International Journal of Current Research and Review
DOI: https://doi.org/10.31782/IJCRR.2025.17602

Case Report

IJCRR
Section: Healthcare

Anatomical and Embryological Aspects of Sprengel Scapula–A Case Report

Gagandeep Kaur[1], Seema Sehmi[2]

[1]PG student, Department of Anaesthesia, Christian Medical College, Ludhiana, India; [2]Professor, Department of Anatomy, Sri Guru Ram Das Institute of Medical Sciences and Research, Amritsar, India.

ABSTRACT

Introduction: Sprengel scapula is an uncommon congenital abnormality in which scapula fails to descend down to the normal position.

Case Report: A boy aged 8-year- old presented with Sprengel scapula on right side. Posteroanterior Chest radiograph was taken and this anomaly was confirmed.

Conclusion: A detailed knowledge of anatomical basis will help orthopedic surgeons to improve the success rate and lowering the complications of different surgical procedures.

Key Words: Sprengel, Embryology, Congenital, Scapula, Anatomical, Surgical Procedures

INTRODUCTION

Sprengel deformity is a rare congenital deformity of shoulder girdle in which there is undescended, hypoplastic and malrotated scapula.[1,2] Female to male ratio of this deformity is about 3:1.[2] Most limited shoulder movement in this anomaly is abduction movement.[3] Initially it was diagnosed by Eulemberg in 1863.[4] Later on Willet and Walsham[5] identified two cases explaining the anatomical basis and its association with rib and spine deformity. Sprengel[6] in 1891 described four cases and disease was named after his name. The scapula develops along with upper limb during fifth week in lower cervical or upper scapular region and descends to final anatomical position by 12th week of gestation.[7] Sprengel scapula is a small deformed scapula with horizontal diameter that exceeds vertical diameter[8], forward curvature of the superior angle of the scapula over the apex of thorax, abutment of the medial scapular border against spinous processes of adjacent vertebrae, anterior curving of supraspinous portion of the scapula and prolongation of superior medial scapular angle with omovertebral bone.[9] Broca described a scapular index to describe relationship between the vertical and horizontal diameters of scapula. Others used subscapular angle, supraspinous angle and infraspinous angle as an indicator of deviation from normal.[8] The proportion of bilateral and unilateral Sprengel scapula cases is 1:10.[10] Cavendish[11] classified the deformity into four grades describing the indications for treatment. They are-

Grade I –Shoulders at the same level. Deformity invisible under clothing.

Grade II- Shoulders at the same level. Deformity visible when patient is dressed as lump at the back of neck.

Grade III- Asymmetric shoulders. The deformed shoulder is raised by 2-5cm.

Grade IV- The affected shoulder is raised by more than 5cm so that superior angle is close to occiput. Rigault and Poliquen[12] gave radiological grading depending on the position of medial angle of scapula.

Grade I –Between the transverse processes of T2 and T4.

Grade II-Between transverse process of C5 and T2.

Grade III-Above the transverse process of C5.

CASE REPORT

A boy-aged 8-year-old came in outdoor Orthopaedics department at Govt Medical College Amritsar with the elevation of shoulder on right side. There was a lump at the back of the neck with right sided restricted movements of shoulder and

Corresponding Author:
Dr. Seema Sehmi, Professor, Department of Anatomy, SGRDIMSR, Vallah, Amritsar (Punjab).
Ph: 9914754354; Email-drseema16@gmail.com

ISSN: 2231-2196 (Print) **ISSN:** 0975-5241 (Online)

Received: 12.02.2025 **Revised:** 05.03.2025 **Accepted:** 16.03.2025 **Published:** 23.03.2025

arm. No other associated musculoskeletal abnormality was noted. X ray chest PA view was done and Sprengel scapula on the right side was diagnosed. (Fig 1)

Figure 1: Radiograph showing Sprengel Scapula on right side.

DISCUSSION

Embryology

Scapula appears as a mesenchymal mass at fourth and fifth cervical vertebral level at 6th week of intrauterine life. It starts migrating caudally to its definitive anatomical position with inferior scapular angle between sixth and eighth thoracic vertebra by 12th week of intrauterine life. During caudal migration scapula undergo morphological changes to adapt to prehensile functions of upper limb.[13] If this migration is interrupted, the condition is known as Sprengel Scapula. The cause is still unknown and it can be vascular [14] as interruption of blood supply from subclavian artery lesions[15] or some familial cases are seen suggesting genetic transmission[16] or as autosomal dominant by some authors.[17] This is also associated with Klippel-Feil Syndrome and muscle abnormalities around shoulder girdle[18,19], cardiopulmonary and genitourinary abnormalities.[20,21] The abnormal bone known as omovertebral bone is an abnormal connection between posterior cervical spine and superomedial border of scapula, is often associated with Sprengel deformity. Possible etiology of omovertebral bone includes overgrowth of spinous process of scapula and ossification of connective tissue in intermuscular planes between spine and scapula.[19] A number of bone abnormalities associated with Sprengel abnormalities include costal elements like aplasia, hypoplasia, synostosis, bifidity, supernumerary ribs, clavicular anomalies like variations in shape and position, vertebral abnormalities like cervical and thoracic spina bifida occulta, hemivertebrae and secondary scoliosis. Extraosseous abnormalities associated with Sprengel scapula include Diastematomyelia, torticollis, Cleft Palate, *Pterygium colli*, renal abnormalities, Poland syndrome and Gorlin-Goltz Syndrome.[16]

Anatomical Basis of Treatment

Most important muscle resisting upward directed forces of levator scapulae and Rhomboids on scapula is Trapezius muscle and this muscle is found atrophied in Sprengel deformity.[11] A number of factors affect the success of surgical outcome like age of patient, functional impairment as well as co morbities like torticollis, short neck, severe congenital scoliosis and neurological involvement. Surgery should be avoided in patients with short neck and Grade 4 deformity.[19] To prevent brachial plexus injury while mobilizing the scapula in patients more than eight years, a 2-cm mid clavicular resection osteotomy is recommended. The resected part can be morcellized and placed back into periosteal sheath. In younger patient, one cm clavicular resection osteotomy is performed without replacing the bony gap.[22] Initially procedures involved release of medial muscle attachments, lowering the scapula bone and anchoring the lower pole of scapula to the rib or surrounding muscles. Procedures involved were Putti or Putti-Schrock and Green's procedure. Putti procedure involved lowering of the scapula and fixing to the rib and is uncommon and obsolete now. Schrock's modified procedure involved osteotomy of the supraspinous region and acromion base to facilitate scapular descent. [13] Green technology involved extra periosteal detachment of superior and medial scapular muscles, excision of supraspinous portion, lowering of the scapula and reattachment of the muscles. Modified Green's procedure included the placement of the inferior angle of scapula in a pocket created beneath latissimus dorsi.[23,22] This was considered as a difficult technique associated with wide ugly keloid. In order to simplify the technique with least complications, Woodward technique was employed. It involved the release of trapezius, levator scapulae and rhomboids from the origin from spinous processes, taking the scapula lower down in a pocket and reattaching all the muscles. But this technique did not cause any change in muscular insertion and scapular rotation.[24,25] Mears's procedure involved muscular detachment at scapular origin and excising superomedial angle of scapula. It also included osteotomy through body of the scapula to avoid any impingement of the residual portions during abduction. This required the release of the long head of triceps which act as passive limitation of the abduction.[26,27] Another approach is partial Scapulectomy which included the removal of superior angle of the scapula. This excision can be done separately or in association with other procedures. It had a role in mild disfigurement otherwise it did not improve the scapular dynamics.[28] Less commonly used procedure was vertical scapular osteotomy involving vertical paramedian incision in which scapula is osteotomized about 1cm from the vertebral border. The muscles attached to the superomedial angle of the scapula were detached. The major lateral portion is lowered

and reattached. Most important technique in this procedure was Konig-Wittek technique.[29] But there was poor functional improvement, difficulties in osteotomy, postoperative keloid formation and bony irregularities.[30]

CONCLUSION

Sprengel deformity is a rare congenital abnormality of shoulder girdle causing functional and cosmetic deformity. The other associated congenital anomalies often determine the management and outcome of treatment. Anatomy of this deformity as well as surgical techniques discussed provide a wide review helping the orthopaedic surgeons to adapt appropriate line of treatment for successful outcome with low complication rates.

ACKNOWLEDGEMENT

We acknowledge the immense help from the authors whose articles are cited here.

Conflict of Interest: No conflict of interest

Source of Funding: None

Authors' Contribution:

Concept, Design and Drafting of the manuscript-Gagandeep Kaur

Supervision-Seema Sehmi

REFERENCES

1. Harvey EJ, Bernstein M, Desy NM, Saran N, Ouellet JA. Sprengel Deformity: Pathogenesis and management. J Am Acad Orthop Surg.2012; 20:177-86.
2. Grogan DP, Stanley EA, Bobechko WP. The congenital undescended scapula. Surgical correction by the Woodward procedure. J Bone Joint Surg Br.1983; 65: 598-605.
3. Walstra FE, Alta TD, Vander Eijken JW, Willems WJ, Ham SJ. Long-term followup of Sprengel's deformity treated with the Woodward procedure. J of Shoulder and elbow Surg. 2013;22(6):752-59.
4. Eulenberg M. Beitrag Z. Dislocation of the scapula. Official report on the German National and Physician Association for the years. 1863; 37:291-94.
5. Willett A, Walsham WJ. A second case of malformation of the left shoulder girdle, removal of abnormal portion of bone with remarks on the probable nature of the deformity. Med Chir Trans. 1883; 66:145-58.
6. Sprengel RD. Congenital Upward Displacement of the scapula Arch Clin Surg.1891; 42: 545-49.
7. Tsirikos AI, McMaster MJ. Congenital anomalies of the ribs and chest wall associated with congenital deformities of the spine. J Bone Joint Surg Am 2005; 87: 2523-536.
8. Horwitz AE. Congenital elevation of the scapula-Sprengel's deformity. J Bone Surg Am. 1908; s2-6:260-311.
9. Carson WG, Lovell WW, Whitesides TE. Congenital elevation of the scapula. Surgical correction by the Woodward procedure. J Bone Joint Surg Am. 1981; 63: 1199-207.
10. Sulamaa M, Wallgren GR. Congenital High Scapula (Sprengels Deformity) Acta Orthopaedica 1954; 24(1-4): 195-206.
11. Cavendish ME. Congenital elevation of the scapula. J Bone Joint Surg Br. 1972; 54(3):395-408.
12. Rigault P, Pouliquen JC, Guyonvarch G, Zujovic J. Congenital elevation of the scapula in childhood. Rev Chir Orthop Reparatrice Appar Mot. 1976;62: 5-26.
13. Mallet JF, Bronfen C. Malformations of the shoulder girdle in children and adolescents. EMC-Locomot Syst 2002,7[15-202-A-10].
14. Floemer O, Magerkurth C, Jauckus J, Lutschg JF. Schneider Klippel Feil Syndrome and Sprengel deformity combined with an intraspinal course of the left subclavian artery and a bovine aortic arch variant. AJNR. Am J Neuroradiol. 2008; 29:306-7.
15. Fontecha CG, Navarro Cano EN, Soldaro F, Barber I. Severe Sprengel's deformity associated with Klippel-Feil Syndrome and a complex vascular abnormality that determined the corrective surgery technique. J Pediatr Orthop. B. 2014; 23:589-93.
16. Genim G. Anomalies osseuses constitutionnelles Sauramps Medical, Paris. 2008.
17. Wilson MG, Mikity VG, Shinno NW: Dominant inheritance of Sprengel's deformity. J Pediatr 1971; 79(5):818-21.
18. Hensinger RN, Lang JE, MacEwen GD. Klippel –Feil syndrome: a constellation of associated anomalies. J Bone Sur Am. 1974; 56: 1246-53.
19. Harvey EJ, Bernstein M, Desy NM, Saran N, Ouellet JA. Sprengel deformity: pathogenesis and management. J Am Aca Ortho Surg. 2012; 20; 177-86.
20. Stelzer JW, Flores MA, Mohammad W, Esplin N, Mayl JJ, Wasyliw C: Klippel Feil Syndrome with Sprengel deformity and extensive upper extremity deformity: A case report and literature review. Case Rep Orthop. 2018:579730.
21. Satis S, Alparslan N, Tuna M, Dere O, Yetisgin A. Bilateral multilevel cervical rib and bilateral omovertebra in Klippel –Feil Syndrome. World Neurosurg. 2020; 136:62-65.
22. Andrault G, Salmeron F, Laville JM. Green's Surgical procedure in Sprengel's deformity: Cosmetic and Functional results. Orthop Traumatol Surg Res. 2009; 95: 330-35.
23. Wada A, Nakamura T, Fujii T, Takamura K, Yanagida H, Yamaguchi T et al. Sprengel deformity: Morphometric assessment and surgical treatment by the Modified Green procedure. J Pediatr Orthop. 2014; 34: 55-62.
24. Ashok A, James D, Gahukamble A, Palocaren T, Madhuri V. Modified Woodward's procedure confers functional improvement in Sprengel's deformity. J Pediatr Orthop B. 2020; 30;585-92.
25. Alsiddiky AM, Rafiq Z, Bakarman KA, Alhuzaimi FS, Asif M. A novel modification of Woodward Procedure for correction of Sprengel deformity by application of anchoring sutures. Ind J Orthop. 2021; 55: 189–198. [CrossRef] [PubMed]
26. Mears DC. Partial resection of the scapula and a release of the long head of triceps for the management of Sprengel's deformity. J Pediatr Orthop. 2001;21: 242-45.
27. Masquijo JJ, Bassini O, Goyeneche R, Miscione H. Congenital elevation of the scapula: Surgical treatment with mears technique. J Pediatr. Orthop. 2009; 29: 269-74.
28. Dhir R, Chin K, Lambert S. The congenital undescended scapula syndrome: Sprengel and the cleithrum: A case series and hypothesis. J Shoulder Elbow Surg. 2018; 27:252-59.
29. Konig F. Operations performed in congenital scapular dysplasia. Zentralbl. Chir.1913; 40: 1186.
30. Greitemann B, Rondhuis JJ, Karbowski A. Treatment of congenital elevation of the scapula; 10(2-18) years follow up of 37 cases of Sprengel deformity. Acta Orthop Scand. 1993; 64:365-68.