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SPRENGEL'S DEFORMITY WITH ASSOCIATED CERVICAL DYSRAPHISM AND OMOVERTEBRAL BONE: A PEDIATRIC CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Sprengel's deformity is a rare congenital anomaly characterized by the failure of the scapula to descend during normal embryonic development, resulting in a high-riding, hypoplastic, and malrotated shoulder blade [1]. This condition frequently manifests with cosmetic asymmetry and functional limitations, notably restricted shoulder abduction [1]. It is also commonly associated with musculoskeletal malformations, including omovertebral connections, rib fusions, and cervical spine defects such as spina bifida occulta and Klippel-Feil syndrome [1]. This article reviews the clinical presentation, diagnostic imaging modalities (incorporating standard radiography and three-dimensional computed tomography), and management considerations for pediatric Sprengel's deformity [2.3].

KEYWORDS

Sprengel , Omovertebral bone , CT, 3D

MAIN ARTICLE

INTRODUCTION

Sprengel's deformity, or congenital high scapula, is the most common congenital defect of the shoulder girdle [4]. Normally, the scapula originates opposite the cervical spine during early embryonic life and descends to its definitive thoracic position by the third month of gestation. Arrest of this caudal migration leads to a persistently elevated and medially rotated scapula [1]. First comprehensively described by Otto Sprengel in 1891, the condition can occur sporadically or via familial inheritance [1, 4]. Beyond cosmetic deformities, patients often experience functional deficits, muscular hypoplasia, and secondary spinal imbalances [1].

CLINICAL PRESENTATION

A 4-year-old child was presented with a congenital elevation of the scapula and a clinical suspicion of an omovertebral bone. Cervico-thoracic computed tomography (CT) acquisition without intravenous contrast, utilizing multiplanar reconstructions and parenchymal and mediastinal window readings, was performed for precise anatomical evaluation. The scan revealed a markedly elevated left scapula with the individualization of a distinct omovertebral bone extending from the superomedial border of the scapula to the laminae and left hemi-spinous processes of C5 and C6 (figure 1). Additionally, imaging demonstrated a midline clefting defect at the spinous processes of C5, C6, and C7 consistent with a spina bifida occulta (figure 2), confirming a complex Sprengel's deformity associated with underlying posterior arch cervical dysraphism.

DISCUSSION

Sprengel's deformity represents a complex developmental arrest of the shoulder girdle, posing significant clinical challenges regarding both functional restoration and cosmetic outcomes [2]. The pathophysiology is rooted in the disruption of normal embryonic caudad migration between the ninth and twelfth weeks of gestation [1]. Clinically, the severity of the condition is frequently classified using the Cavendish grading system, which categorizes

deformities from Grade 1 (very mild) to Grade 4 (severe, with the superior angle near the occiput) [2].

A hallmark complication contributing to restricted kinematics is the presence of an omovertebral bone or fibrous band, reported in approximately 25% to 50% of cases [1]. This anomalous structure—such as the cervicothoracically bridged bone identified in our patient linking C5-C6 laminae to the scapular border—anchors the scapula rigidly to the cervical spine, neutralizing the natural gliding plane of the scapulothoracic articulation. Furthermore, Sprengel's deformity is frequently intertwined with systemic or regional musculoskeletal anomalies, most notably Klippel-Feil syndrome, cervical spina bifida occulta, rib hypoplasia, and secondary progressive thoracic scoliosis [1, 2, 3].

Diagnostic protocols have evolved significantly with cross-sectional imaging [2]. While standard anteroposterior radiographs remain mandatory to establish baseline metrics, advanced imaging modalities—specifically multidetector computed tomography (MDCT) with multiplanar reconstructions and three-dimensional (3D) volume rendering—are indispensable for surgical planning, providing high-resolution mapping of anomalous bone structures like the omovertebral connection, evaluating periscapular muscles, and screening for intraspinal anomalies [2, 3].

Therapeutic management is dictated by the functional deficit and aesthetic impact [2, 4]. Mild cases (Cavendish Grades 1 and 2) are managed conservatively with physical therapy, whereas moderate-to-severe cases (Cavendish Grades 3 and 4) warrant surgical intervention—such as the Woodward or Green procedures—typically performed between the ages of 3 and 8 years to restore alignment and function while avoiding neurovascular injury [2, 4].

CONCLUSION

Sprengel’s deformity remains a rare and intricate congenital anomaly requiring a careful multidisciplinary approach to diagnosis and treatment [2]. Comprehensive radiological assessment using modern cross-sectional CT imaging is essential for identifying associated structural malformations like omovertebral bones, spinal clefts, and cervical fusions [2, 3]. Early identification and strategic surgical intervention can successfully mitigate functional deficits and improve the quality of life for pediatric patients [2].

FIGURES:

Figure 1 : Three-dimensional (3D) CT skeletal reconstruction (posterior view). The image demonstrates a markedly elevated and hypoplastic left scapula compared to the contralateral side. A prominent osseous bridge (omovertebral bone, yellow arrow) extends from the superomedial border of the left scapula to the posterior elements of the mid-cervical spine (C5–C6 laminae)

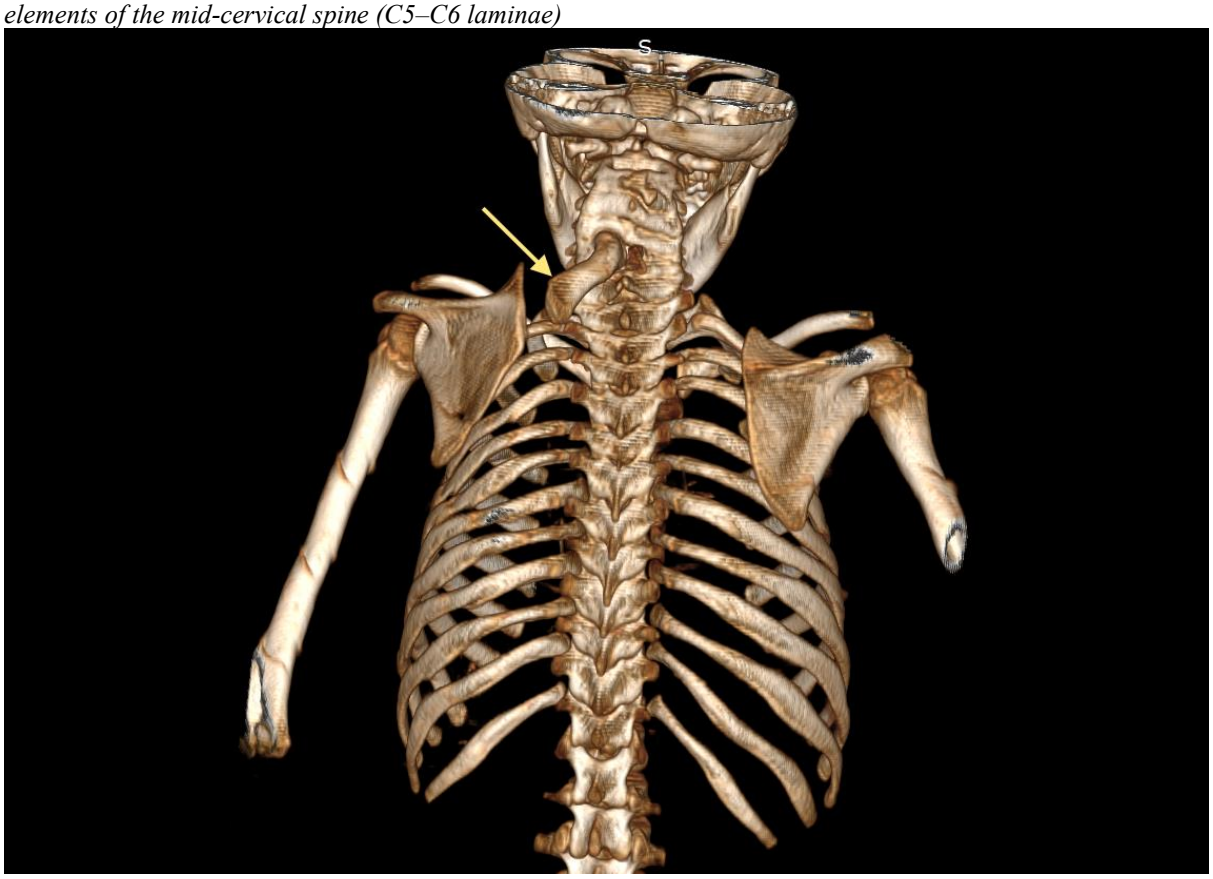
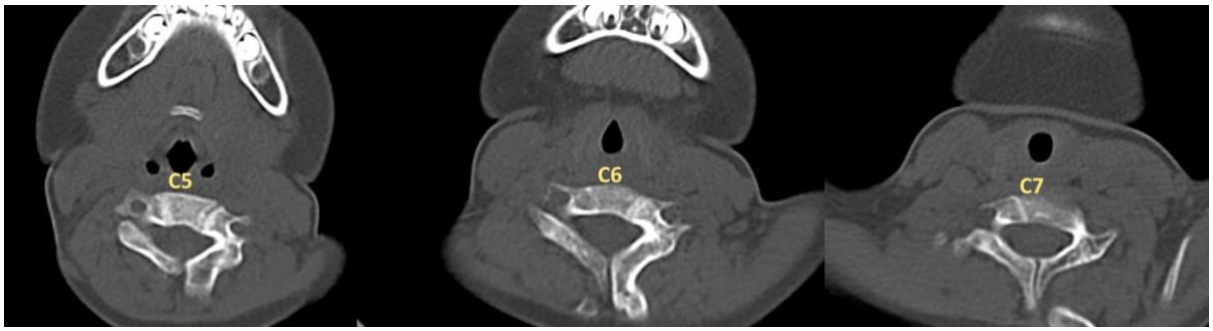


Figure 2 : Axial non-contrast CT of the cervical spine (C5, C6 and C7 levels), demonstrating o midline posterior arch defect (arrow) consistent with spina bifida occulta.



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