

Shoulder duplication in constriction band syndrome: a case report

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Abstract

A 2.5 year old girl is presented with both hands constriction bands leading to distal amputations and the rare deformity of shoulder duplication in the right side accompanying constriction skin marking over the affected shoulder. The cephalomedial scapula articulated with the clavicle and the caudolateral scapula articulated with humeral head. The most important physical finding which could explain the pathophysiology of this rare anomaly, was constriction band marking over the right shoulder. Shoulder range of motion was limited but still functional and no surgical intervention was required for the scapular duplication.

Keywords: Shoulder duplication, constriction band syndrome.

Introduction

Complete scapular duplication is an exceedingly rare anomaly, in which only 4 cases have been reported in the literature, three of which involved the right and one involved the left scapula. There have been associated malformations in all 4 cases including foot deformities in three, hand syndactylies in two, and constriction band in shoulder girdle area in one case. We report a case of complete duplication of right scapula with hand syndactylies (acro-syndactyly in the left) and sign of constriction band in right shoulder girdle area.

Case report

The patient was a 2.5 year-old girl with no familial history of congenital anomaly and product of an uneventful normal vaginal delivery. The patient had been referred

for treatment of both hands syndactylies. On physical examination she had a bony prominence in right shoulder girdle corresponding with distal clavicle and a dimple on the skin of proximal arm and shoulder as a remnant of a constriction band (Fig. 1). Elevation of the right shoulder was limited to 80° but shoulder rotation was minimally affected. Also the elbow and wrist had full active range of motion. The right hand was involved with pre-axial polydactyly and symbrachydactyly (two phalanx fingers) (Fig. 2). The left hand had acrosyndactyly and constriction band tip computation of the fingers. The shoulder as well as elbow and wrist were normal in the left side and both upper extremities had equal length. There was no spine-pelvis or lower extremity abnormality.

Radiograph of the affected shoulder is presented in Fig. 3. There were two completely separated scapulas in the right side;

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Fig. 1. Right shoulder deformity. Arrowhead: Prominence of the right clavicle; Arrow: Remnant of constriction band and skin dimple.

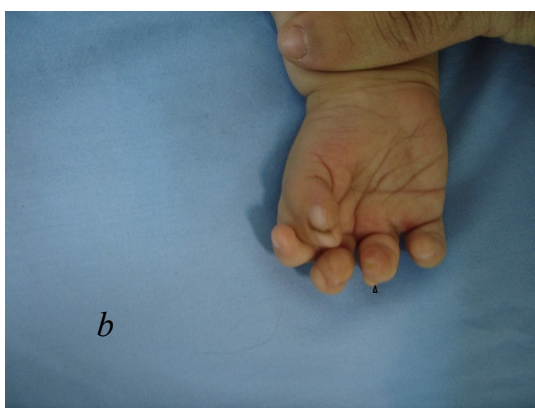


Fig. 2 **a**: Dorsal view of the right hand. Consider constriction bands; **b**: Volar view of the right hand. Consider preaxial polydactyly and skin dimples (arrowhead).

the larger craniomedial scapula was deficient in glenoid and articulated with the clavicle through a hypoplastic acromion. The lesser caudolateral scapula had a well differentiated acromion and glenoid and articulated with the proximal humerus. The

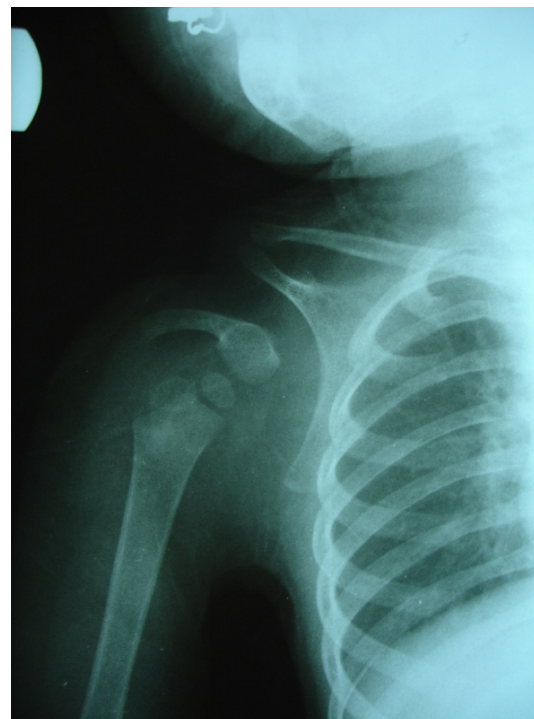


Fig. 3. Radiography of the right shoulder. The scapula is duplicated; the larger medial part containing coracoid is articulated with clavicle and the smaller lateral part is articulated with proximal humerus.

latter scapula showed no body or spine development. There was no bony connection between these two scapulas.

Discussion

The scapula begins to differentiate from the fourth, fifth and sixth cervical vertebrae and develops through one primary ossification center which appears between 45th to 60th days of gestation and seven secondary ossification centers [1,2]. Only the body and the spine of the scapula are ossified at birth. The majority of the scapula forms by intramembranous ossification. All secondary ossification centers fuse as late as age 22.

Congenital abnormalities of the shoulder are rare; Sprengle's deformity is the most common one. These deformities are frequently unnoticed by both physicians and patients. To date only 4 cases of complete scapular duplications have been described; Martini and Neusel first reported on two Children, both males who had duplication of their right scapulas [3]. In the first,

there was a constriction band presented in the region of the right shoulder and also acrosyndactyly of the right middle and ring fingers. The scapula was normal in the left side and the patient had scoliosis as well as bilateral club feet. He underwent scapular cerclage wiring when he was 9 month-old.

The second case was a 4 year old male again demonstrated right scapular duplication, and a bony bridge connected the two scapulas. Both hands had syndactylies, scoliosis and neural arch defects were presented as well as left foot hypoplasia and fragmented left capital femoral epiphysis. This boy also underwent surgery when he reached 5 and the bony bar resected and the two scapulas were joined. Stacy and Yousefzade at 1999 reported on a malformation of left upper extremity in a 2 month old patient with left doubled scapula [4]. The proximal humerus did not articulate with any of the scapulas, resembling proximal femoral focal deficiency. The larger craniomedial scapula articulated with the left clavicle.

The last case was presented by M. L. Sanchez Alegre et al in 2003 which was a 5 month old girl with duplicated right scapula [5]. The only associated malformation was a

congenital equinovarus foot deformity.

The case that we report here is similar to the first case described by Martini and Neusel in regard to involvement of the right side, evidence of constriction band in the affected shoulder, acrosyndactylies of the hands and radiographic characteristics. In our patient the shoulder was functional and no surgery required. Also there was no spinal or lower extremity malformation.

Although abnormalities of epiphyseal growth explain some anomalies of the scapula, we agree with M. L. Sanchez in that the constriction band could be regarded as the probable etiology in our case.

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