Case Report

Spinal canal stenosis at the level of atlas

Suchanda Bhattacharjee, Vijayasaradhi Mudumba, Purohit K. Aniruddh

Department of Neurosurgery, Nizam’s Institute of Medical Sciences, Hyderabad, India.
Corresponding author: Dr. Suchanda Bhattacharjee, Department of Neurosurgery, Nizam’s Institute of Medical Sciences, Hyderabad – 500 082, India.
E-mail: suchandab@gmail.com

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Abstract

We report here a rare case of high cervical stenosis at the level of atlas who presented with progressively deteriorating quadriparesis and respiratory distress. A 10-year-old boy presented with above symptoms of one-year duration with a preceding history of trivial trauma prior to onset of such symptoms. Cervical spine MRI revealed a significant stenosis at the level of atlas from the posterior side with a syrinx extending above and below. High-resolution computed tomography of the above level yielded an ill-defined osseous bar compressing the canal at the level of C1 posterior arch, which appeared bifid in the midline. The patient was immediately taken up for surgery in view of his respiratory complaints. The child showed an excellent recovery after excision of the posterior arch of atlas and removal of the compressing osseous structure.

Key words: Atlas, posterior arch, spinal stenosis

INTRODUCTION

Stenosis of the cervical spine at the level of atlas is a rare entity in children. There are scattered case reports in the medical literature but little is mentioned in the text books.[1] They are usually seen in association with other anomalies of the CV junction. However spinal canal stenosis of the atlas in adults is not very uncommon.

We report this case of a 10-year-old boy who presented with progressively deteriorating quadriparesis which was preceded by a trivial injury.

CASE REPORT

A 10-year-old boy presented with complaints of progressively increasing weakness of all four extremities for one year. Weakness was more on the left side then the right. Patient had a spastic circumducting gait on the left side. Complaints of painful and restricted neck movements both on the vertical and horizontal axis were present since one year. Child had wasting of left hand and forearm muscles and occasional symptoms of L’Hermitte’s phenomenon. There were no complaints of sphincter disturbances. The child was having breathing difficulty for the last 20 days. There was a history of trivial trauma three months prior to the onset of complaints. The child fell from a running bullock cart but had no symptoms at the time of fall.

On examination the child was tachypneic with the entire accessory muscles in action. There was wasting of trapezius, sternocleidomastoid, suboccipital group of muscles on the left side along with left forearm and hand muscles wasting. Spastic quadriparesis with patellar and ankle clonus was present. Posterior column sensation was impaired and pain and temperature was lost from C2 to T4 dermatome on the left side. Radiogram of the CV junction did not reveal any abnormality on the neutral or flexion and extension views. High-resolution thin-section computed tomography of the CV junction revealed a fusion defect of atlas posterior arch in the center with a faint bony bar in the canal [Figures 1 and 2]. Magnetic resonance imaging showed a posteriorly compressing atlas stenosis with...
a syringomyelia extending from medulla to C5 vertebra level [Figure 3]. Surgical decompression was done by a posterior approach. Part of occiput up to C3 spinous process was exposed. On dissection we found that there was no fusion defect of C1 arch, rather the posterior arch was incurved inward towards the canal in an odd fashion, which was severely compressing the cord. The incurved part was soft and cartilaginous. The posterior arch was excised in the midline along with the compressing incurved element and immediately the pinched cord was released of compression and started pulsating wonderfully. Dura mater was not opened.

A per-operative flexion – extension radiograph of the CV junction – was taken to look for any instability which was not there and therefore no instrumentation was attempted.

Post-operatively the child did well. There was immediate improvement in his respiratory embarrassment and gradually the spasticity came down. Sutures were removed on the seventh post-operative day and the child was discharged with advice of physiotherapy. The child has improved completely on one-year follow-up.

**DISCUSSION**

A whole range of anomalies occur in the craniovertebral junction which cause spinal stenosis. Dysphasia of the atlas is infrequently encountered. Fissures and defects of atlas arch are very rare but when present they are commoner in posterior arch compared to anterior arch. The incidence of posterior arch defect is 0.69% on a study of 1440 lateral cervical film.[1] Posterior midline cleft is the most common defect of all posterior arch defect comprising more than 90% of all posterior arch defect and an incidence of 3 to 4%.[1]

According to Hinck, the sagittal diameter of spinal canal at the level of C1 ranges between 15 and 20 mm.[2] In our patient the diameter of spinal canal at the level of atlas was only 7 mm on thin cut high-resolution computed tomography. The imageological picture in this case was suggestive of atlas stenosis.

There are sporadic case reports but most of them are in adults.[3-8] Radiologically similar picture has been mentioned by Sharma et al.[9] Only a single case report of a similar fashion was reported in 2000 by Po-Chou Lilang, et al.[6]

Developmental stenosis usually occurs below the C3 vertebral level and rarely above C2.[10-12]

In children the atlantal stenosis may be due to a variety of congenital anomalies ranging from achondroplasia, aplasia, dysplasia or hypoplasia of the atlas arch. Various defects have been mentioned by A. Currarino et al., which is shown in [Table 1].

The rare anomalies range from fissures to defects. The defect of this child do not fit in any of this described categories by Currarino et al. This patient radiologically has the appearance of a bifid arch but on operative view it was rather an incurved.

| Type | Description                                      |
|------|--------------------------------------------------|
| A    | Failure of posterior midline fusion of the two hemiarches |
| B    | Unilateral defect                                 |
| C    | Bilateral defects                                 |
| D    | Absence of the posterior arch, with persistent posterior tubercle |
| E    | Absence of the entire arch, including the tubercle |
oblong-shaped posterior arch where it was continuous with no evidence of any discontinuity. The incurved part was hence not picked up in CT scan and there was an impression of bifid arch. Histopathology of the excised posterior arch did not reveal any abnormal structure other than the osseous components. Compression due to bifid posterior arch has been reported in five children by Devi et al.[13] Goel et al reported on the association of spinal stenosis due to posterior compression in three patients with atlantoaxial dislocation.[14]

They may remain asymptomatic and diagnosis may not be made up to 5–10 years of age as semi-arches may remain unfused normally.[1] On the other hand they may be symptomatically critical as they may cripple patients completely. This child was already having respiratory embarrassment and further delay in treatment could have proved fatal.

The contribution of trauma cannot be ruled out as there was a history of trivial injury but there was no radiological or per-operative evidence for a traumatic origin. Congenital anomaly is the assumed pathogenesis with trauma being the inciting factor. Etiological hypothesis remains to be lack of chondrification or even hereditary factors.[14,15] The inward mobility of the posterior fragment during extension of the cervical spine is well demonstrated in two patients in a study by Sharma et al.[9] This could be the possible reason in our patient in response to the trivial trauma. Compensatory hypertrophy of C2 spinous process may occur due to insertion of ligament and musculature, which would normally have been inserted in the C1 posterior tubercle.[15] Impingement of the cord due to buckling of the posterior arch leads to compressive symptoms.[16,17]

Radiograph remains the first line of investigation but in this case the defect was not very clearly defined. Computed tomography vividly revealed bony pathology at the level of atlas when thin high resolution section was taken. On retrospective review of the scan postoperatively, we could make out the curved shape of the posterior arch compressing the cord. MRI is without any doubt most reliable and rules out any soft tissue pathology. Syrinx of the cord could be correlated with the clinical manifestations in this patient. Surgery is relatively simple and a mere excision of the incurred arch suffices.

CONCLUSIONS

Isolated stenosis of the atlas in children is a rare event and the incidence is not known so far. Reporting of such cases brings awareness of such existing etiologies.

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