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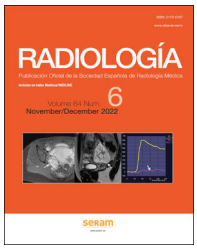

SCIENTIFIC LETTER

Sprengel's deformity: Two illustrative cases in paediatric patients

Deformidad de Sprengel: dos casos ilustrativos en pacientes pediátricos

Introduction

Sprengel's deformity, also known as congenital elevation of the scapula, is a rare congenital deformity that affects one or both scapulae and is usually noticed at birth. It is caused by the failure of the scapula to migrate during the early embryonic stage.[1]

This deformity is more than just an aesthetic issue; it is also associated with restricted mobility in the shoulder and the cervical spine. It can appear in isolation or associated with other abnormalities, the most frequent of which are Klippel-Feil syndrome, scoliosis and rib defects.[1]

Computed tomography (CT) with three-dimensional reconstructions (3D-VRT) and magnetic resonance imaging (MRI) are today fundamental in the assessment of the deformity and any other co-existing abnormalities, as well as in the planning of treatment.[2]

Case presentation

Case 1

Six-year-old girl undergoing monitoring from the age of 10 months due to delayed psychomotor development, progressive increase in head circumference (> p99) and cyst-like malformation of the posterior fossa. She was taken to the child trauma centre due to clavicle length asymmetry, winged scapula and possible Sprengel's deformity. Additional tests were requested due to the patient's progressive inability to reach the higher grades of shoulder abduction, as well as cervical pain. The patient is currently being managed conservatively, with periodic 6–12 month check-ups.

In the CT a curved bar of bone is observed, suggestive of the omovertebral bone, fused to the C7 left lamina. This bone points towards the medial aspect of the scapula body, with no sign of osseous bridges between the two (Fig. 1A–C). The left scapula is more elevated than the right, and its superomedial border is located at the C5 spinous processes (Fig. 1D).

Case 2

Five-year-old boy with no relevant personal or family history, assessed at 7 months because of difficulties in raising his left arm. Physical examination revealed an elevated left scapula, with the patient unable to complete abduction or extension of the shoulder, which suggests Sprengel's deformity. CT with 3D reconstructions were requested to assess the presence of the omovertebral bone and to plan surgery. Finally the patient was operated on using the Woodward technique with no complications.

In the CT, elevation of the left scapula may be observed, attributable to Sprengel's deformity. (Fig. 2C). Furthermore, an omovertebral bone is observed that articulates at the top with the C5 spinous process/left lamina (Fig. 2A) and at the bottom with the medial border of the scapula at D2, with no fusion of the bones (Fig. 2B). The superomedial border of the left scapula is located at the level of the C4 spinous processes (Fig. 2D).

Discussion

Both cases stand out because they demonstrate bone abnormalities typical of Sprengel's deformity, such as the elevated scapula and the omovertebral bone. They therefore provide valuable training material, particularly for radiologists who are not familiar with this entity.

Sprengel's deformity, also known as congenital elevation of the scapula, is a deformity caused by the failure of the scapula to descend as it normally would during embryonic development. Although the pathophysiology is unknown, the hypothesis is that the scapula fails to migrate between the ninth and twelfth week of development.[3]

It occurs unilaterally in up to 90% of cases. It is more common in girls (with a proportion of 1:3) and is diagnosed in early infancy/childhood. In up to 30% of cases there is an omovertebral bone, comprising bone, cartilage or fibrous tissue, between the superior angle of the scapula and the spinous process of the cervical vertebrae.[1]

As we see, the CT, particularly using the 3D-VRT reconstructions, is particularly useful in patient manage-

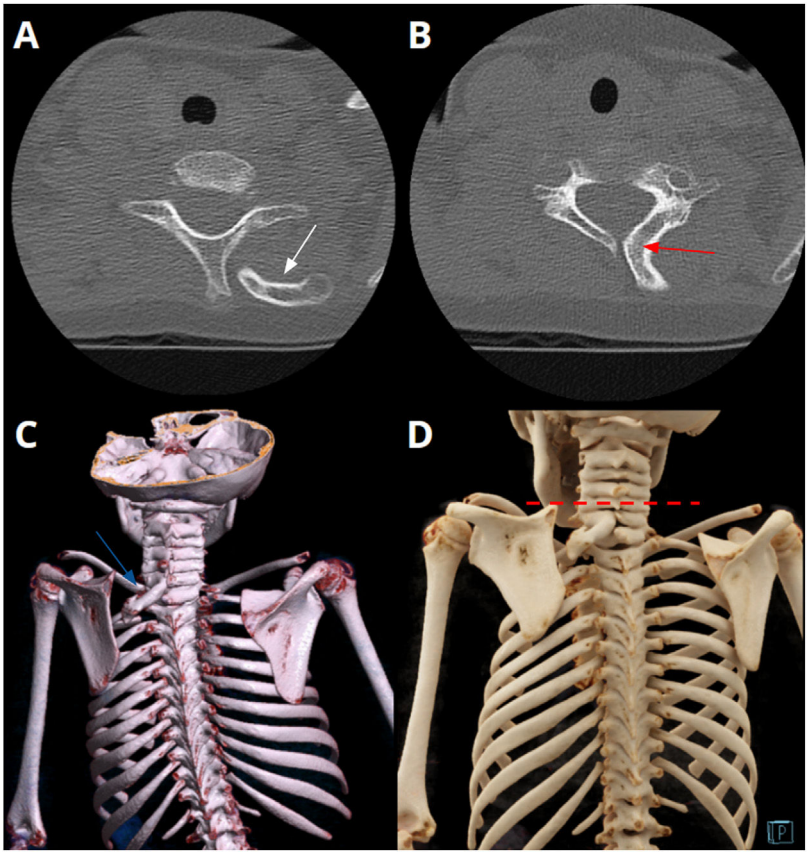

Figure 1 A and B) Simple CT of the cervical spine, axial plane in bone window, with curved bone structure (white arrow) fused to the vertebral lamina of C7 (red arrow), suggestive of an omovertebral bone. C and D) 3D-VRT reconstructions in which the morphology and direction of the omovertebral bone can be observed in more detail (blue arrow) and the elevation and well-defined rotation of the left scapula, whose superomedial border lies above the C5 spinous process (dotted red line) (Rigault classification Grade III).

ment because it allows orthopaedic surgeons to locate and analyse the deformity accurately. The omovertebral bone may be connected with fibrous, bony and/or cartilaginous tissue. It is also interesting, therefore, to perform an MRI to assess any potential fibrous/cartilaginous element.[4]

The differential diagnosis should include rickets, osteomalacia, deformity secondary to long thoracic nerve paralysis, and scapular fracture.[3]

The Rigault radiological classification divides patients into three grades[5]:

1 Grade 1: Superomedial angle lower than T2 but above the T4 transverse process.
2 Grade 2: Superomedial angle located between C5 and T2 transverse process.
3 Grade 3: Superomedial angle located above C5 transverse process.

Non-surgical treatment is reserved for children with a mild deformity, minimum shoulder dysfunction and minimal aesthetic deformity. Surgery forms the basis for treatment of all but minor cases, and is performed to improve the aesthetics and function of the shoulder. Surgical techniques (Woodward and modified Green procedures) include the resection of the elevated portion of the scapula, the removal of the omovertebral bone and the moving of the scapula to a more caudal position.[4]

Conclusion

Radiology plays a fundamental role in identifying and monitoring congenital abnormalities, and Sprengel's deformity is no exception. This rare condition has significant implications for the anatomy and alignment of the spinal column in paediatric patients. This article focuses on two clinical cases that illustrate the importance of radiology in the assessment and early diagnosis of Sprengel's deformity, highlighting that imaging tests play a crucial role in the multidisciplinary treatment of this type of patients.

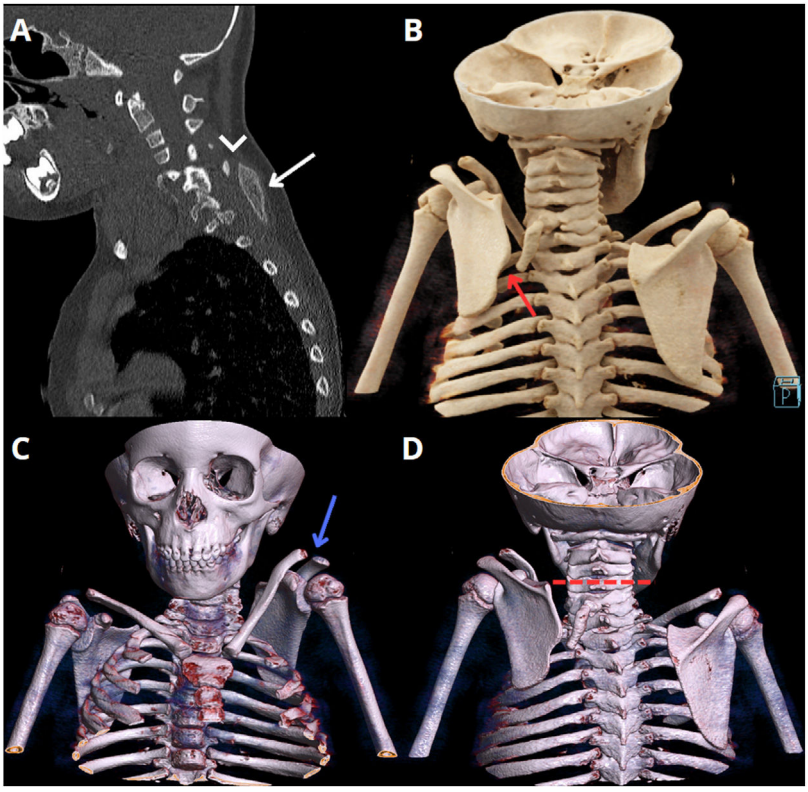

Figure 2 A) Simple cervicodorsal spine CT, sagittal plane in bone window, with omovertebral bone (white arrow) that at its superior border is joined to the C5 transverse process/lamina (white arrowhead). B) 3D-VRT reconstruction which shows the medial border of the scapula and lowest edge of the omovertebral bone, and the absence of osseous bridges between the two (red arrow). C) 3D-VRT reconstruction in anterior plane, with a marked elevation of the left scapula and clavicle (blue arrow), attributable to Sprengel's deformity. D) 3D-VRT in posterior plane, which shows that the superomedial border of the scapula is at the level of C4 (dotted red line) (Rigault's classification Grade III).

CRediT authorship contribution statement

The authors state that they contributed equally to the different sections of this study.

Declaration of competing interest

The authors declare that they have no conflicts of interest.

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