Case Report

Unilateral lateral mass hypertrophy: An extremely rare congenital anomaly of atlas

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Abstract

A wide variety of congenital anomalies are observed around the craniovertebral junctional area. However, hypertrophied unilateral lateral mass of atlas in association with chiari-1 malformation leading to myelopathy is extremely uncommon. Herein we report a case of 28-year-old female who presented to us with a high cervical compressive myelopathy. Imaging revealed bony hypertrophy involving right sided C1 lateral mass along with chiari malformation-type 1. She underwent transoral as well as posterior decompression followed by occipito-cervical fusion. The authors discuss their case in light of other such reported cases and present a review of the literature.

Key words: C1 lateral mass, chiari malformation type 1, compressive myelopathy, hypertrophy, unilateral, surgery

INTRODUCTION

A host of congenital anomalies are observed around the craniovertebral junction (CVJ) in general and the atlas in particular. Some of these include various grades of C1 arch hypoplasia, assimilation of atlas, anomalies involving the odontoid process, congenital atlanto axial dislocations, basilar invagination etc.[1-6] In addition to these bony anomalies, various vascular and soft-tissue anomalies are also very frequent in this crucial area harboring the transition of brain into the spinal cord. However, hypertrophied unilateral lateral mass of atlas in association with chiari-1 malformation leading to myelopathy is extremely uncommon. Herein we report a 28-year-old female who presented to us with high cervical compressive myelopathy. Imaging revealed bony hypertrophy involving right sided C1 lateral mass along with chiari malformation-type 1. She underwent transoral as well as posterior decompression followed by occipito-cervical fusion. The authors discuss their case in light of other such reported cases and present a review of the literature.

CASE REPORT

The present case report is about a 28-year-old female patient who presented with posterior cervical pain of 2 years duration along with ascending spastic quadriparesis for 1 year. On examination, she had subtle quadriparesis (power 4/5 on the right upper and lower limbs with 4+/5 power on the left side). Deep tendon reflexes were uniformly exaggerated and plantars were bilaterally up going. There was graded sensory impairment below C4 dermatome (30-50%) and posterior column impairment was noted in upper and lower limbs. She had left sided torticollis and neck movements, particularly rotation, were restricted toward the right side. There were no lower cranial nerve dysfunction and no stigmata pertaining to the CVJ anomalies were found on general examination.

She was investigated with computed tomography (CT) scan and magnetic resonance imaging (MRI) of the CVJ. On CT scan, there was hypertrophy involving the right lateral mass of atlas causing impingement into the bony cervical canal and
consequent narrowing of the transverse canal diameter. The bony overgrowth was similar in density as the rest of the C1 ring. Posterior arch of C1 was hypoplastic and left hemi-arch was absent. There was no evidence of atlanto axial dislocation or basilar invagination [Figure 1a-c]. MRI of this area showed compression of the spinal cord from right lateral aspect with distortion of the cord. Cerebellar tonsils were seen impacted at the foramen magnum with obliteration of the cistern magna. The tonsilar descent was seen up to the level of C1. There was T2 hyperintensity inside the substance of the cord at the level of C2 body [Figure 1d and e].

As the patient had both lateral and posterior compression on the spinal cord, we performed transoral decompression of the hypertrophied lateral mass followed by removal of posterior lip of foramen magnum, excision of hypoplastic posterior arch of atlas and occipito-cervical (C2) fusion with titanium cable. The histopathological examination of the hypertrophied lateral mass revealed the presence of fibro-osseous tissue and no evidence of malignancy was found.

Post-operatively, the patient had relief of the neck pain and reduction in severity of spasticity. Post-operative reconstructed CT of CVJ revealed adequate bony decompression and the posterior construct in situ [Figure 2a-c]. The patient had an uneventful post-operative period and was discharged in stable condition. At the time of writing this report, she had completed 12 months follow-up and she had complete recovery of power in her limbs.

**DISCUSSION**

Myelopathy due to extramedullary compression at the level of CVJ is very common in clinical practice. Most such cases are due to atlanto axial dislocation, basilar invagination, odontoideum, hypertrophied odontoid, chiari malformation, primary atlantal hypoplasia etc.,[1-6]

In this report, we have presented a case of myelopathy due to a rare combination of CVJ anomaly consisting of unilateral lateral mass hypertrophy, deficient atlantal hemiarch and tonsilar ectopia. Such a combination of CVJ anomaly presenting with compressive myelopathy has not probably been described earlier. The hypertrophied lateral mass impinged on the bony cervical canal and led to C1 stenosis in the transverse plane. In addition to this static element, motion of the spinal cord within the narrowed cervical canal during neck movements might also have perpetuated the cord damage in our patient. In addition, our patient had soft-tissue anomaly in the form of tonsilar ectopia and crowded posterior fossa. This anomaly also probably contributed to the myelopathy and impending syringomyelia.

Various congenital anomalies are known to involve the atlas,[3-6] Embryologically, proatlas as well as the first cervical sclerotome contribute to the genesis of the atlas. Although proatlas contributes to the upper part of the posterior arch, remaining portion of C1 is derived from the first cervical sclerotome. The anterior arch ossifies from a separate center and two separate ossification centers give rise to the lateral masses and the posterior arch. Progressive spread of the ossification dorsomedially gives rise to the posterior arch. Although both hemiarches unite in the midline at birth, final ossification is usually delayed until 5-7 years of life.[6] As a result, bifold posterior arch is the most common anomaly involving the atlas (90% of all C1 anomalies, overall incidence 3-4%), whereas the anomalies involving the anterior arch are extremely uncommon.[4] Curranino studied hypoplasias affecting the atlas and classified them into five different types [Table 1].[6] Our patient had Curranino type B hypoplasia (affecting one posterior hemiarch). There are various reports of primary atlantal stenosis and myelopathy due to intact, but hypertrophic posterior atlantal arch, also called primary C1 stenosis (Normal sagittal diameter

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**Figure 1:** Hypoplastic C1 posterior arch with normal atlanto dental interval on flexion (a) and extension (b) sagittal computed tomography images. Axial section through the C1 shows hypertrophy affecting right sided lateral mass with deficient left half of the posterior arch (c). Magnetic resonance (MR) imaging of the craniocervical junction shows ventral compression on the neuraxis with hyperintensity within the cord against the C2 body suggestive of early syrinx formation (d). Axial MR section through the same level shows anterolateral indentation on the spinal cord by the bony hypertrophy (e).

**Figure 2:** Post-operative computed tomography scans of the craniocervical junction shows evidence of anterior transoral decompression with removal of lateral mass hypertrophy on the right side (a-c). A capacious lateral gutter is seen and O-C2 sublaminar cable wiring is seen (c).
of C1 16-22 mm, symptoms appear with a diameter below 10 mm.[7,8] Although the absence of C1 hemiarch in our patient appeared to compensate for the lateral mass hypertrophy by natural posterior decompression, in actuality, there remained fibrous constricting bands in place of the actual arch, which needed to be divided during posterior decompression.

To the best of our knowledge, only 4 other cases of unilateral C1 lateral mass hypertrophy have been described in the literature.[5] All of these patients presented with progressive myelopathy and torticollis. The patients ranged from 21 to 54 years of age and were distributed equally in both sexes. Two of these patients had associated syringomyelia and one patient had bony C1 posterior arch. The authors have also argued that this unilateral atlantal lateral mass hypertrophy could be a defined clinical entity and developmental in origin. Similar to theirs, our patient also presented with myelopathy and torticollis. In addition to C1 lateral mass hypertrophy and syringomyelia, our patient also had associated chiari-I malformation and unilateral absent C1 hemiarch. Hence, the constellation of anomalies in our patient is different than those reported by Goel et al. We also encountered one case report of isolated C1 posterior arch hypertrophy leading to myelopathy.[3] However, there was no lateral mass hypertrophy in that patient. The authors suggested a possibility of prior injury with subsequent periosteal reaction and bony growth as an explanation for the isolated posterior arch hypertrophy. However, we agree with Goel et al. who believe that this rare condition could be developmental in origin. Presence of other anomalies in the CVJ in our case further fostered our assumption as to the etiology of this extremely rare atlantal anomaly. There are reports of compensatory hypertrophy of C1 anterior arch or C2 spinous process in cases of C1 posterior arch hypoplasia.[9] Such findings have been attributed to the increased muscular attachments and attendant increased vascularity. This possibility remained strong in our patient as the C1 lateral mass hypertrophy was on the right side whereas the C1 posterior arch was deficient on the left side. However such compensatory lateral mass hypertrophy has not been described previously.

As far as the treatment is concerned, all four patients reported by Goel et al. were operated from posterior approach only.[5] Although only posterior decompression was done in three patients, direct excision of the hypertrophied lateral mass was done in the remaining patient. The last patient was reported to have improved most among 4 and hence Goel et al. maintained that direct excision of the hypertrophied bone is the ideal treatment in such cases. In our patient, we aimed to excise the hypertrophied bone transorally.

The transoral route was chosen because it allowed us to drill the hypertrophied bone under direct vision. Moreover, presence of the transverse ligament acted as a safeguard for the cord during the procedure. After dealing with the hypertrophy anteriorly, we did posterior decompression. The remaining C1 hemiarch was removed followed by posterior fusion (O-C2 wiring). We do concede that, the bony hypertrophy probably could have been decompressed from a posterior only approach as mentioned by Goel et al.[3] However the transverse ligament would anyway have become incompetent considering the fact that the inner surface of the lateral mass provides attachment to the ligament. Hence, iatrogenic instability was anyway going to occur whether anterior elements were removed or not. Rather removing the anterior arch and odontoid allowed wider working space in order to drill the impinging hypertrophied lateral mass. Moreover, the transverse ligament, lying anterior to the cord, provided a safeguard against retraction and drill bit related injury to the anterolateral aspect of the cord whereas no such protection is available posteriorly.

**CONCLUSION**

Unilateral lateral mass hypertrophy of the atlas is an extremely uncommon developmental anomaly and may cause myelopathy either on its own or in combination with other anomalies like tonsillar ectopia. Decompression of the bony overgrowth impinging the cord is the most effective treatment and can be accomplished either anteriorly or posteriorly. More such cases need to be reported for having further insight into this rare anomaly and formulating a uniform management plan for such patients.

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