American Journal of Medical Case Reports, 2021, Vol. 9, No. 12, 702-704
Available online at http://pubs.sciepub.com/ajmcr/9/12/10
Published by Science and Education Publishing
DOI:10.12691/ajmcr-9-12-10

Sprengel Deformity with Cervical Myelopathy a Rare Entity: A Case Report

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Received July 23, 2021; Revised September 02, 2021; Accepted September 12, 2021

Abstract Sprengel's Deformity is a congenital condition characterized by a small and undescended scapula often associated with scapular winging and scapular hypoplasia. Diagnosis is made clinically with a high-riding, medially rotated, triangular-shaped scapula, with associated limitations in shoulder abduction and flexion. Treatment is observation in the absence of shoulder dysfunction. Operative management is indicated in the presence of severe cosmetic concerns or functional deformities (abduction < 110-120 degrees). We herewith present a case of this deformity seen in our center in patient of cervical stenosis with myelopathy.

Keywords: *Sprengel deformity, cervical myelopathy, scapular winging*

Cite This Article: Sharif Al Qadhi, Neeraj Salhotra, Bashar Al Tunbi, C. Livingston, Fahad Al Kheder, Mohammad Hashim, and Kauthar Al Zakwani, "Sprengel Deformity with Cervical Myelopathy a Rare Entity: A Case Report." *American Journal of Medical Case Reports*, vol. 9, no. 12 (2021): 702-704. doi: 10.12691/ajmcr-9-12-10.

1. Introduction

Sprengel's deformity is the congenital failure of descent of the scapula. Eulenburg first described this in 1863. Sprengel described 4 cases in 1891; hence, the anomaly has been called Sprengel's deformity. Other terms for this entity are Sprengel's anomaly, Sprengel's shoulder, congenital high scapula, and undescended scapula. In this entity, varying degrees of scapula elevation and scapula hypoplasia occur. In approximately one third of cases, Sprengel's deformity is accompanied by an accessory ossicle, the omovertebral bone, which articulates between the medial border of the scapula and 1 or more of the cervical vertebrae. In the absence of an omovertebral bone, a fibrous fascial sheath extends from the superior angle of the scapula to the spinous process, the lamina, or the transverse process of 1 or more lower cervical vertebrae. This fixation accounts for the medial position and elevation of the scapula.

We herewith report a case of Sprengel deformity in a adult presented to us with cervical myelopathy

2. Case Report

Our patient was a 54 yr old male with complaints of right sided limb weakness since 6 months. Examination revealed power on right side 3/5 with hyperreflexia and increased tone with positive Hoffman test. Patient underwent a CT and MRI cervical spine. CT revealed a Sprengel deformity and bifid cervical spinous process at C4 and 5 and Sprengel deformity joining the bifid spinous process on right side. MRI revealed gross spondylitiv myelopathy at C3 to 5 level with cord changes with canal stenosis.

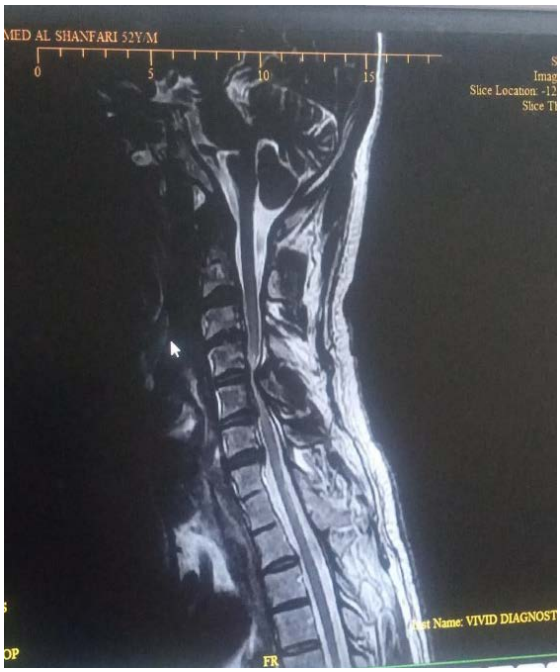

Figure 1. Preoperative MRI showing severe cervical canal stenosis and myelomalcia at C4/5 level

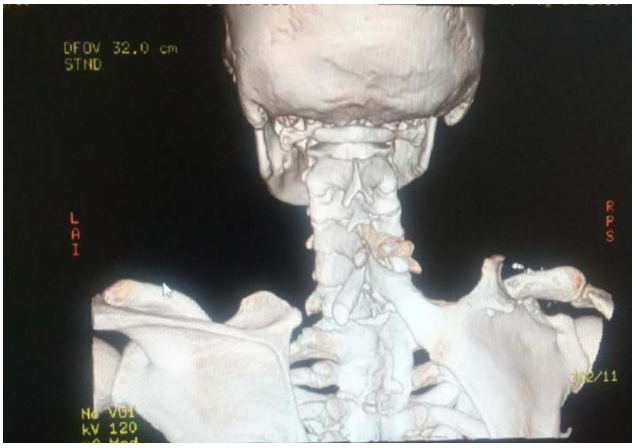

Figure 2. CT reconstruction showing the sprengel's deformity of bony bar connecting the bifid spinous processes of C4/5 region to the scapula

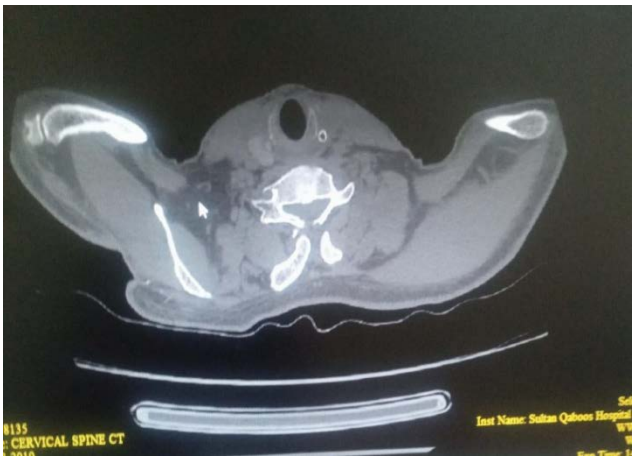

Figure 3. CT spine revealing sprengel's deformity

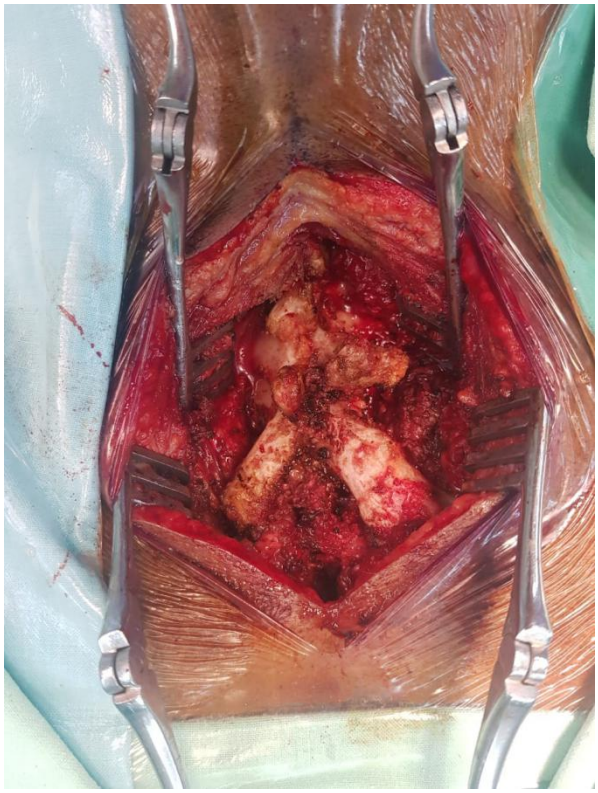

Figure 4. Interaoperative picture revealing the bony bar of sprengel's deformity

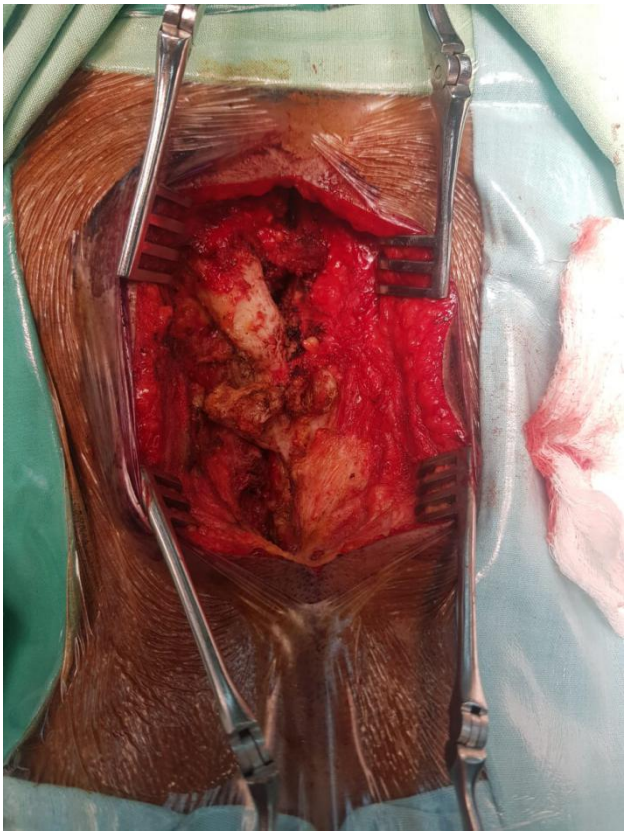

Figure 5. Interaoperative picture of sprengel's deformity after excision of the bony bar

After explaining the family all possible risks and benefits and once family agreed patient underwent posterior decompressive laminectomy of cervical 4 and 5 and excision of the bony bar joining spinous process to scapula. Postoperative period was uneventful. Patient was discharged home after suture removal and is on physiotherapy and on follow up in OPD.

3. Discussion

In 2018 Khan Durrani MY et al discussed the pathophysiology and managment of spengel's deformity in detail. [1] In 2020 Patwardhan et al described surgical correction of Sprengel's deformity by modified Woodward procedure. [2] In 2013 VanAalst et al described sprengle's deformity association with spinal dysraphism. [3] In 2013 Mittal N et al also described the association of sprengel's deformity with tethered cord. [4] In 2001 Chinn DH et al described prenatal diagnosis of sprengel's deformity by ultra sound. [5]

4. Conclusion

Sprengel's deformity is generally seen in children and their association with spinal dysraphism is common. In our patient who neglected this deformity in childhood but presented only once started having cervical spondylitic myelopathic features. Patient underwent standard posterior decompressive cervical laminectomy along with excision of the bony bar connecting the bifid spine to the right sided scapula. Patient is doing well and is on follow up in OPD.

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