



(CASE REPORT)



Deformation of Sprengel in children: About three cases and review of the literature

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Abstract

We report three cases of congenitally elevated scapula or Sprengel deformity diagnosed and followed in our hospital. This very rare congenital deformation of the shoulder girdle is characterized by more or less deformed and abnormally high shoulder blades. It is often found in young children, causing functional and aesthetic problems. When surgery is indicated, imaging examinations are recommended to diagnose structures from the scapula to the cervical spine, ossified (omo vertebrae) or non-ossified (fibrous and/or cartilaginous junctions). Ultrasound is readily available and, above all, easy to perform for young children. The CT scan is essential to look for an omovertebra and associated vertebral anomalies. Magnetic resonance imaging is useful for evaluating possible fibrous and/or cartilaginous components.

Keywords: Deformation; Malformation; Sprengel; Elevation; Scapula

1. Introduction

Congenital elevation of the scapula (SCS), called Sprengel deformity or malformation, is characterized by a dysmorphic scapula in a high position. This rare congenital condition, of unknown etiology, is the consequence of a cessation of caudal migration of the scapula during embryogenesis. In 25 to 50% of cases, a bony structure (also called “omo - vertebral bone”), fibrous and/or cartilaginous comes between the scapula and the cervical spine [1,2]. Imaging is then essential to confirm the existence of this omovertebral formation and generally relies on CT scan. The latter allows us to better appreciate the ossified component, unlike MRI and ultrasound and its relationship with the scapula and spine. To our knowledge, the contribution of ultrasound has only been evaluated once in the literature [1]. Through this observation, we report the aspects encountered in imaging and highlight the interest of imaging in this condition.

2. Case 1

Four-year-old girl referred for pediatric surgery consultation for shoulder asymmetry. The clinical examination revealed an ascension of the left shoulder compared to the contralateral side without a scoliotic attitude, demonstrated by the tomography (Fig. 1 and 2), the medial angle of the left scapula was located between the transverse processes of the second and fourth dorsal vertebrae.

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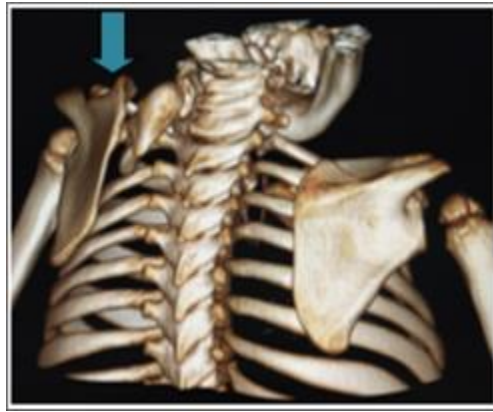


Figure 1 3D reconstruction showing the omovertebral bone between the left scapula and the cervical spine (arrow)



Figure 2 Coronal section showing abnormal ascension of the left scapula : grade 1 according to the Rigault and Pouliquen classification (arrow)

3. Case 2

16-year-old girl with shoulder asymmetry from a young age. A scoliotic attitude was clinically detected two years ago (Fig. 3). The anteroposterior radiograph of the left shoulder revealed an elevation of the left scapula (Fig. 3). Chest CT scan showed an elevated and dysmorphic left scapula (Figs. 4 and 5).



Figure 3 Clinical appearance of elevation of the left shoulder and frontal x-ray of the shoulder showing abnormal rise of the scapula

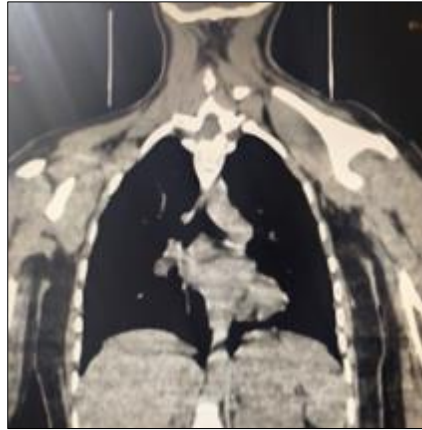


Figure 4 Coronal reconstruction showing a raised left scapula compared to the contralateral side without omovertebral bone

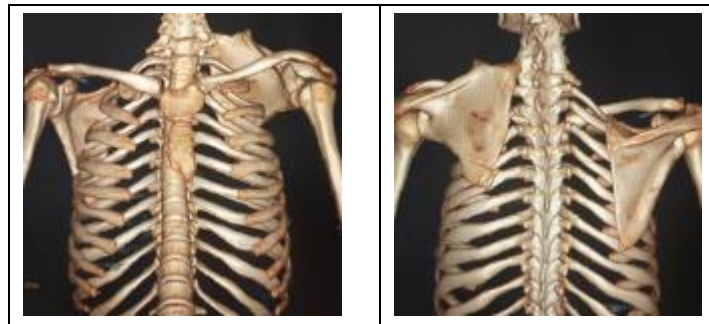


Figure 5 3D reconstruction showing a raised left scapula compared to the contralateral side without omovertebral bone

4. Case 3

Four-year-old boy with a vicious attitude of the right shoulder. Chest CT scan showed an ascended and dysmorphic right scapula with a medial angle of the scapula between the transverse processes of the second and fourth dorsal vertebrae, a bifid rib and a synostosis between the sixth, seventh and eighth right ribs (Fig. 6). There was no vertebral block or omo -vertebral bone.

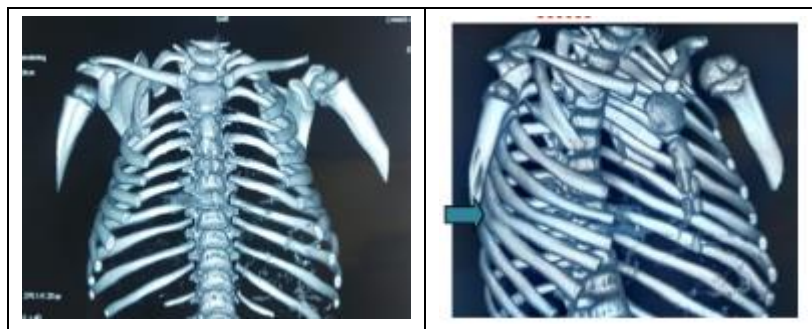


Figure 6 3D reconstruction showing a raised right scapula associated with a bifid rib and a synostosis between the 6th, 7th and 8th right ribs (arrow)

5. Discussion

SCS was first described in 1863 by Eulenberg, but it was not until 1891 that Sprengel gave it its name by reporting four observations [3]. SCS results from an interruption of this caudal migration whose exact cause remains unknown, possibly vascular [5,6]. A few familial forms of SCS have been described and suggest the possibility of genetic transmission [6]. In case of SCS, the scapula is in a high position and appears more or less hypoplastic and dysmorphic, wider than high, with a convex medial edge and a concave lateral edge. This is associated with hypoplasia of the scapular muscles. The scapula on the opposite side, in normal position, is sometimes slightly dysmorphic [7]. SCS is frequently associated with other abnormalities. Among the bony anomalies, let us cite costal anomalies (aplasia, hypoplasia, synostoses, bifidity, supernumerary ribs, etc.); clavicular (abnormalities of shape and/or position) and above all, vertebral (Klippel-Feil syndrome, spina bifida cervical and/or thoracic occulta, hemivertebrae, secondary scoliosis, etc.). Among extra-osseous anomalies, diastematomyelia, cervical muscle hypoplasia, torticollis, pterygium colli, cleft palate and renal abnormalities may be observed [6]. Several cases of SCS have also been described in association with other syndromes. The most frequent association, however, concerns the omovertebral bone, even if it is inconsistent. Its origin remains controversial: the omovertebral bone could develop from the posterior arc of the vertebrae, from the scapula, or correspond to acquired ossification [8]. It is usually single but can be double. Its size varies, of the order of a few centimeters. Most often, it is interposed between the spinous processes, posterior arches or transverse processes of the last cervical vertebrae and the superomedial border of the scapula. A case of omo-vertebral bone stretched between the clavicle ("omo-clavicular" bone), the occiput and the medial edge of the scapula has been reported [9]. SCS is usually unilateral (90% of cases), more common in girls than in boys (sex ratio of 1/3), as in our case, with or without side predominance according to the authors [1,6,8]. Parents consult because of aesthetic damage (scapular asymmetry, posterior thoracic swelling corresponding to the protrusion of the tip of the scapula) and/or a limitation of active abduction of the shoulder. Multidirectional shoulder instability may be associated. The Cavendish classification is clinical. The diagnosis of SCS is confirmed by radiographs which show a scapula in a high position. The radiological classification of Rigault and Pouliquen takes into account the importance of this elevation on standard images: discreet elevation (grade I, omovertebral bone generally absent); usual (degree II) or serious (degree III) [11]. It is sometimes difficult to confirm the possible presence of an omovertebral bone on an x-ray due to bony superpositions. The scanner and its 3D reconstructions are then very useful for searching for this omovertebral bone; specify the seat and size; evaluate the position of the scapula in space (lateral tilt in the frontal plane, anterior in the sagittal plane and medial in the transverse plane) and the importance of scapular dysmorphia; look for associated bony anomalies, particularly vertebral ones, and plan the surgical intervention as best as possible [4,7]. The CT scan occupies a preponderant place in the preoperative assessment but does not make it possible to satisfactorily assess a possible non-ossified component or the presence of a fibrous or cartilaginous structure. Conversely, MRI can be useful, on the one hand, to analyze this fibrous and/or cartilaginous component and, on the other hand, to search for associated spinal cord abnormalities. Ultrasound, on the other hand, is simple, easy to use, and also makes it possible to easily evaluate the ossified and non-ossified components of a superficial supernumerary structure. It could be performed in cases of SCS without detectable omovertebral bone on radiographs and preferred to MRI in young children for the reasons specified above. On ultrasound, the cartilaginous component has a characteristic appearance: rounded or oval structure, well defined, hypoechoic. The only ultrasound observation reported in the literature concerns a structure that is both fibrous and cartilaginous, the morphological appearance of which differed depending on the tissue concerned: oval formation, well limited, hypoechoic, similar to our observation (cartilage) and linear formation, well limited, hypoechoic (fibrous tissue) [1]. However, pathological correlations are lacking.

6. Conclusion

SCS is characterized, to varying degrees, by a dysmorphic scapula in a high position. An ossified supernumerary structure (omovertebral bone) or not is often associated with it and sits between the medial edge of the scapula and the cervical spine. The diagnosis is raised clinically and early due to aesthetic (asymmetry of the shoulders) and/or functional (limitation of abduction) damage. X-rays make it possible to confirm the diagnosis and to objectify a possible omo-vertebral bone but they are sometimes difficult to interpret due to bony superpositions. The complementary imaging assessment is mainly based on the scanner and its 3D reconstructions. These specify the location and dimensions of the omovertebral bone as well as the often associated vertebral anomalies. In the absence of omovertebral bone, ultrasound can be useful to look for a fibrous or cartilaginous structure. MRI can also assess the possible existence of such a structure but it is often technically difficult to achieve in a young child. Ultrasound could in the future constitute a significant adjunct in the therapeutic management of this condition.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

The present research work does not contain any studies performed on animals/humans subjects by any of the authors

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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