital scoliosis is a three-dimensional spinal deformity involving longitudinal and rotational imbalances due to cell migration abnormalities in the embryonic sclerotome. Diagnosis is usually made in the first two years of life, with an incidence of 0.5 to 1/1,000 births worldwide.

The causes of these congenital anomalies of the spine are still unclear, but it has been reported that there is a 5-10% risk of CS being genetic in origin.

Spinal anomalies may be isolated or associated with other syndromes such as VACTERL syndrome.

Diagnosis should involve a simple standard X-ray of the front and side to measure the Cobb angle of the curve in order to assess the CS and study its evolution.

This is followed by a CT scan to assess complex anomalies prior to surgery, in order to identify anatomical features and locate bony malformations in the surgical area. Analysis of the thorax and lung highlights deformations of the thoracic wall: costal synostoses, rib hypoplasia or agenesis, intracanal protrusions through the root foramens, etc.

In order to classify all the different vertebral anomalies, they are first classified into scoliosis caused by longitudinal imbalance and scoliosis caused by rotational imbalance, the latter being divided into vertebral traction anomalies, vertebral thrust anomalies and mixed anomalies. The longitudinal imbalance group is divided into four groups which are scoliosis caused by: segmentation failure, formation

failure, mixed defects and unclassifiable complex defects.

Some patients may present with several combinations of anomalies at the same time: hemi vertebrae, bars, butterfly vertebrae (vertebrae with two hypoplastic ossification centers in the vertebral body, giving a butterfly appearance on a frontal X-ray) and even costal fusions in certain spondy-locostal dysostoses. These multiple anomalies, which cannot be systematized, are sometimes referred to as the 'vertebral puzzle', as in the case of our patient.

MRI allows exploration of the spinal cord from the cranio-occipital hinge to the terminal cone. It has been shown that curvatures that reach a Cobb angle greater than 20° before the age of 10 have a prevalence of spinal cord abnormalities ranging from 17.6 to 26%. Because of this high incidence, screening MRI of the spinal cord is recommended in patients with early-onset spinal deformity and curvatures greater than 20°, even in the absence of abnormal neurological findings on physical examination.

Congenital scoliosis encompasses a range of abnormalities with different patterns of presentation and progression. The complexity of the disease makes its management difficult and variable. The ultimate aim of treatment, whether observational or surgical, must be to prevent progression of the curvature while achieving balance of the spine. In order to achieve these objectives, the age of presentation, the location and the nature of the deformity are all taken into account.

Over 70% of congenital scoliosis develops aggressively and requires surgery. However, it is known that certain deformities such as block vertebrae and wedge hemivertebrae can be managed conservatively.

The essential criterion for choosing the right time for surgery is the extent of the scoliotic curve. This is assessed by measuring the Cobb angle. Up to 40°, the patient is carefully monitored periodically, every 4 to 6 months. Above 40°, surgery is required.

Four surgical principles are applied: fusion with or without instrumentation, convex hemipysiodesis, vertebrectomy in the case of hemivertebra and rib distraction.

Conclusion

Congenital scoliosis is defined as a three-dimensional spinal deformity present from birth.

Diagnosis is usually made within the first two years of life but may be delayed in the absence of clinical symptoms.

The type of malformation determines the potential evolution of congenital scoliosis.

Medical imaging is essential for diagnosis and follow-up, as is the search for other associated malformations.

Management can be either abstention and monitoring by imaging in the majority of cases, or surgical treatment, which is reserved for patients whose deformity is progressing uncontrollably [1-6].

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