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Case Report

Rare case of unilateral hypertrophy of left lamina and spinous process of C6 with associated C3-C4 block vertebra: A case report

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ABSTRACT

We report a rare case of congenital cervical anomaly in a 32 yr old female, who was referred in our department for the complaints of chronic cervical and shoulder pain with a hard cervical mass. Anteroposterior and lateral radiographs of the cervical spine were performed, which show a rib like bony projection, in the left side of neck and on the lateral radiograph, it was corresponding to a hypertrophied C6 spinous process. CT study was done for better delineation of this congenital anomaly, which showed hypertrophied left lamina of C6 and spinous process, crossing midline and towards the left side, extending inferiorly up to the level of thoracic inlet. There was associated fusion of C3 and C4 vertebrae, resulting in block vertebra (Klippel-Feil syndrome Type 1) and unfused spinous process of C6 vertebra. This case report is the first case, where hypertrophied lamina and spinous process of C6 with unfused spinous processes and block vertebra, congenital cervical anomalies were seen in the same patient.

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1. Introduction

Congenital abnormalities of cervical spine are not a rare entity. Relatively common developmental disorders of the cervical spine include persisting apophysis of the transverse process, vertebral platyspondyly, fusion of vertebral bodies at various levels, vertebral hypoplasia etc.¹

Cervical spine abnormalities are associated with a variety of pathologies like Morquio's syndrome, Spondyloepiphyseal dysplasia, Diastrophic dwarfism, Osteogenesis imperfecta, Klippel feil syndrome.² These congenital abnormalities are often seen incidentally on radiologic studies and are rarely associated with clinical symptoms. In this case report, we present a very rare congenital spinal anomaly- marked unilateral hyperplasia of lamina and spinous process of C6 vertebrae associated with schisis defect and block vertebra at the level of C3-C4 level. This is an extremely rare congenital anomaly which has been described only twice in the literature. The rarity of

2. Case Report

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A 32 yr old female was referred for evaluation of a hard cervical mass and chronic cervical pain. On examination, a hard, non mobile mass was localized to the posterior neck without any overlying redness or tenderness. The neck movements were normal without any restriction.

Radiographs of the cervical spine were done, in AP and lateral projections, which revealed an elongated bony protrusion arising from the C6vertebra. Partial fusion of the body of the C3 and C4 vertebrae was seen (Figure 1).

CT scan of the patient was performed to further evaluate the patient which revealed unilateral hypertrophy of the left spinous process of C6 vertebra with unfused posterior process at the same level. Partial fusion of the C3-C4 vertebral bodies was also seen (Figure 2). 3D reformats were also performed (Figure 3).

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the case and its presentation for the first time in the Indian region prompted us to the present report.

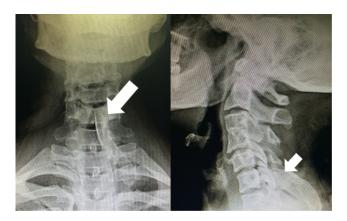


Fig. 1: Radiographs of the cervical spine was done before MRI, in AP and lateral projections, which revealed an elongated spinous process of C6 vertebra, crossing the midline and seen up to the level of left 1^{st} rib

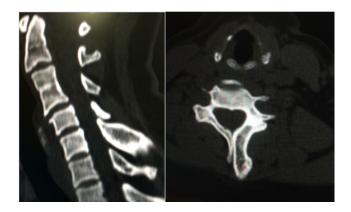


Fig. 2: Block vertebra of C3-C4 is evident on Sagittal sections. Axial CT scans passing through C6 vertebra show the absence of pseudoarticulations at the base of the spinous process and within it. Schisis of the vertebral posterior arch and hypertrophy of the left lamina are also evident

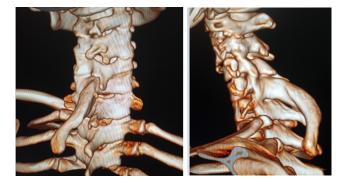


Fig. 3: 3D reconstruction (images of cervical spine) with elongated spinous process of C6 vertebra

Plain MRI was performed to rule out any other intraspinal anomalies, Chiari malformation, diastematomyelia, or other spinal cord malformations, however no such anomalies were detected on MRI. Mild T2W hyperintense signal was seen within the soft tissue surrounding the boney outgrowth. Mild degenerative changes were also seen within the cervical spine (Figures 4 and 5).

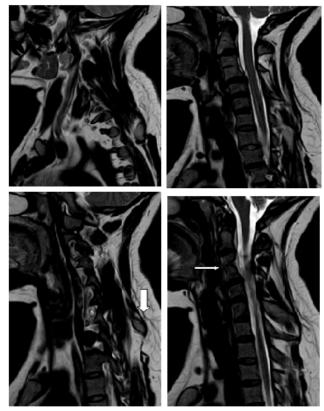


Fig. 4: Sagittal T2W images show a hyperplastic spinous process of C6 vertebra. Sagittal T2 images at the level of hyperplastic spinous process (Thick arrow) show the presence of normal appearing spinal cord, normal intervertebral disc spaces and anterior & posterior epidural spaces. Block vertebra at C3-C4 is also evident (thin arrow)

Whole body imaging was performed upon the patient to look for other anomalies, however no other associated anomaly could be visualized in the patient.

Surgical resection of the bony outgrowth was performed and the resected segment of bone was sent for histopathological evaluation which revealed normal bone tissue.

3. Discussion

Congenital abnormalities of the cervical spine can cause multiple symptoms such as limitation of cervical movement, pain, neurological deficit and aesthetic anomalies.³

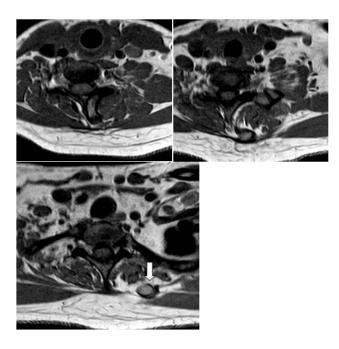


Fig. 5: Axial T1W images show a hyperplastic and elongated spinous process of C6 vertebra and left lamina, extending to the left of the midline and terminating in subcutaneous tissues(arrow)

Diagnostic evaluation of the vertebral anomalies is primarily performed by radiological investigations. Although conventional radiographs depict the anomalies in most of the cases, however differentiation between congenital anomalies and traumatic causes can pose a difficulty at times, hence, in cases of uncertainty, CT scan is indicated. MRI is also indicated in patients with spinal anomalies because of the high incidence of intraspinal pathologies even in absence of clinical signs.^{4,5}

Unilateral hyperplasia of lamina and spinous process of C6 vertebra is not a common entity, only a very few cases have been reported in literature so far. In none of the cases reported so far, existence of vertebral fusion abnormality with elongated spinous process and schisis, has been seen in a single patient. In utero, the development of the vertebral arch begins in the 3rd to 6th gestational week and is completed by the second decade when fusion of the secondary ossification centers occurs.¹ The exact etiology of unilateral hyperplasia of lamina and spinous process is unknown however, abnormal chondrification and ossification of one vertebral arch with normal development of the other arch best explains the anomaly and the finding is usually detected incidentally. The schisis defect observed is the most probably due to the non union of the secondary ossification centers of the spine (which usually completes in the second decade). Schisis is the simplest neural tube defect which is caused due to the failure of the foetus's spine to close properly during the first 3-4 weeks of gestation, which usually completes in the second decade. Schisis in the lower cervical spine is a common finding which is

usually associated with congenital absence of a cervical spine pedicle⁶ and cervical spondylolysis.⁷ Giuseppe et al. described a similar case of marked unilateral hyperplasia of a left posterior cervical arch with associated schisis at the same level.² There is also a case report showing hyperplasia of the C7 spinous process in a 24-year-old women published in the year 2007.⁵

Various developmental disorders of the cervical spine are described in literature, which include Klippel- Feil Syndrome, occipitalisation of atlas, basilar invagination, congenital absence or hypoplasia of pedicle, hemivertebra or butterfly vertebra, but unilateral hypertrophy of spinous process is extremely rare. Klippel- Feil Syndrome (KFS) is a syndrome that is characterized by classical triad decreased cervical motion, short neck and low-set posterior hairline.¹

In our patient due to chronic cervical pain and hard mass, the patient underwent surgery and spinous process was excised.

4. Conclusion

We describe the first case of unilateral giant hyperplasia of posterior cervical arch with schisis at the same level and cervical block vertebrae. This finding may should be taken into account by radiologists, neurologists and orthopaedic surgeons, while dealing with abnormalities of the cervical spine, the possibility of such anomalies should be considered and the treatment planned accordingly. Since congenital hyperplasia of a spinous process does not cause any limitation of cervical spinal movement or neurological deficit, it is therefore imperative to differentiate it from other cervical abnormalities, which need surgical management.

5. Source of Funding

The authors declare that there is no conflict of interest.

6. Conflicts of Interest

None.

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