International Journal of
Clinical & Medical Images

ISSN : 2376-0249
Vol 5 • Iss 1• 1000593 Jan, 2018
DOI: 10.4172/2376-0249.1000593

Clinical Image

Sprengel Deformity and Omovertebral Bone

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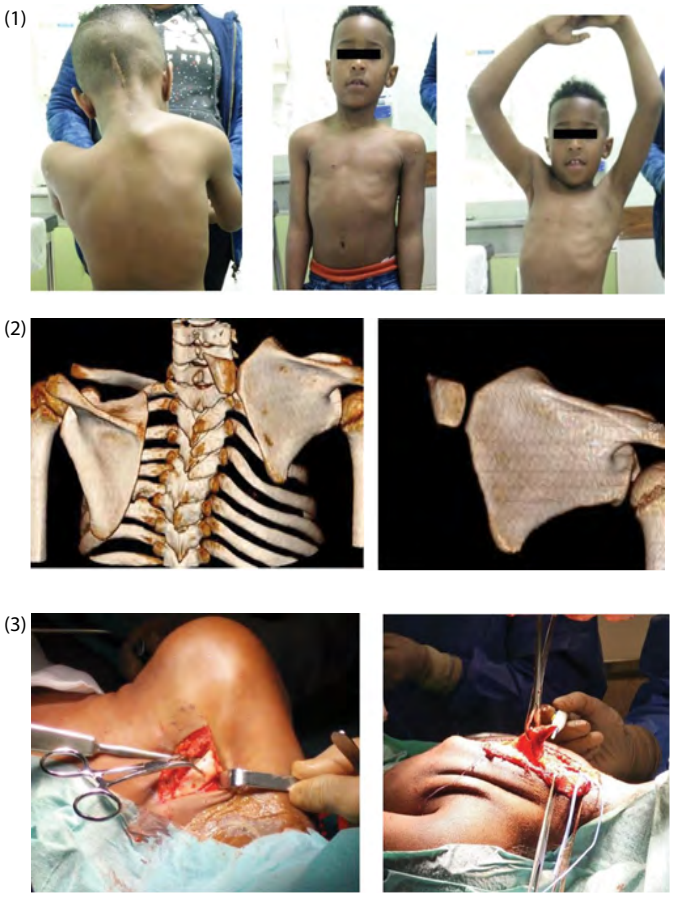

Figure 1: Sprengel's deformity is shown.
Figure 2: Scapular elevation was confirmed and an anomalous omovertebral bone was observed.
Figure 3: Clavicular osteotomy, omovertebral bone resection and woodward procedure were performed.

Abstract

Sprengel deformity is the most common congenital abnormality of the shoulder girdle, resulting from an elevated, dysplastic, and variably fixed scapula. It has cosmetic implications that make it readily identifiable, and may be also associated with functional impairment. It may associate with other structural abnormalities, such as the presence of an omovertebral bone, or bar. When present, it adds to the immobility of the scapula. We present the images relating to the case of a 8-year old boy, with previous history of spina bifida and hydrocephalus. He presented with scapular elevation and limitation of shoulder motion, and was diagnosed with a Cavendish 4 type Sprengel deformity of the scapula. An associated omovertebral bone connecting the superomedial angle of the scapula and cervical spine was detected on CT scan. 3D reconstruction of the images allowed accurate definition of the deformity and the relations of the abnormal bone. The deformity was afterwards surgically treated, by means of a clavicle osteotomy, omovertebral bone resection, and Woodward procedure. The patient had an eneventful recovery, and had satisfactory functional and cosmetic gains.

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***Citation:** Pato M, Jordão P (2018) Sprengel Deformity and Omovertebral Bone. Int J Clin Med Imaging 4: 593. doi:10.4172/2376-0249.1000593*

Keywords: Sprengel deformity; Omovertebral bone; Shoulder girdle; Paediatric orthopaedics

Case Presentation

An 8-year old boy, previously surgically treated for hydrocephalus and spina bifida, was referred to our Paediatric Hospital for evaluation of the fixed elevation and limited motion of the right shoulder (Figure 1). A clinical diagnosis of Sprengel Deformity was made, being a Cavendish 4 grade [1] in severity. In the investigations ordered scapular elevation was confirmed and the presence of an anomalous omovertebral bone was noted between the superomedial angle of the scapula and C6 spinous process (Figure 2).

Surgery was proposed, and a clavicular osteotomy, omovertebral bone resection and Woodward procedure were performed (Figure 3). Sprengel Deformity is a complex congenital pectoral girdle deformity, with cosmetic and functional implications. It is associated with other deformities, such as spinal dysraphism (as spina bifida in this case), scoliosis, and rib and vertebral segmentation abnormalities, such as Klippel-Feil. [2]

The omovertebral bone is an anomalous bone connecting the elevated scapula to the cervical spine, being called omovertebral bar should it be fused between them. It contributes to the immobility of the scapula and the functional impairment, and its resection should be performed if surgical treatment is pursued [1]. It may not be always visible on plain x-rays, and so more advanced imaging is recommended prior to surgery for assessment and planning, such as CT (ideally with 3D reconstruction) [3].

Clavicle osteotomy is advised either in the treatment of associated clavicle deformity, or as to lower the chances of brachial plexus injury [4] in higher grade corrections.The child in the case described had an uneventful postoperative course, and received rehabilitation treatment, having later satisfactory functional and cosmetic gains.

Conclusion

Sprengel deformity is a complex shoulder girdle abnormality. If surgical treatment is considered, associated abnormalities such as the omovertebral bone should be sought - CT scan with 3D reconstruction is advised. When found, omovertebral bone should be excised during surgery.

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