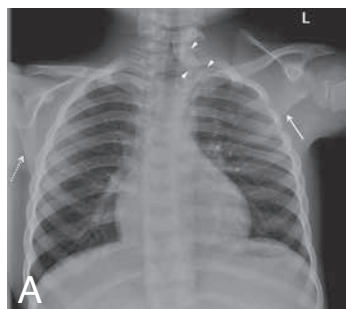


IMAGES IN CLINICAL RADIOLOGY



Sprengel's shoulder

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An otherwise healthy 5-year-old boy was referred for shoulder asymmetry and limited range of motion on the left. According to the boy's mother, he hardly ever complained about it, and was a good swimmer. Physical examination revealed left scapular elevation; glenohumeral function was good, apart from forward elevation (150 degrees). Frontal chest radiograph (Fig. A) and left shoulder plain film (Fig. B) showed elevation of the left scapula with medial rotation of its inferior angle, pointing towards the spine (arrow). This was connected to the posterior elements of the sixth cervical vertebra by a bony bar (arrowheads), which was better appreciated on a lateral radiograph of the cervical spine (Fig. C). The right shoulder girdle was normal (Fig. A, dotted arrow). No other abnormality was seen.

Comment

Sprengel's deformity, named after one of the doctors who drew attention to the condition in 1891, is a rare skeletal abnormality. It is, nevertheless, the most common congenital malformation of the shoulder girdle, resulting from failure of the scapula to descend from its position in the neck to its normal position in the posterior thorax during embryological development.

This complex anomaly consists of scapular malposition and dysplasia: the affected scapula appears not only elevated with its inferior part medially rotated, but is also smaller and more cephalad than the normal contralateral one. Regional muscle hypoplasia or atrophy is usually present, which adds to the disfigurement and limitation of shoulder movement.

The deformity can be unilateral, with the left side being more commonly affected than the right, or bilateral, which is cosmetically more acceptable but functionally more disabling. It may occur in isolation or associated with other anomalies, including Klippel-Feil syndrome (the most frequent one, characterized by the congenital fusion of any two of the seven cervical vertebrae), spinal dysraphism, vertebral and rib segmentation or fusion abnormalities.

An omovertebral bone – a rhomboid- or trapezoid-shaped fibrous/cartilaginous or osseous bar connecting the superomedial border of the affected scapula to the spinous processes, lamina or transverse processes of the cervical spine (most commonly the 4th to 7th vertebrae) – is present in 30% of patients, which is best visualized on a lateral or oblique radiograph of the cervical spine.

Cavendish classified the deformity into four grades, based on its severity: grade 1 – very mild deformity, with the shoulders almost at the same level, not noticed with the clothes on; grade 2 – mild deformity, but the superomedial portion of the high scapula is visible as a lump; grade 3 – moderate and visible deformity, with the affected shoulder 2-5 cm higher than the opposite shoulder; grade 4 – severe deformity, the scapula is very high, with the superomedial angle at the occiput, with neck webbing and brevicollis.

Surgical correction of the deformity should be considered in patients with moderate or severe cosmetic and functional compromise, and it should be performed between 3 and 8 years of age. Techniques described in literature involve a combination of scapular lowering with shift of the origin or the insertion of the scapular muscles on the spine/scapula, resection of the superomedial border, and omovertebral bar resection. Although multiple procedures have been described, the Green and the Woodward procedures remain the gold standards for correction of the condition, and their modified forms (with strategies to reduce complications, including morcelisation of the middle third of the clavicle) are most popular presently. The Green procedure detaches the muscles from the scapula whereas the Woodward procedure detaches the origins of the trapezius and rhomboids

from the spinous processes. Postoperative complications include winging of the scapula, prominent scars, and neurovascular injury (which is minimized by clavicle osteotomy).

In this case, the Sprengel's deformity was moderate (grade 3 on the Cavendish grading system). Nevertheless, the glenohumeral function was relatively preserved, with restriction of motion only on forward elevation of the arm, and apparently causing no significant impact in the boy's routine. The mother's main worry was cosmesis/functional compromise in the future, but she was reluctant to proceed directly to surgery. Therefore, a conservative approach was adopted for the time being, and contact between the mother and people who had/had not surgery was arranged.

Reference

1. Kadavkolan A.S., Bhatia D.N., Dasgupta B., Bhosale P.B.: Sprengel's deformity of the shoulder: Current perspectives in management. *Int J Shoulder Surg*, 2011, 5: 1-8.

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