IPSILATERAL OMOVERTEBRAL BONES IN THE LEVATOR SCAPULAE MUSCLE AND THE RHOMBOID MUSCLE IN A SPRENGEL DEFORMITY : CASE REPORT

E. BAULOT *, P. TROUILLOUD **, E. A. GIROUX **, P. M. GRAMMONT *

Our interest was stimulated by the uncommon case of a 4-year-old girl who presented a Sprengel deformity associated with two omovertebral bones on the same side. The first omovertebral bone was situated in the levator scapulae muscle and the second omovertebral bone was lying in the rhomboid muscle. The removal of these two bones was combined with a Woodward procedure to obtain a good correction.

Keywords : omovertebral bone ; levator scapulae muscle ; rhomboid muscle ; Sprengel deformity.
Mots-clés : os omo-vertébral ; muscle levator scapulae ; muscle rhomboïde ; déformation de Sprengel.

INTRODUCTION

Although the omovertebral bone in Sprengel's deformity is well known, we know of no reports in the literature of an omovertebral bone lying in the levator scapulae muscle associated with another omovertebral bone on the same side. Gallien (8) already reported the sole observation of a accessory bone at the insertion of the levator scapulae muscle without any other omovertebral bone.

It may be significant in our patient that the two abnormal bones were associated with a very severe deformity, grade 4 on the Cavendish score (3).

CASE REPORT

A 4-year-old girl with a bilateral Sprengel deformity was seen in our institution. The cosmetic severity of the deformity was major on the left side (grade 4 on the Cavendish score (Table I), and was a grade 2 on the right side. Functional impairment consisted in severe limitation of abduction and inability to rotate or to bend the head to the left side.

There was no family history of musculoskeletal abnormality.

On physical examination, an omovertebral bone was felt extending from the superomedial border of the left scapula to the spinous process of the upper cervical spine.

Preoperative radiographs showed an omovertebral bone near the superomedial angle of the

Table I. — The Cavendish score (1972)

Grade 1 very mild	The shoulder joints are level and the deformity is invisible, or almost so, when the patient is dressed.
Grade 2 mild	The shoulder joints are level or almost level, but the deformity is visible when the patient is dressed, as a lup in the web of the neck
Grade 3 moderate	The shoulder joint is elevated 2 to 5 centimeters. The deformity is easily visible
Grade 4 severe	The shoulder is greatly elevated, so that the superior angle of the scapula is near the occiput, with or without neck webbing or brevicollis.

* Department of Orthopedic and Pediatric Surgery, University Hospital, Bocage Center, 10 Boulevard Marechal de Lattre de Tassigny, 21034 Dijon Cedex, France.
** Laboratory of Anatomy, University Hospital, 2 Boulevard Jeanne d'Arc, 21000 Dijon, France.
Correspondence and reprints : E. Baulot.

left scapula on the lateral view (fig. 1). The bone had a bifid cranial extremity in contact with the spinous process of the second cervical vertebra. The anteroposterior view disclosed on the same side a second omovertebral bone lying in the rhomboid muscle between the medial border of the scapula and the left transverse process of the fifth cervical vertebra (fig. 2). This view also showed the very high position of the left clavicle. Preoperatively, magnetic resonance imaging eliminated diastematomyelia.

At the time of surgery, a Woodward procedure was performed on the left side and was combined with removal of these two omovertebral bones. The bifid bone was lying in the exact course of the levator scapulae muscle, and the second omovertebral bone was situated in the rhomboid muscle. The increase in postoperative abduction was 40° and the patient demonstrated improvement to grade 2 using the Cavendish score (3). Although the clavicle was a component of the deformity (fig. 2), an osteotomy was not necessary to achieve significant improvement. The postoperative neurovacular state was normal.

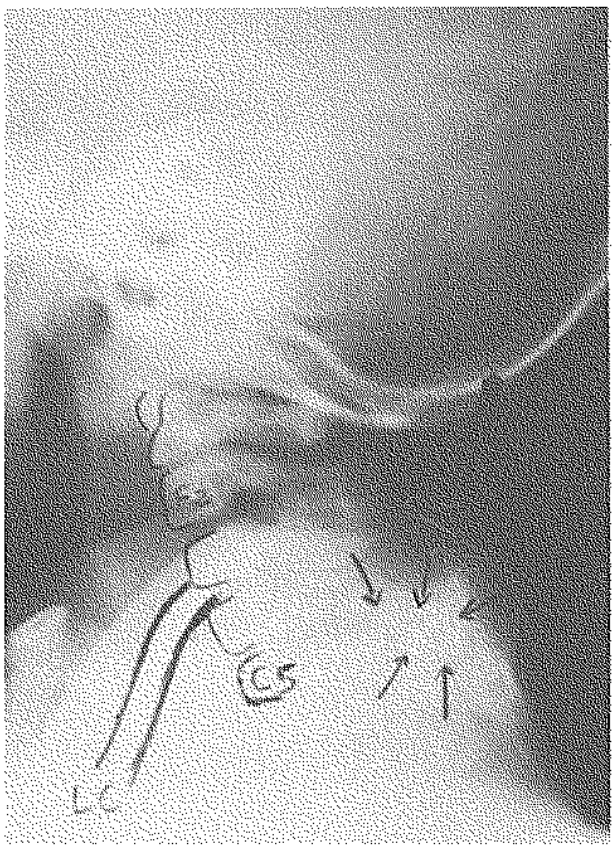

Fig. 1. — Preoperative lateral view showing a bifid omovertebral bone lying between the superomedial border of the scapula and the spinous process of the second cervical vertebra. The second omovertebral connection is also visible between the scapula and the posterior arch of the fifth cervical vertebra (large arrows). The left clavicle is short and high, up to the level of the third cervical vertebra. C2 : second cervical vertebra ; LC : left clavicle.

DISCUSSION

The case of a 4-year-old girl who presented a Sprengel deformity stimulated our interest because of an omovertebral bone lying in the levator scapulae muscle associated on the same side with a second omovertebral bone lying in the rhomboid muscle. To our knowledge such unusual findings were not reported in the literature previously.

Congenital elevation of the scapula (C.E.S), usually known as Sprengel's deformity, was first described by Eulenberg in 1863 (7). Among the numerous hypotheses regarding the pathogenesis of this anomaly the most comprehensive ones are those by Horwitz (9) and Engel (6). The arrest in caudal migration of the scapula probably occurs

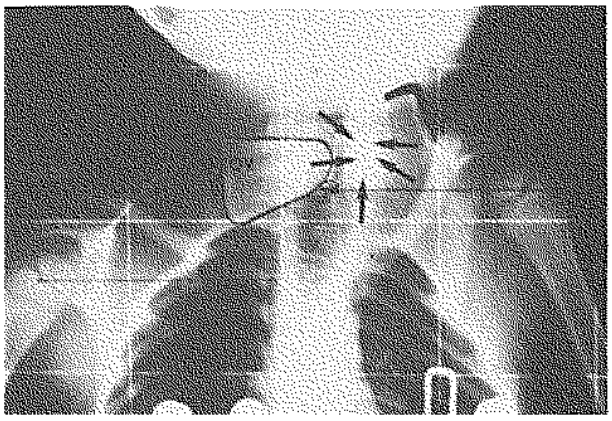

Fig. 2. — Preoperative anteroposterior radiograph showing the Sprengel deformity of the left shoulder with one of the two omovertebral bones extending from the medial border of the scapula to the transverse process of the fifth cervical vertebra (arrows). In this view, the omovertebral bone which was lying in the levator scapulae muscle is not visible. On the right side a single and large omovertebral bone (ov) is also visible on this anteroposterior view. The two superomedial angles of the two scapulae are underlined. C6 : 6th cervical vertebra.

between the ninth and the twelfth week of gestation (11).

An omovertebral bone associated with C.E.S was first reported by Willett and Walsham (15). It appears as a chondro-osseous structure usually lying in the rhomboid the trapezius muscle. Omovertebral bone usually connects the superomedial border of the scapula to the spinous process, lamina or transverse processes of the lower cervical vertebrae (11), mostly between the fourth and the seventh cervical vertebrae (2).

The percentage of omovertebral bones associated with C.E.S is variable in the literature (19% Greitemann (10), 24% Borges (1), 24% Ross (13), 30% Carson (2), 47% Crogan (5)). When present this bone is best visualized on lateral or oblique radiographs (2).

Numerous other congenital abnormalities have been described in association with CES, such as absent or fused ribs, chest wall asymmetry, Klippel Feil Syndrome, or cervical spina bifida (2, 11). For Borges *et al.* (1) congenital scoliosis is the most frequent anomaly associated with CES. Von Bazan (14) recently demonstrated a close association between CES and diastematomyelia.

For treatment, the Woodward procedure (16) seems to be the best and most popular at present for correction. This procedure allows sufficient downward relocation of the scapula after detachment of the spinal origins of the trapezius and rhomboid muscles from the spinous processes and refixation in a more caudal position, without stripping muscles from the scapula. The levator scapulae muscle is released. If present, the omovertebral bone must be systematically resected.

Many authors routinely supplemented the Woodward procedure with a clavicular osteotomy (2, 4, 10), which allows greater correction and prevents damage to the brachial plexus or compression of the subclavian artery (12) between the clavicle and the first rib. For others clavicular osteotomy must be associated in a few cases only (1, 5). This osteotomy may be performed in the middle third of the clavicle. In a very severe deformity, Jeannopoulos (11) performed an osteotomy through the base of the acromion. In our case, despite the high position of the clavicle, osteotomy of this bone was not performed and correction was fair without neurovascular complication.

CONCLUSION

In Sprengel's deformity, it is well known that impairment is never severe unless the deformity is severe. In consequence, when deformity and functional impairement are mild, it is clearly admitted that no surgical treatment is indicated. In severe cases where surgery is recommended, the orthopedic surgeon must be aware that no operation achieves perfect appearance because even if the scapula can be brought to the same level, the affected scapula remains small and deformed, and the scar is often unsightly. If present, omovertebral bone must be excised as a necessary adjunct to any surgical procedure.

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SAMENVATTING

E. BAULOT, P. TROUILLOUD, E. A. GIROUX, P. M. GRAMMONT. Sprengeldeformatie geassocieerd aan 2 ipsilaterale omovertebrale beenderen in de levator scapulae en de romboideus.

De auteurs rapporteren een originele observatie van een meisje met Sprengeldeformatie graad IV met 2 omovertebrale beenderen gelegen in de m. levator scapulae en de m. romboideus. Dit werd niet eerder beschreven. Ter gelegenheid van deze observatie vestigen de auteurs de aandacht op om een dergelijk os omovertebrale preoperatief te detecteren, daar de verwijdering ervan deel uitmaakt van de definitieve behandeling.

RÉSUMÉ

E. BAULOT, P. TROUILLOUD, E. A. GIROUX, P. M. GRAMMONT. Maladie de Sprengel associée à deux os omovertébraux homolatéraux dans le muscle levator scapulae et le muscle rhomboïde.

Les auteurs rapportent l'observation originale d'une fillette atteinte d'une maladie de Sprengel de grade 4, associée à 2 os omovertébraux homolatéraux localisés dans le muscle levator scapulae et dans le muscle rhomboïde. Une telle association n'a jamais été décrite dans la littérature. A l'occasion de cette observation les auteurs insistent sur la valeur de la détection préopératoire des os omo-vertébraux dans la maladie de Sprengel car leur excision fait partie du traitement.