Congenital Elevation of the Scapula Associated with a Dislocated Glenohumeral Joint

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Congenital elevation of the scapula (Sprengle's deformity) is a rare entity resulting from an aberration in the caudal descent of the scapula. The glenohumeral articulation is intact but the range of motion may be restricted. Surgical interventions are aimed at improving cosmesis or improving range of motion. We present the case of a six years old male child with congenital elevation of his left scapula in association with a dislocated left glenohumeral joint and absent range of motion that significantly improved following a scapular resection.

Keywords: congenital elevation, dislocated glenohumeral joint, glenohumeral joint, scapula.

A six-year-old male child presented to us with a huge lump on the left side of his neck with loss of the left shoulder contour since birth **(Figure 1A & 1B)**. The patient did not have any active movements of his affected non-dominant left shoulder, but passive motion was possible. History revealed that the lump had been present since birth and gradually enlarged causing cosmetic concerns. There had been no active range of motion in the affected extremity, but neurovascular status was intact. Plain radiographs and CT studies showed a high riding left scapula with complete dislocation of the left glenohumeral articulation with the humeral head lying below the inferior angle of

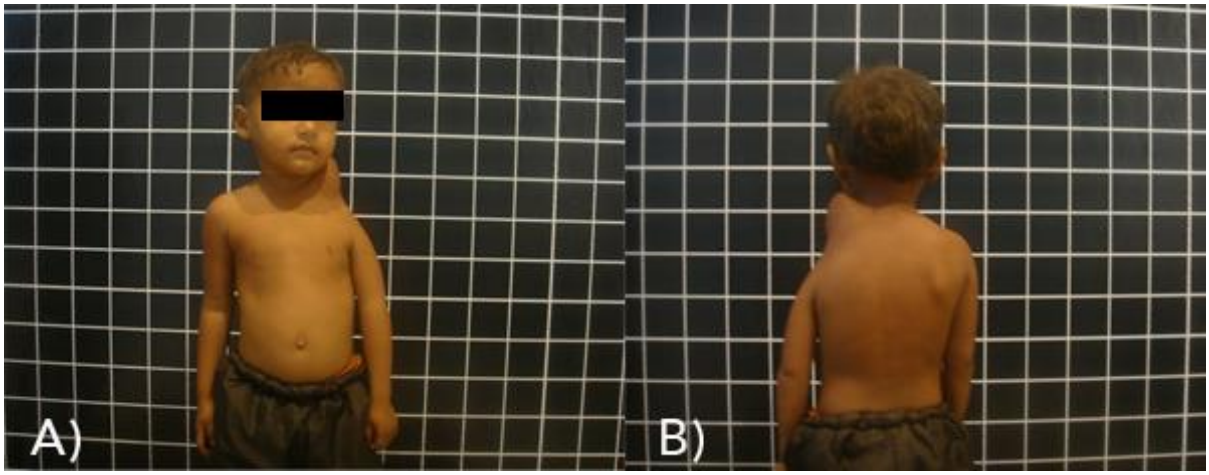

Figure 1: The large lump on the left neck and shoulder region as seen from A) the front and B) behind. Glenohumeral motion was absent on this side.

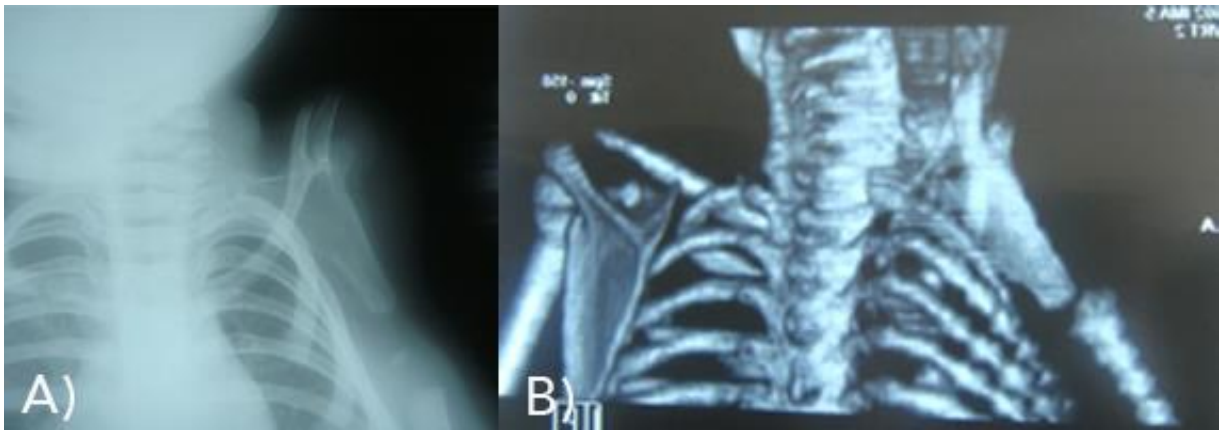

Figure 2: A) Anteroposterior radiograph and B) 3-D CT reconstruction showing an elevated left scapula with complete dislocation of the left glenohumeral joint.

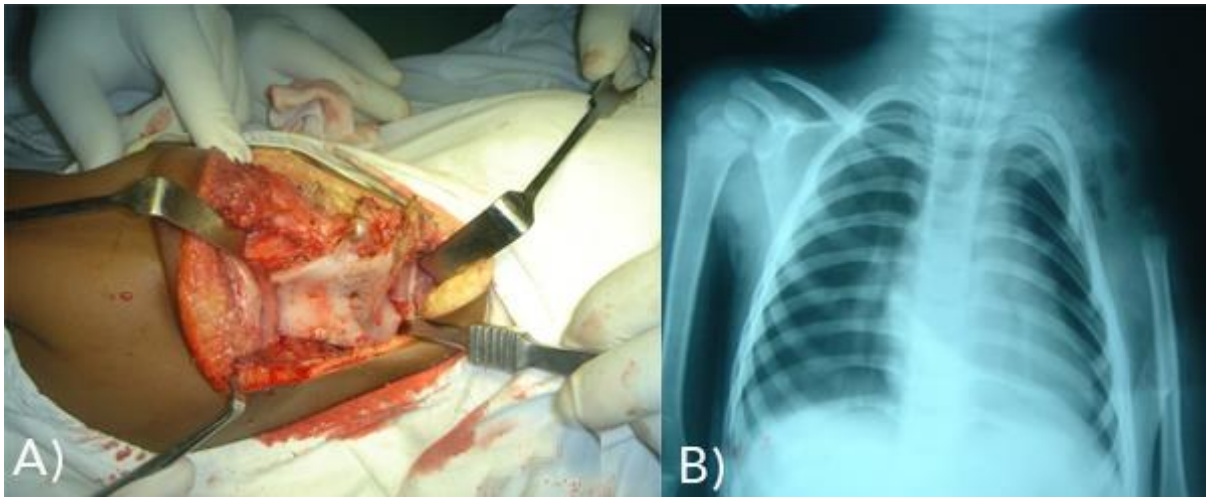

Figure 3: A) Left scapulectomy in progress; removed specimen showed a hypoplastic scapula with dysplastic glenoid and aplastic coracoid B) Post-scapulectomy radiograph.

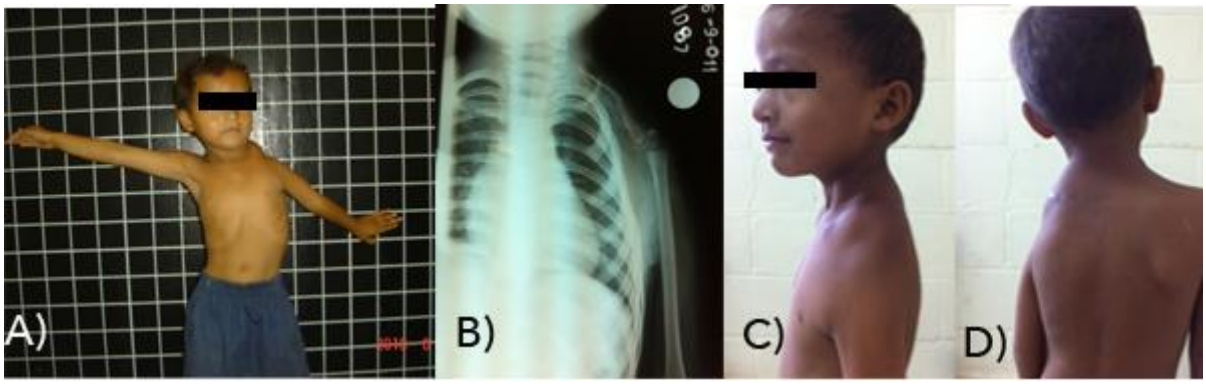

Figure 4: A) Six months post surgery, the child had regained functional range of motion with 800 of abduction B)Looked cosmetically better but C) Left shoulder contour remained flattened D) Radiograph at six months showing regeneration of the left scapula in the periosteal sleeve.

scapula **(Figure 2A & 2B)**. There was no associated omovertebral connection. The patient didn't have any other associated anomalies. In view of the cosmetic deformity, lack of range of motion of the affected shoulder, restricted neck range of motion on the affected side and concerns of neurovascular impingement if the mass was allowed to progress, a decision to undertake surgical resection of the mass was reached. With the patient in a lateral decubitus position, an incision was made over the mass and a left claviculo-scapular resection was performed subperiosteally **(Figure 3A & 3B)**. The removed specimen revealed a hypoplastic scapula with dysplastic glenoid and poorly defined coracoid. Post-operative recovery was uneventful and range of motion exercises was instituted. To our great surprise, the patient regained functional range of motion of the affected shoulder and neck, including active abduction of 800 **(Figure 4A)**, over a period of six weeks and the cosmetic appearance was much better **(Figure 4B)** although shoulder contour remained flat **(Figure 4C)**. At six months follow-up the child was fully functional with the affected extremity and could carry out all activities of daily living consistent with his age. Radiographs at six months showed evidence of regeneration of the left scapula and clavicle in the periosteal sleeve **(Figure 4D)**.

Discussion

High riding scapula (Sprengle deformity) is the commonest congenital anomaly affecting caudal descent of the scapula during development, often associated with other congenital anomalies like Klippel-Feil syndrome and congenital chest wall anomalies.[1-3] A bony, cartilaginous or fibrous omovertebral connection linking

the superomedial scapula to the lower cervical spine, the omovertebral body,[4] is present in up to 50% of cases and rarer variants like omo-clavicular and omo-occipital connections of the undescended scapula have also been described.[5] The etiology of failure of descent of the growing scapula is thought to be a result of omovertebral connections resulting from abnormal epiphysis originating from the superior angle of scapula or by factors such as increased intrauterine pressure or increased CSF permeability of the fourth ventricle leading to aberrations in mesodermal development.[6,7] Cavendish1 classified these lesions depending on their clinical appearance whereas Rigault[8] classified them based on the degree of proximal arrest. The main problems with this deformity are cosmetic unsightliness and limitation of shoulder motion, mainly abduction. To the best of our knowledge, there has been no report of a high riding scapula in association with a completely dislocated glenohumeral joint.

The indications to intervene surgically, as was in our case, are cosmetically unacceptable deformity and lack of range of motion of the affected shoulder girdle and neck. We also felt that leaving the lesion as it were could risk traction on the neurovascular structures as the mass enlarged. Surgical treatments range from excision of the prominent medial part of the scapula to more extensive procedures where the scapula is brought down to the desired level and muscles realigned to stabilize it there.[9-11] We deemed it necessary to perform a total scapulectomy and partial claviculectomy to ensure the neck range of motion would be unobstructed by the mass, to eliminate any evolving traction on the neurovascular structures and in the hope of achieving a better cosmetic appearance. The restoration of the shoulder joint motion post-operatively was unanticipated and most gratifying to both the patient and his family and our team. The implications of the re-growth of the removed scapula which became evident by six months is unclear and the question of whether an extraperiosteal excision should have been carried out will only be answered on longer term follow up.

Conclusion

In summary, congenital elevation of scapula is a rare disorder and its association with a dislocated glenohumeral joint has not been described in the literature to the best of our knowledge. Total resection of the scapula has led to excellent range of motion gain from zero to functional active motion of the affected shoulder and a better cosmetic result at short term follow up.

Longer term follow up until skeletal maturity will define the role and

effectiveness of such radical surgery in patients presenting with this rare disorder.

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