Pediatric Oncall Journal
DOI: https://doi.org/10.7199/ped.oncall.2023.4

PEDIATRIC ONCALL
CHILD HEALTH CARE

IMAGES IN CLINICAL PRACTICE

ONE TOO MANY BONES

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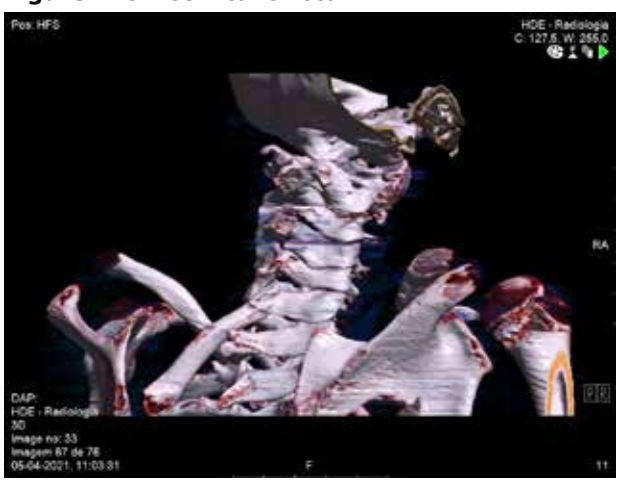
KEYWORDS
Sprengel deformity, Klippel-Feil Syndrome, shoulder asymmetry, omovertebral bone

ARTICLE HISTORY
Received 13 November 2021
Accepted 20 December 2021

A 6-year-old girl with a personal history of global development delay, epilepsy and cardiopathy was referred to the Orthopaedic Department due to shoulder asymmetry. She complained of functional limitation in daily activities, no pain registered and, recently, frequent falls with lower extremity weakness. On physical examination, the left shoulder and scapula were at a higher level and limited abduction was noted on the left shoulder. Spine x-ray revealed toraco-lombar scoliosis. Cervical computed tomography (CT) and magnetic resonance with three-dimensional reconstruction revealed multiple vertebral malformations and an omovertebral bone that runs from the superior internal border of the scapula to the posterior arch of C6 (Figure 1). She required a surgery were an 8 cm omovertebral bone exeresis was performed, without complications. On follow-up, neurologic symptoms resolved and the patient described no pain and improved shoulder abduction mobility and cosmetics.

Figure 1. 3D Cervical CT scan



What is your diagnosis?

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Sprengel deformity is a congenital elevation of the scapula.[1,2,3,4,5] Although rare, it is the most common congenital malformation of the shoulder.[1,3,4,5] It is unilateral in approximately 90% of the cases3 with a predilection for left side[1,4] and it has a male predominance (3:1).[1,3] It occurs as a result of failure of the scapula to descend to its correct position during intrauterine development1-5 being characterized by abnormal position of scapula and dysplasia with muscular atrophy[1,3]; its cause is still unknown.[3,4]

Even though the deformity appears randomly, familial cases have been reported.[1,4]

It can present as a single entity or associated with other syndromes, the most common being Klippel-Feil Syndrome[1,3,5] – an anomaly characterized by congenital fusion of a variable number of cervical vertebrae – present in about 19-27% of patients with Sprengel's deformity.[1,3] These associated vertebrae malformations can be explained by the common embryonic origin of the scapula and cervical spine.[3]

In its most severe form (in 25-50% of the cases), there is an omovertebral connection where the scapula is connected to the cervical spine by a congenitally anomalous ossification (fibrous, cartilaginous and/or osseous connection), leading to a stiff and elevated scapula with limited range of motion.[2,3,5]

Other linked bony and spinal cord abnormalities include spina bifida, congenital scoliosis and diastematomyelia.[1,2] In this case report, multiple vertebral anomalies suggestive of Klippel-Feil Syndrome was found as well as left hemi-medular myelomalacia in C4-C5. Anteroposterior X-ray imaging can confirm the diagnosis. However, computed tomography and magnetic resonance scans with three-dimensional reconstruction are nowadays used to detail the degree of the deformity, evaluate the omovertebral connection, diagnose concomitant abnormalities and plan appropriate treatment.[3,4,5]

Treatment can be conservative or surgical. Conservative treatment is reserved for patients with minor deformity and no functional limitation in daily activities.[1,3] Surgery is considered for patients with severe limitation of scapular function, neck pain or to improve cosmetic.[1,3,5] It is usually performed between the ages of 3 and 8 years old.[1,3,5]

In this case report, due to the intracanal omovertebral

bone localization, local myelomalacia and left hemiparesis, she needed surgical correction. An 8 cm omovertebral bone exeresis was performed, without complications. On follow-up, neurologic symptoms resolved and the patient described no pain and improved shoulder abduction mobility and cosmetics.

Compliance with ethical standards

Funding: None

Conflict of Interest: None

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