

Case Report

Sprengel shoulder: a case report

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ABSTRACT

Sprengel shoulder is a rare deformity of shoulder girdle of unknown etiology. Eulenburg in 1863, had first reported this rare congenital anomaly of the scapula. Sprengel described it in 1891, and hence this deformity got its' name. Autosomal dominant inheritance has been reported for this abnormality. Abnormal descent of the scapula during 9th to 12th weeks of gestation leads to this congenital anomaly. It is a cosmetic deformity. Elevation of scapula is the characteristic feature. A lump at the back of the neck with restricted movements of the shoulder and or arm are the presenting finding. The affected scapula is rotated on its sagittal axis. The superior and the vertebral border comes nearer to the medial line and to the axilla respectively. Severity of this deformity varies widely being almost invisible when covered with clothes to the shoulder being elevated over 5 centimeters along with the presence of webbing of neck. This congenital deformity can be associated with other musculoskeletal abnormalities such as Klippel-Feil syndrome, scoliosis and rib anomalies. In 30% of cases of Sprengel shoulder, an omovertebral cartilaginous, fibrous or bony bar connecting superomedial border of scapula to spine, transverse process or lamina of cervical vertebra can be seen and this causes restriction of movements around the shoulder girdle. Surgical management is considered in severe cases. Surgical management involves excision of the protruding portion of scapula and inferior translation of scapula. This enhances shoulder functions as well as general appearance of the patient.

Keywords: Shoulder, Ribs, Spine, Scapula, Scoliosis

INTRODUCTION

Sprengel shoulder is a rare deformity of shoulder girdle of unknown etiology. Eulenburg in 1863, had first reported this rare congenital anomaly of the scapula.¹ Sprengel described it in 1891, and hence this deformity got its' name.² Autosomal dominant inheritance has been reported for this abnormality. Abnormal descent of the scapula during 9th to 12th weeks of gestation leads to this congenital anomaly.³ It is a cosmetic deformity. Elevation of scapula is the characteristic feature. A lump at the back of the neck with restricted movements of the shoulder and or arm are the presenting finding. The affected scapula is rotated on its sagittal axis. The superior and the vertebral

border comes nearer to the medial line and to the axilla respectively. Severity of this deformity varies widely being almost invisible when covered with clothes to the shoulder being elevated over 5 centimeters along with the presence of webbing of neck. This congenital deformity can be associated with other musculoskeletal abnormalities such as Klippel-Feil syndrome, scoliosis and rib anomalies.⁴ In 30% of cases of Sprengel shoulder, an omovertebral cartilaginous, fibrous or bony bar connecting superomedial border of scapula to spine, transverse process or lamina of cervical vertebra can be seen and this causes restriction of movements around the shoulder girdle.⁷

Surgical management is considered in severe cases. Surgical management involves excision of the protruding portion of scapula and inferior translation of scapula. This enhances shoulder functions as well as general appearance of the patient.⁵

CASE REPORT

An 8 years old male child was examined in the department of pediatrics, coming with bilateral webbed neck more pronounced on the right side. There was a slight degree of abduction of right shoulder joint. Examination from the back revealed hypoplastic shoulder more pronounced on the right side.

On plain radiograph, it was seen that the superior border of both the scapula are higher up more on the right side. There was lateral bending of the cervical column of vertebrae with convexity on right side. There was crowding of the transverse processes of the cervical vertebrae along with fusion of their lateral masses on the left side. There was decrease in distance between convex medial borders of both the scapula. Inferior angles of both the scapula were found at the level of T4 on right side and T6 in case of left side and the angles are very prominent. On radiograph no omovertebral bar was seen. No cardiopulmonary abnormality was seen and all movements were intact except abduction at shoulder joint. No associated other anomalies were found.



Figure 1: Sprengel's shoulder.



Figure 2: Radiograph of Sprengel shoulder.

DISCUSSION

Harvey et al study shows Sprengel shoulder shows decrease cosmetic deformity and shoulder function.⁵ From the physical examination cosmetic deformity and shoulder dysfunction have been observed in this case too. We can say it as mild Sprengel deformity if after clothing no cosmetic deformity seen but severe if cosmetic deformity is visible after clothing along with shoulder dysfunction. Mild cases do not require surgery but severe cases are required surgical approach.

According to Sulamaa et al it is a very rare skeletal deformity. It may be both congenital and acquired but congenital variety is most common. The proportion of bilateral and unilateral cases is 1:10.⁶ But from our present finding we have found this deformity as bilateral, more pronounced on the right side. Here it is congenital which is due to non-descendent of shoulder blade.

According to Pellegrin et al this deformity is characterized by high elevation of scapula with medial rotation of inferior angle.⁸ This finding coincide with our present findings. Due to elevation of scapula on right side the clavicle is found as more vertical position than on the left side along with higher elevation of right shoulder joint.

Bejiqi et al found fusion of two or more vertebrae with short neck and limited neck movement.⁹ This coincide with our finding. There is overlapping of transverse processes of cervical vertebrae on left side with shortening and webbing of neck on left side which may lift up the shoulder girdle on right side.

CONCLUSION

With the help of plain radiograph and clinical examination physician can come into instant diagnosis of lump on back of the neck associated with difficulty in lifting arm in some daily work for an example combing hair by brush or lifting objects. With this knowledge pediatric surgeons can plan for operative maneuver to remedy it.

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