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Sprengel's deformity of the shoulder: A Case report

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Abstract

Congenital elevation of the scapula i.e. 'Sprengel's shoulder' is a congenital anomaly of the shoulder girdle that is associated with abnormal descent, and malrotation of the scapula.

Conventional anteroposterior radiography of the chest including both shoulders along with CT or MRI is required for diagnosis and proper management. Most common therapeutic choice is surgical intervention for cosmetic and functional recovery of the shoulder.

Surgical procedure: With the patient in prone position, in general anaesthesia, Skin incision was given paraspinal area, along superior-medial angle and medial border of the left scapula. After detaching supraspinatus muscle from the scapula along with its periosteum, omovertebral bar was excised, leaving remnant at the connection from the spinous process of c3 vertebra. Then muscles were reattached to the scapula in corrected position.

Result: As follow-up after 6 weeks, the patient's active range of motions improved. Forward flexion was improved from 60° pre-operatively to 120° at six months follow-ups. Similarly abduction improved from 60° to 130°, internal rotation from 15° to 20° and external rotation 0° to 30°. Neurovascular evaluation was within normal limits. X-rays confirmed the position of scapula about 3 cm lower than the pre-operative position which is less than 1 cm higher as compare to right scapula.

Keywords: Floating hip, acetabulum fracture, femur fracture

Introduction

Congenital elevation of the scapula i.e. 'Sprengel's shoulder' is a congenital anomaly of the shoulder girdle that is associated with abnormal descent, and malrotation of the scapula. The deformity is usually associated with muscle hypoplasia or atrophy. Combination of these bone and muscular factors results in deformity and functional limitation of the shoulder [1].

Failed migration of the scapula from neck to thoracic region during early embryonic life [2] leads to the scapula that protrudes in the neck. However, the deformity is not only a cosmetic concern, but more importantly it causes restricted range of motion of the affected shoulder and the cervical spine [3]. Sprengel deformity appears either as a single abnormality or in association with other abnormalities, like- Klippel-Feil syndrome, spina bifida, kyphoscoliosis, torticollis, underdevelopment of clavicle or humerus and rib defects [4].

Cavendish's classification is based on clinical findings of sprengel's shoulder. It affects males and females equally. It is associated with an omovertebral connection in between cervical spine and superior-medial angle of scapula in 20-25% of cases [5]. The omovertebral connection can be fibrous or osseous.

Conventional anteroposterior radiography of the chest including both shoulders is a simple and effective means of diagnosing Sprengel deformity, particularly when combined with appropriate clinical examination. Computed tomography (CT) with three-dimensional (3-D) reconstruction and magnetic resonance imaging (MRI) is done nowadays to diagnose coexisting abnormalities and treatment planning. Although the management of Sprengel deformity depends on the severity of the abnormality and the degree of motion restriction, the most common therapeutic choice is surgical intervention for cosmetic and functional recovery of the shoulder [4].

Case Report

A 10-year-old girl presented with a short neck and lumps on left side of her neck. On physical examination, neck deviated towards same side and scoliosis towards right side. She had limited range of movement of the cervical spine, 60° abduction, 60° forward flexion, 0° of external rotation of left shoulder, without neurovascular compromise. [Fig-1, 2, 3] The shoulder asymmetry was of 6 cm, i.e. grade-4 according to Cavendish's classification.

She was first advised an anteroposterior X-ray of the thorax and cervical spine, which demonstrated asymmetrically high-positioned scapulae, in conjunction with cervical vertebrae synostosis and thoracic cage deformities and presence of omovertebral bar. [Fig-4] Surgery was decided and a CT scan was performed to delineate the anatomy in detail, assess the degree of asymmetry, and provide the details for morphometric analysis and surgery planning. [Fig-5, 6]

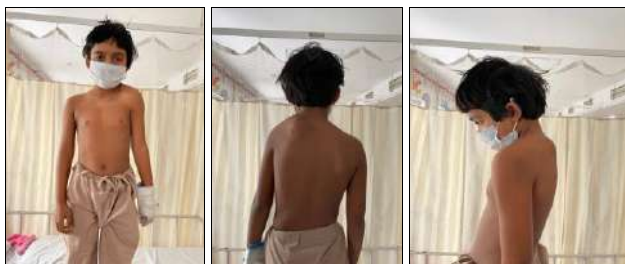


Fig 1, 2, 3: shows Deformity of left shoulder with deviation of neck towards left and Kyphoscoliotic changes. (Cavendish's: grade-3)



Fig 4: X-Ray shows left side elevated scapula with omovertebral bar

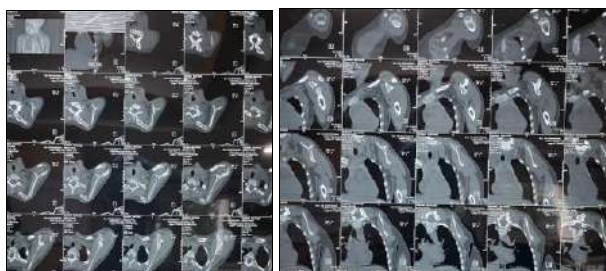


Fig 5, 6: CT scan of Shoulder Showing Sprengel deformity of left shoulder associated with cervical and thoracic spine deformity. Rigault classification grade 3 (superio-medial angle above C5 transverse process)

Surgical Procedure

With the patient in prone position, in general anaesthesia, (Fig- 7) Painting and draping was done to have an aseptic field. Skin incision was given paraspinal area, along superior-medial angle and medial border of the left scapula. Muscles were detached from their scapular insertion, the trapezius muscle was then elevated extraperiosteally and reflected medially, thus exposing underlying muscles and the scapula. The supraspinatus muscle was then detached from the scapula along with its periosteum. The omovertebral bone was excised, leaving remnant at the connection from the spinous process of c3 vertebra.(Fig-8,9)

The resected bone was of 5 * 2 cm in size.(Fig-10) The insertions of the levator scapulae muscles on the superior angle of the scapula and of the rhomboid muscles on the medial border of the scapula were dissected. The supraspinous fossa of the scapula was resected, taking care to avoid injury to the suprascapular neurovascular bundle. The scapular attachments of the latissimus dorsi and serratus anterior muscles were detached from the anterior aspect of the scapula. The scapula was then displaced distally down to the level of the normal side using the superomedial angle of the scapula as a landmark rather than its tip because of constant scapular hypoplasia. Once the scapula was in its corrected position, the muscles were reattached to it, the supraspinatus muscle to the base of the scapular spine. Rhomboid muscles were reinserted without compromising the new position of the scapula in the frontal plane by lengthening it if necessary, in order to achieve good orientation of the glenoid and therefore to increase the range of abduction. (Fig-11)



Fig 7: Positioning of the patient under General anaesthesia. The operating upper limb should be free to check intra-operative gliding movements of scapula



Fig 8, 9: Exposure of the bone connection in between scapula and transverse process of c3 vertebra



Fig 10: Excised bony bar (omovertebral bar)

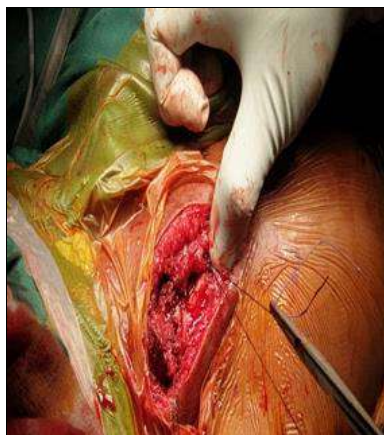


Fig 11: Repairing of Supraspinatus and Rhomboid muscles to the lowered down scapula

Follow-Up

Patient was discharged on the 3rd Post-Operative day. The sutures were removed on 14th post-operative day.

Patient was followed up in total 6 visits over 6 months. Check X-ray were done post-operatively and at 6 weeks & 6 months. (Fig-12,13,14) Physiotherapy in the form of shoulder range of motion exercises were started after 6 weeks.

Table 1: Shoulder range of motions in pre-operative and post-operative follow-ups

Follow-ups	Forward Flexion	Abduction	Internal Rotation	Ext. Rotation
PRE-OP	60°	60°	15°	0°
POD-6 weeks	90°	80°	15°	10°
POD-3 months	110°	110°	20°	30°
POD- 6 months	120°	130°	20°	30°



Fig 12, 13, 14: post-operative (6 month) follow-up range of motion of shoulder

Results

As follow-up after 6 weeks, the patient's active range of motions improved. Forward flexion was improved from 60° pre-operatively to 120° at six months follow-ups. Similarly abduction improved from 60° to 130°, internal rotation from 15° to 20° and external rotation 0° to 30°. Neurovascular evaluation was within normal limits. X-rays confirmed the position of scapula about 3 cm lower than the pre-operative position which is less than 1 cm higher as compare to right scapula.

Discussion

Sprengel deformity is rare condition, but it is the commonest congenital defect of the scapula. The cause of

sprengel deformity is unknown and there have been only very few cases reported of familial Sprengel deformity, but association is not established yet. Although Kadavkolan reports that it occurs equally in both sexes but most authors agree that the undescended scapula affects female three times more often than men [6]. It can occur on both sides concurrently but most often it is unilateral, with a predilection for the left side. Estimation of the height difference between the normal positioned scapula and elevated scapula using the inferior angle of the scapulae on an anteroposterior X-ray of the chest is not always accurate for the estimation of the severity of the deformity and for choosing the appropriate treatment. Detailed anatomy of abnormal scapula and glenoid orientation with CT scan and 3D reconstruction should be advised followed by proper management.

Gain in range of abduction's mean value ranges from 19° to 70° in various studies, with a greater gain seen in patients who has more severe preoperative restriction [7, 8, 9].

In this case Forward flexion was improved from 60° pre-operatively to 120° at six months followups. Similarly abduction improved from 60° to 130°, internal rotation from 15° to 20° and external rotation 0° to 30°, which is comparable to the previous studies.

In many studies, surgical complications documented were scar-related complications, brachial plexus palsy, brachial neuritis, winging of the scapula, regeneration of the omovertebral bar, recurrence of the deformity and prominence of the sternoclavicular joint [10, 11].

In this case, Neurovascular evaluation was within normal limits, without any major complication. Patient's functional outcome improved in satisfactory level.

We need to take longer follow-ups till adulthood for this child. More number of cases need to be enrolled to establish effectiveness of this procedure.

Disclosure

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Conflict of Interest: None

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