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Case Report

Sprengel Deformity with Omovertebral Bone and Vertebral Anomalies: Imaging Findings in a Case Report

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Abstract

We report the case of a patient presenting with Sprengel syndrome, also known as congenital elevation of the scapula, a rare developmental anomaly resulting from defective caudal migration of the scapula during embryogenesis. It is usually diagnosed during childhood and clinically manifests as shoulder asymmetry and limitation of shoulder abduction.

This condition may occur as an isolated anomaly or in association with other congenital musculoskeletal abnormalities. One of the characteristic associated findings is the presence of an osseous, fibrous, or cartilaginous omovertebral structure between the scapula and the cervical spine, which may limit scapular mobility.

Imaging plays a central role in diagnosis and therapeutic planning. Standard radiography is often the first-line examination. Computed tomography (CT), particularly with three-dimensional reconstructions, provides excellent anatomical assessment of the scapula, the cervicothoracic junction, and possible osseous omovertebral connections. Magnetic resonance imaging (MRI) complements CT by enabling detailed evaluation of soft tissues and detection of non-ossified fibrous or cartilaginous bands, while also assessing potential associated spinal cord or muscular abnormalities.

Therapeutic management mainly depends on the patient's age, the degree of functional limitation, and the severity of the deformity according to the Rigault and Pouliquen classification.

Keywords: *Sprengel Deformity; Omovertebral Bone; Vertebral Anomalies; Magnetic Resonance Imaging (MRI); Computed Tomography (CT)*

Introduction

Sprengel deformity is a rare congenital anomaly characterized by an abnormally high scapula [1]. It is the most common malformative pathology of the shoulder in children [2].

This deformity leads to a cosmetic deformity of the shoulder and a limitation of its movement, and is often associated with vertebral or muscular anomalies [1].

It occurs as a result of the failure of the scapula to migrate caudally during embryologic development [3].

Diagnosis is primarily clinical, but imaging, particularly 3D CT scan, plays a major role in confirming, classifying, and deciding on treatment options [4].

MRI, on the other hand, is becoming the gold standard for characterizing unossified omovertebral structures and the search for associated anomalies, particularly spinal cord anomalies [4].

Case Presentation

A 3-year-old girl was referred to the pediatric orthopedic surgery department for marked shoulder asymmetry and progressive limitation of left upper limb mobility. Clinical examination revealed a clear elevation of the left shoulder, associated with restriction of left shoulder abduction to 90°, as well as a short neck (Figure 1).

Figure 1: *Asymmetry of the shoulders Cavendish grade III with a short and wide appearance of the neck.*

Initial chest radiography demonstrated elevation of the left scapula (Figure 2). This finding was further evaluated by CT with three-dimensional reconstructions, which confirmed the scapular elevation, with the superomedial angle positioned at the level of the transverse process of C4 (Figure 3 and 4).

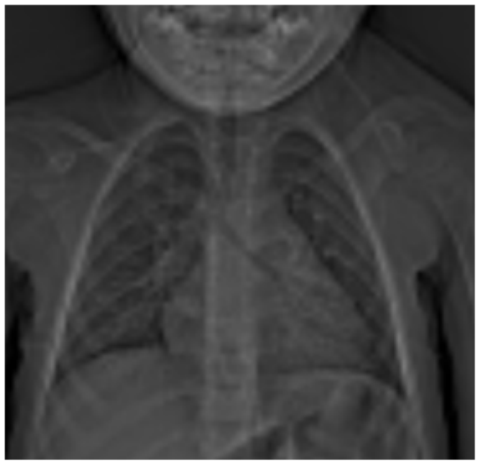

Figure 2: *Chest radiograph demonstrating elevation of the left scapula.*

CT additionally showed that the left scapula was enlarged compared with the contralateral side, with a concave lateral border and an inferiorly oriented glenoid cavity (Figure 4). An incomplete omovertebral bone was also identified, appearing as a bony outgrowth arising from the left lamina of C6 (Figure 3 and 4).

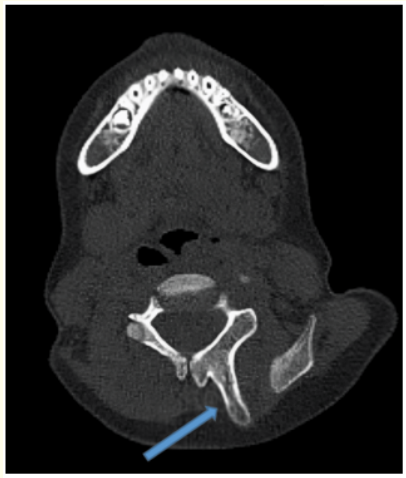

Figure 3: *Cervical CT scan, axial section in bone window.*

Presence of an incomplete omovertebral bone (Blue arrow) originating from the left lamina of C6.

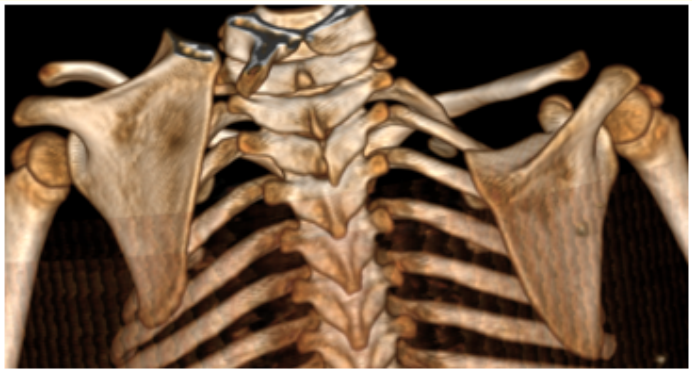

Figure 4: *3D reconstruction CT scan.*

We observe an asymmetry in the height of both scapulae, with marked elevation of the left scapula (Asterix), which appears larger than its contralateral counterpart and presents a concave lateral border (Orange arrow) with a glenoid cavity oriented downward (Yellow arrow).

We also note the presence of an incomplete omovertebral bone arising from the left lamina of C6 (Blue arrow).

Associated vertebral anomalies were present, including non-fusion of the posterior arches of C6 and T2, as well as a bifid spinous process at C6 (Figure 5). Based on these findings, the diagnosis of Sprengel syndrome associated with cervical malformations was established.

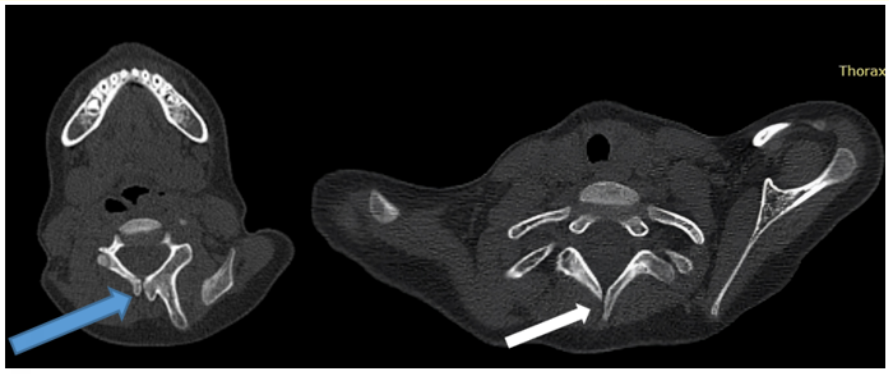

Figure 5: *Axial cervicothoracic CT scan, showing associated vertebral abnormalities.*

Congenital fusion anomalies of the cervicothoracic posterior vertebral arches were also identified, including non-fusion of the posterior arch of C6, which showed a bifid spinous process (Blue arrow), as well as non-fusion of the posterior arch of T2 (White arrow).

The case was classified as grade III according to the Rigault and Pouliquen radiological classification. Surgical management was proposed to the family at the age of 5 years; however, they declined the procedure. The patient subsequently underwent physiotherapy aimed at improving shoulder mobility.

Discussion

Sprengel's deformity was first described by Otto Sprengel in 1891 [3]. This deformity affects the musculoskeletal development of the shoulder, and has both functional impairment and cosmetic issues [5]. It is rarely bilateral, has no clear propensity for either side, and is more prevalent in females [6].

The scapula begins to form at five weeks' gestation as a mesenchymal mass at the level of C4-C5 vertebrae. It begins to migrate caudally at six weeks, forming normal shape and position by twelve weeks [6].

The interruption of this migration is what results in Sprengel's deformity, although etiology is currently unknown [2].

The abnormal scapular migration is often aggravated by the presence of an omovertebral connection, which may be osseous, fibrous, or cartilaginous. This connection maintains an abnormal fixation between the scapula and the cervical spine, resulting in medial rotation of the scapula with inferior orientation of the glenoid cavity and consequent limitation of arm abduction [4].

Three main hypotheses have been proposed to explain the development of an omovertebral bone. One suggests a vertebral origin, in which the lesion arises from the spinous process or from excessive development of the posterior lateral processes, referred to as hypapophysis, which may later fuse with the scapula, as observed in our case [5]. Another proposes a scapular origin, in which the bone develops from the posterior border of the scapula and subsequently establishes a connection with the vertebral column through an epiphyseal outgrowth [5]. A third theory describes an independent origin, in which the bone results from ossification of intermuscular connective tissue between the scapula and the vertebrae, or from a rudimentary vestigial scapular element related to an early embryonic cleft [5].

The omovertebral bone is typically unique and connects the spinous processes, posterior arches, or transverse processes of the lower cervical vertebrae to the superomedial border of the scapula. However, rare cases reported in the literature have described atypical connections between the scapula and the clavicle or the occiput [4].

The omovertebral bone can also be incomplete and fail to fully connect the scapula to the cervical vertebrae, as we have documented in our case. It can stop short of making either connection, manifest as a discontinuous rudimentary fragment, or be incomplete with fibrous or cartilaginous components [1,5].

Sprengel's deformity may occur as an isolated anomaly or in association with other skeletal or extraskeletal abnormalities. Among the most frequently reported associations are cervical vertebral anomalies, particularly Klippel-Feil syndrome, which corresponds to cervical vertebral fusion and is observed in approximately 20 - 24% of bilateral cases [2]. Additional associated skeletal anomalies include clavicular abnormalities, scoliosis, spina bifida as observed in our case, and rib anomalies such as supernumerary ribs, hypoplasia, or bifidity. Extraskeletal associations have also been described, including diastematomyelia, hypoplasia of cervical musculature, congenital muscular torticollis, cleft palate, and renal anomalies [3,6].

Anteroposterior radiography including both shoulders and the cervicothoracic spine on the same image constitutes the first-line examination. It allows confirmation of the abnormally elevated position of the scapula but remains limited because of overlapping bony structures, particularly for assessing the severity of the deformity and identifying associated anomalies [4,6].

Computed tomography (CT) plays a major role in the preoperative evaluation of Sprengel deformity and allows for a more precise anatomical assessment than conventional radiography, particularly through two-dimensional and three-dimensional reconstruction [1,4]. These reconstructions allow for an accurate assessment of the degree of scapular elevation, as well as a detailed evaluation of scapular morphology, including its shape, size, spatial orientation, contours, and glenoid cavity orientation. Although the scapula may appear hypoplastic, it is in fact often larger than the contralateral side, dysmorphic, and characterized by a configuration that is wider than it is tall, with a convex medial border and a concave lateral border [1,6].

CT is also highly valuable for identifying an omovertebral connection and characterizing its origin, extent, and nature, whether osseous, cartilaginous, or fibrous. In addition, it facilitates the detection of associated skeletal abnormalities, especially vertebral malformations [1,4].

In our case, 3D reconstructions showed a scapula larger than the contralateral side and higher than wide, with a concave lateral border and a downward-oriented glenoid cavity. It also allowed us to demonstrate the presence of an incomplete omovertebral bone arising from the left lamina of C6 (Figure 4).

When the omovertebral connection is fibrous or cartilaginous, MRI is the reference imaging modality [6]. For optimal visualization, sedation and dedicated patient positioning are recommended, including lateral flexion of the neck toward the contralateral side and elevation of the affected shoulder [4]. MRI also allows evaluation of associated spinal cord or vertebral anomalies [4,5].

In the prenatal setting, ultrasound allows diagnosis as early as the second trimester of pregnancy through the identification of an elevated scapula, asymmetry of the nuchal soft tissues, and, in some cases, the presence of an omovertebral bone confirming the anomaly. Early detection facilitates the investigation of associated malformations and contributes to genetic counselling [3].

In the postnatal period, ultrasound represents a valuable complement to radiographic and CT examinations, particularly for the assessment of cartilaginous and fibrous structures that are not visible on plain radiographs [4]. It is a non-invasive complementary tool useful for initial screening and for the dynamic evaluation of scapular mobility as well as associated muscular abnormalities, such as hypoplasia of the scapular muscles [4].

Clinical assessment is primarily based on the Cavendish classification, which evaluates the cosmetic severity of the deformity into four grades [6]. Grade I corresponds to a mild deformity that is not visible under clothing, grade II is characterized by level shoulders with a visible prominence at the base of the neck, grade III corresponds to a shoulder elevation of 2 to 5 cm, and grade IV to an elevation greater than 5 cm [6].

Radiological assessment is based on the Rigault and Pouliquen classification, which relies on the position of the superomedial angle of the scapula relative to the cervical transverse processes on standard radiographs or computed tomography [6]. Grade I corresponds to a position between T2 and T4, grade II between C5 and T2, and grade III above C5 [6].

Our case was classified as Cavendish grade III and Rigault-Pouliquen grade III.

Surgical treatment is typically performed between the ages of 3 and 7 for cosmetic and functional reasons [4,5]. It is generally indicated in Rigault and Pouliquen types II and III, as well as in Cavendish types III and IV [6]. In contrast, surgery is not recommended in Cavendish grade I cases, in which the cosmetic and functional impact is minimal or absent [6].

When indicated, several surgical techniques may be used, including the Woodward procedure, which consists of releasing the parascapular muscles and reattaching them at a lower level on the scapula [3], and Green's technique, which involves resection of part of the superomedial scapula [3].

Removal of the omovertebral bone, or fibrous/cartilaginous equivalent, is always performed to allow scapular mobilization and rotation correction [4].

For primarily cosmetic cases, resection of the superomedial scapular angle can also be performed [6].

The major neurological complication associated with surgical correction of Sprengel deformity is injury to the brachial plexus [5]. This complication usually results from stretching or compression within the thoracic outlet during scapular lowering toward a more anatomical position [5].

Other specific nerves may also be affected depending on the surgical procedure performed. The spinal accessory nerve may be injured during resection of the omovertebral bone, whereas the suprascapular nerves are at risk during resection of the superomedial angle of the scapula [6].

Several factors influence the risk of neurological complications, including the patient's age, the degree of scapular lowering, and the surgical technique used [2]. The risk increases significantly in children older than 6 to 8 years [6]. In addition, because the scapula is often hypoplastic, attempting to achieve perfect symmetry with the contralateral side may result in excessive traction on the brachial plexus [6].

To minimize these risks, several preventive measures have been recommended, including clavicular osteotomy to reduce tension on neurovascular structures, intraoperative neurophysiological monitoring using somatosensory evoked potentials or electromyography, and clinical monitoring of the radial pulse during scapular lowering maneuvers [6].

For our patient, surgical treatment was proposed; however, it could not be performed because the family declined the procedure.

Conclusion

Sprengel's deformity is an uncommon but important congenital shoulder girdle deformity.

Imaging, especially CT, is essential for full characterization of the deformity and associated anomalies, particularly the presence of an omovertebral bone.

Early diagnosis and appropriate surgical intervention can improve cosmetic appearance and function, highlighting the importance of a multidisciplinary approach.

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