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Case Report

Congenital Elevation of the Scapula (Sprengel Malformation): Imaging Contribution

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Abstract: Congenital elevation of the scapula, also known as Sprengel's malformation, is a rare congenital anomaly of the shoulder girdle secondary to a defect migration of the scapula during embryonic life. The pathophysiology remains poorly understood. Diagnosis is made in early childhood. Imaging, CT in particular with multi-planar reconstruction, plays a fundamental role in the positive diagnosis and assessment of the disease. Ultrasonography and MRI are second-line examinations. Standard X-rays can be used for positive diagnosis, but the assessment of lesions is limited due to the superimposition of different anatomical planes. We report three cases of Sprengel malformation with particularities for each case.

Keywords: Congenital elevation, scapula, vertebrae, Computed tomography (CT).

INTRODUCTION

Congenital elevation of the scapula, also known as Sprengel's malformation, is a rare congenital anomaly of the shoulder girdle secondary to a defect migration of the scapula during embryonic life. The pathophysiology remains poorly understood.

Diagnosis is made in early childhood. Imaging, CT in particular with multi-planar reconstruction, plays a fundamental role in the positive diagnosis and assessment of the disease. Ultrasonography and MRI are second-line examinations.

Standard X-rays can be used for positive diagnosis, but the assessment of lesions is limited due to the superimposition of different anatomical planes. We report three cases of Sprengel malformation with particularities for each case.

Observation 1:

This is a 4-year-old male patient who presented with clinically visible positional asymmetry of the scapulae, ascension of the left scapula, stage III of the Cavendish classification, limited mobility of the left shoulder, and torticollis, in favor of a Sprengel malformation. A CT scan was performed to assess the lesion.

A CT scan of the shoulder revealed an elevation of the left scapula with a superior internal angle opposite to C6 (grade II of the Rigault and Pouliquen classification).

This is associated with a bony structure linking the left posterior hemi-arc of C5 to the medial edge of the scapula, about an omo-vertebral bone.

There are also abnormalities of the cervical vertebrae, such as a lack of fusion of the posterior arches of C5 and C6, explaining clinical torticollis.

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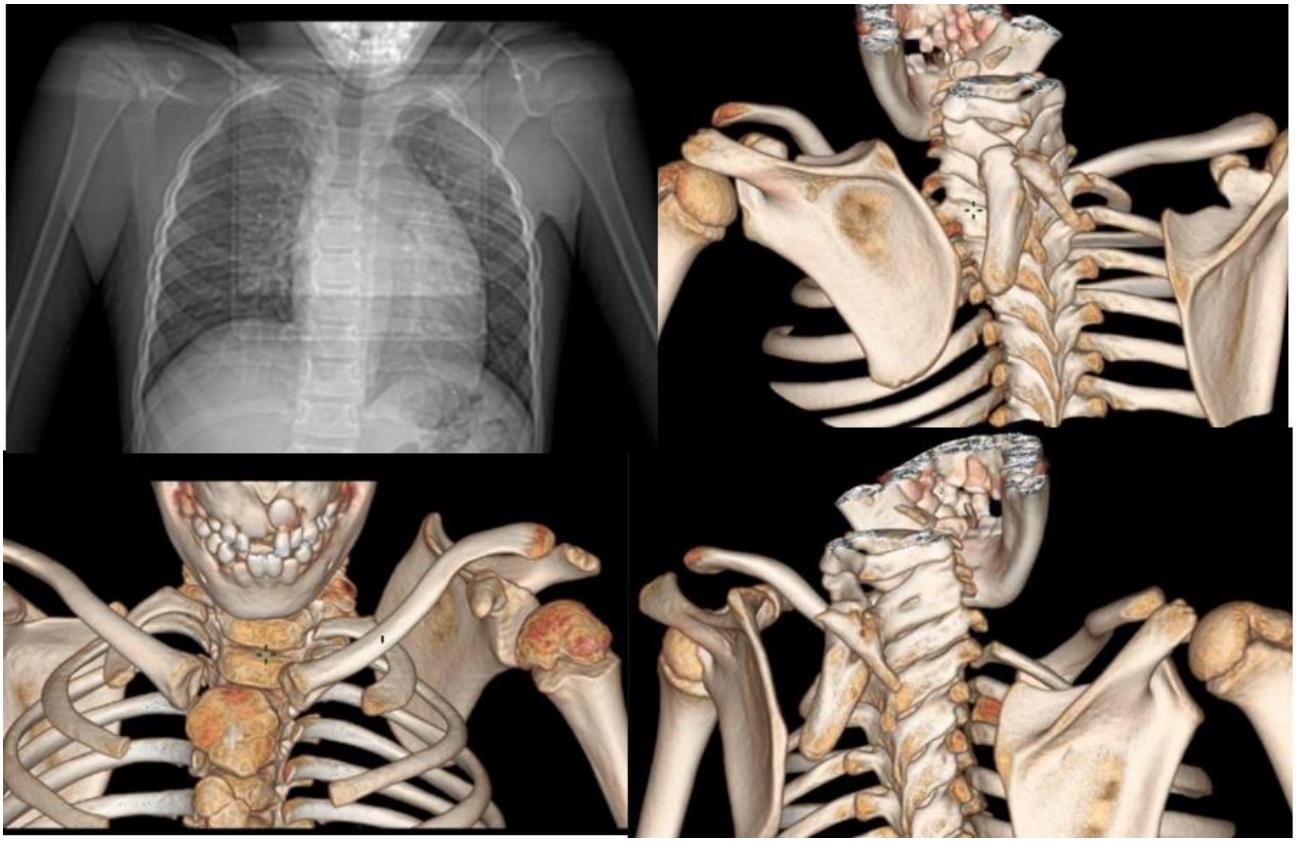

Figure 1: CT images in VR reconstruction showing elevation of the left scapula with its superior-internal border located opposite to C6 and an omo-vertebral bone connecting C5 to the medial border of the scapula

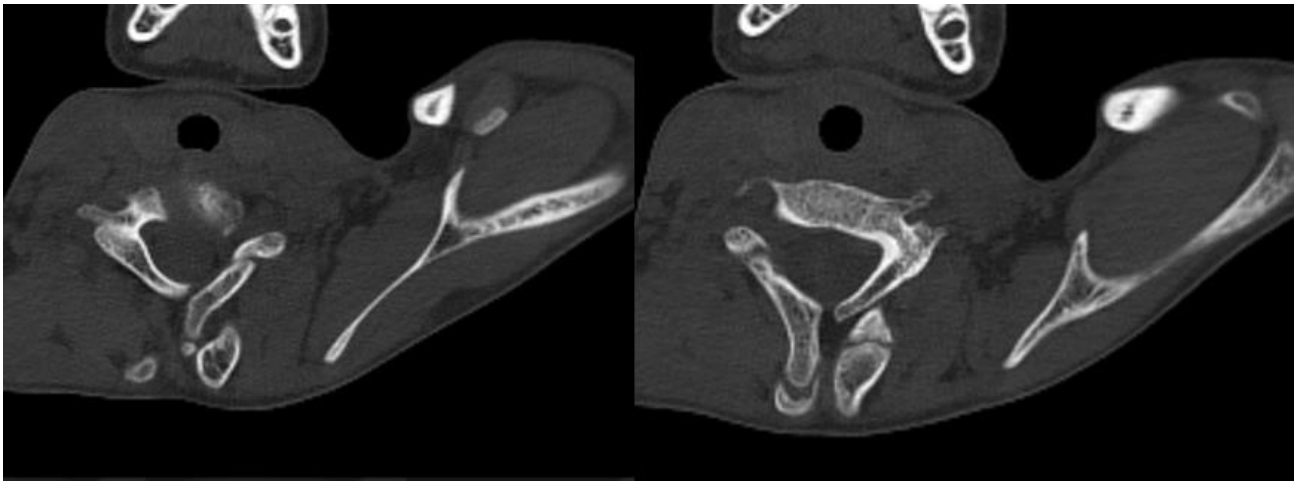

Figure 2: Axial slice CT images in the bone window showing a closure defect in the posterior arches of C5 and C6

Observation 2:

This is a 3-year-old male patient who presented with positional asymmetry of the scapulae, ascension of the left scapula classified as stage III in the Cavendish classification, with limited joint mobility of the left shoulder. A CT scan was performed to assess the lesion.

The scan revealed an elevated left scapula with a superior internal angle opposite C5 (grade II of the Rigault and Pouliquen classification) with an omo-vertebral bone.

Failure fusion of the posterior arches of C5 and C6 and hypertrophied left vertebral laminae.

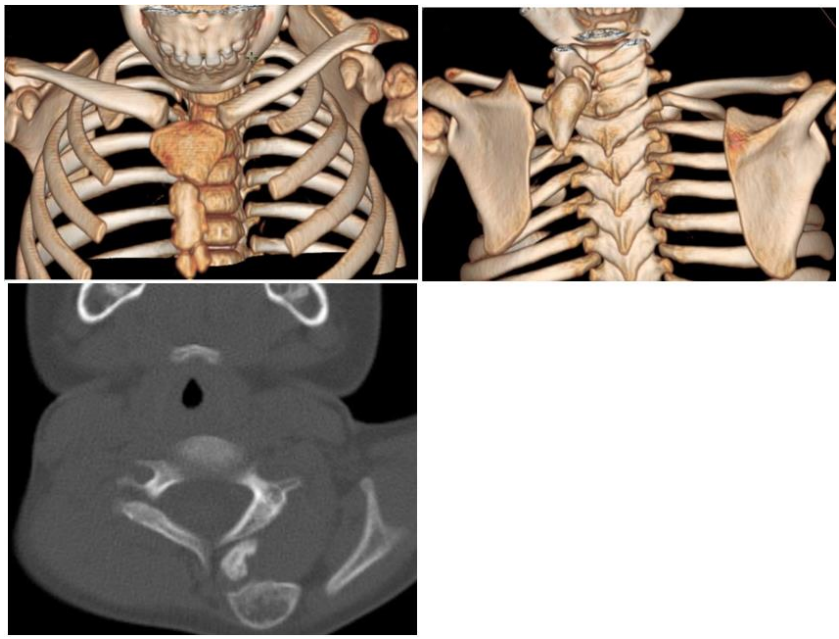

Figure 3: CT images in VR reconstruction and axial section showing a Sprengel malformation, grade II according to the Rigault and Pouliquen classification, with omo-vertebral bone and posterior dysraphism of C5 and C6

Observation 3:

This is a 14-year-old female patient who presented with positional asymmetry of the scapulae with the ascension of the left scapula classified as stage III in the Cavendish classification and a marked torticollis. A CT scan was performed to assess the lesion.

The scan revealed an ascension of the left scapula with a superior internal angle opposite to the transverse apophysis of C6 (grade II of the Rigault and Pouliquen classification). No vertebral bone was identified, and no cartilaginous or fibrous structure was identified on ultrasound.

It is associated with constitutional anomalies of the cervical vertebrae such as agenesis of the hemi-arch of C1, fusion defect of the posterior arch of C6, vertebral block from C2 to C6, hypoplasia of the lateral masses of C2.

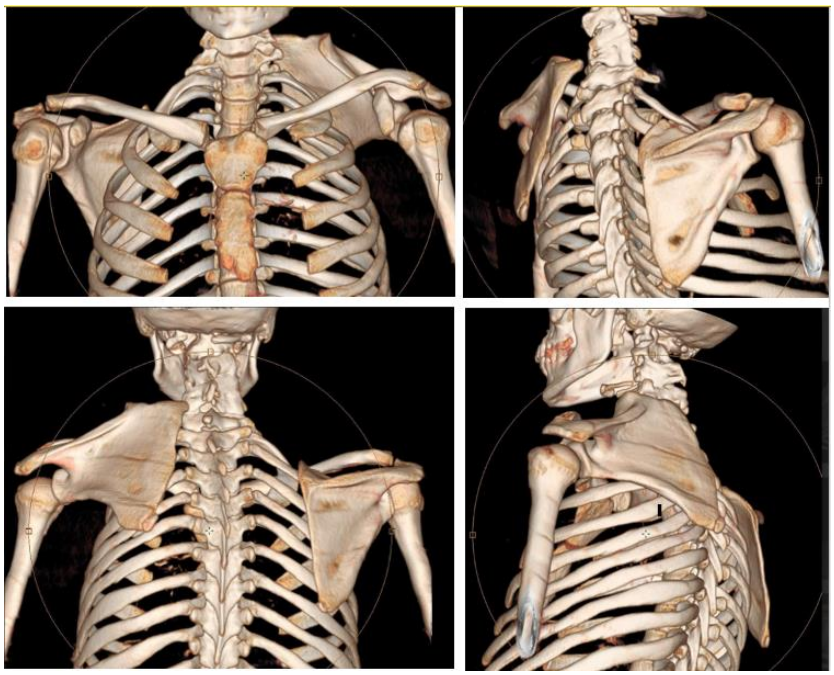

Figure 4: CT images in VR reconstruction showing a grade II Sprengel malformation according to the Rigault and Pouliquen classification, with no detectable omo-vertebral bone or fibrous or cartilaginous structure visible on ultrasound. There is an extensive vertebral block from C2 to C6, which explains the torticollis, as well as agenesis of the left hemi-arch of C1 and a dysmorphic appearance of the spinous processes of the fused cervical vertebrae

DISCUSSION

Congenital elevation of the scapula is a rare malformation of the shoulder girdle. It is secondary to a defect in the migration of the scapula during embryonic life, which may be more or less marked, and associated with a more or less severe scapular dysmorphia [1].

This migration begins at the 9th week in utero, with progressive shaping of the scapula, which is fairly close to its adult form by the 12th week in utero [1].

The pathophysiology of this anomaly remains unknown. A vascular hypothesis has been put forward [2, 3].

Familial forms have been reported, suggesting an underlying genetic abnormality [3]. This condition mainly affects women, in almost 75% of cases, and is almost always unilateral.

Bilateral elevation of the scapulae should be investigated for Klippel Feil malformation [3]. The association with other skeletal and extra-skeletal malformations should not be overlooked.

Cervical malformations such as vertebral blocks, hemi vertebrae or defects in the closure of the posterior vertebral arches are often associated, due to the common origin of the scapula and cervical spine, both of which derive embryologically from the mesoderm [3].

There is also the possibility of costal and clavicular malformations and hypoplasia of the chest wall muscles [3].

Congenital elevation of the scapula is generally diagnosed in infancy (the first two years of life), causing damage that is usually cosmetic but may also be functional due to limited abduction secondary to the presence of the omo-vertebral bone, or torticollis under tension due to associated cervical spine anomalies [3-5].

There are several grades of severity depending on the extent to which embryological migration of the scapula has been arrested and the size of the thoracic swelling.

Based on the extent of swelling and scapula elevation, the Cavendish classification distinguishes four clinical stages of increasing severity, from very mild malformation (stage I) to severe malformation (stage IV) [6].

Imaging plays a fundamental role in the positive diagnosis, the search for associated malformations, and the identification of the redundant bone that frequently connects the superior-medial angle of the scapula to the posterior arch of one of the last cervical vertebrae, most often C4 and C5.

This bone is called the omo-vertebral bone and is more or less ossified. An omo-clavicular bone has also been described [7].

Congenital elevation of the scapula is confirmed by X-rays, which show the scapula in an elevated position. The Rigault and Pouliquen classification takes into account the extent of this elevation on standard X-rays: discrete elevation (grade I, omo-vertebral bone generally absent), habitual elevation (grade II), or severe elevation (grade III) [8].

Computed tomography (CT) is the gold standard because of the volume reconstructions it provides, enabling a comprehensive assessment of bone malformations. In contrast, standard radiography may be limited by the superimposition of different planes masking the presence of any vertebral bone [1, 9].

The origin of the vertebral bone is debated; it may be formed from the posterior arch of a cervical vertebra, or the scapula, or correspond to an acquired ossification [10].

It may be reduced to a fibrous or cartilaginous structure and therefore not seen on standard radiography or CT [10].

Ultrasound and MRI will therefore play a central role in the research for this structure. MRI can look for associated spinal cord malformations such as diastematomyelia or spina bifida occulta [11].

Surgical treatment will realign the ascending scapula and is indicated for stages II and III of the Rigault and Pouliquen classification and stages III and IV of the Cavendish classification [12-14].

CONCLUSION

Congenital elevation of the scapula is a rare malformation of unknown pathogenesis, most often diagnosed in early childhood and responsible for both cosmetic and functional damage. CT scans with multi-planar reconstructions play an important role in assessing the disease. Ultrasound and MRI have their place in highlighting the cartilaginous and/or fibrous component of the omo-vertebral bone, which may also be exclusive. MRI can also be used to assess associated spinal cord anomalies. The contribution of standard radiography remains limited, especially a pre-operative disease assessment, due to the summation of the different anatomical planes.

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