

Malformed Posterior Arch of Atlas Vertebrae and its Possible Clinical Relevance

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ABSTRACT

Introduction: congenital anomalies related to malformation or incomplete fusion of the posterior arch of the atlas bone, are quite common. They are usually asymptomatic and are diagnosed incidentally. They may range from partial defects presenting as clefts, to the complete absence of posterior arch.

Case Report: during routine osteology class for 1st MBBS students we came across a peculiar atlas with unfused posterior arch. Right arch was shorter compared to left one. The groove for vertebral artery was also found to be narrow.

Discussion and conclusion: anomalous Atlas, in some cases, may cause cervical myelopathy, which can result in numbness and problems with coordination. Vascular complications may also occur, as in the cases of the vertebral artery. The vertebral artery passes through the posterior arch, so fusion deformities may cause a deviation from the usual path taken by the artery. This may lead to compression of the vessel causing ischemia and maybe even necrosis, to the regions of the upper spinal cord, brainstem, and posterior part of the brain. This may further cause a condition that could be characterized by mental retardation, and/or a very high chance of brain strokes. Hence awareness regarding various malformed atlas is essential to neurosurgeons, radiologists and orthopaedic surgeons while diagnosing or treating vascular deficiencies or cord compression syndromes.

Keywords: Atlas; Cervical vertebra; Bone defect; Cervical defect.

Introduction

Atlas is a first vertebra in the vertebral column, supporting the head (globe), enabling its movements by rendering support through the atlanto-occipital joint. It is called atypical vertebra because of absence of body, instead it has anterior and posterior arch with lateral masses, bearing articular facets. Anterior arch forms 1/5th of the overall circumference of the ring, while 2/5th is formed by posterior arch. Behind each superior articular facets on the superior surface of the posterior arch lies a groove for the lodgement of 3rd part of vertebral artery^{1(g)}. Rarely this groove may be converted into foramen by the arching of delicate bony spiculum from the posterior aspect of the superior articular facet. It also transmits first cervical nerve. In the literature incidence of variations in the posterior arch of atlas accounts to 0.7-3%. Variations in the formation of posterior arch of atlas or any bony spiculum in that area can disturb the course and efficiency of vertebral artery and result in neurovascular deficits².

Case Series

During routine osteology class for 1st MBBS in the anatomy department, JSS Medical College, Mysore, we came across 2 peculiar Atlas vertebrae.

Case 1 - Here Atlas vertebra had incompletely formed (unfused) posterior arch, an extra projection (bony

spiculum) was arising from the posterior end from the transverse process extending medially towards the superior articular process on the left side (Fig-1). This projection is seen limiting the space available for vertebral artery.



Figure 1. Case 1 showing incompletely formed posterior arch and ✦ showing extra projection (bony spiculum) arising from the posterior end of the right transverse process.

Case 2 - Here Atlas vertebra had incompletely formed (unfused) posterior arch. On the left side the incompletely formed posterior arch was short and its posterior end was placed more medially compared to that of right side (Fig-2). Thus, compromising the space available for vertebral artery to arch over it.



Figure 2. Case 2 of unfused atlas vertebra, ★ showing short, inturned posterior arch on the right side of unfused atlas vertebra.

Discussion

Being a part of the axial skeleton, the vertebral column is playing an important role in weight transmission, body posture, and walking gait cycle. Anomalies like hemivertebrae may affect the normal vertebral curvature which may lead to scoliosis. Overt manifestation of vertebral curvature defect depends on the factors like the type of vertebrae involved, range of vertebral defect, site, number, and its mutual relation with structures nearby³. The malformation of the atlas is a relatively rare phenomenon. The wide range of posterior arch defects identified clinically are including midline, bilateral, unilateral, posterior arch absence with persistent tubercle, and complete arch absence. According to the type and extent of defect; its clinical, diagnostic, and therapeutic implications differ^{4,5}. Among two anomalous vertebrae in the present case, one presented with posterior arch absence, which could be due to bilateral failure in the formation of two lateral ossification centres. And another one is showing a defect on one side of the posterior arch, it is suggestive of lack of formation of another side lateral ossification in that vertebra. Currarino classified the defect into 5 types. Although our case 1 fits to Type A, it has one more projection extending from transverse process to superior articular facet. In case 2, incomplete arch is short and more medially placed.

The occipitalsclerotome contributes to the formation of the anterior arch of the atlas, and the posterior arch receives contributions from both occipital and cervical sclerotomes. The 1st cervical vertebrae atlas develops from 3 ossification centres, 1 anterior and 2 lateral. Two laterals active centres extend laterally and posteriorly to fuse with the fellow of the opposite side for a complete posterior arch of the atlas. One anterior ossification centre appears normally, if it fails to meet with lateral ossification centres on either side may lead to an anterior vertebral arch defect in the atlas. Lack of chondrogenesis or ossification failure could be the probable causative factors involved in posterior arch developmental failure in atlas⁶.

Non-invasive radiological investigations are playing a substantial role in the early identification of foetal anomalies/defects. A retrospective cross-sectional study has revealed the anomalous hemivertebra associated with other co-existing anomalies like Genito-urinary, cardiac, and other skeletal defects. Most of the fetuses identified with co-existing anomalies have met with the ultimate consequence of neonatal loss when compared with the incidences of isolated hemivertebrae alone. Prenatal screening plays an important role in the early recognition of subtle congenital defects and its probable plan of medical management⁷. Posterior arch defects of atlas cervical vertebrae may remain mute for a long duration, such presentations are considered as benign variations, often it is an incidental radiological finding⁸. Incidence of posterior arch atlas defects is relatively common when compared with an anterior arch which is much rarer. Absentia of anterior arch atlas ossification may mislead the diagnosis as Type 1 Jefferson fracture; hence it requires a meticulous clinical and diagnostic evaluation to confirm this anomaly because of its resemblance with the fracture of anterior arch⁹. A clinical case of the hypoplastic complete posterior arch of an atlas with spondylolysis was identified on a cervical radiograph was associated with non-traumatic cervical myelopathy causing spinal stenosis and cord compression. Surgical posterior arch decompression has relieved the symptoms effectively¹⁰.

Congenital or traumatic causative factors are involved in craniocervical junction abnormalities, are likely to involve atlas cervical vertebrae. Such defects may impact the nearest structures like blood vessels, nerves, or brain stem. Defective chondrogenesis or ossification, or defective somites causing disparity in sequential metameric rearrangements may lead to congenital malformations affecting the development of the normal pattern of the vertebral arch which may result in anomalies like hemivertebrae¹¹. In a particular group of people, certain genes are identified to be involved in cervical vertebral malformations are including PAX1, DLL3, SLC35A3, WNT3A, TBX6. Despite understanding the mechanism of formation of vertebrae from somites, diversified factors are involved in congenital vertebral malformations, still, interplay between host and environment resulting in such anomalies remains obscure¹².

Conclusion

The atlas vertebral defects are often asymptomatic, and incidental findings, often considered as benign variations. Clinically, patients presented with bilateral cleft along with persistent posterior arch, and absence of posterior arch with persistent posterior tubercle, they need special attention to avoid sports-related injury, and to prevent untoward events attributing to atlanto-occipital

joint functioning. Often vague musculoskeletal or neurological clinical presentations are associated with a range of cervical vertebral defects. They may pose a diagnostic challenge without radiological

aid; hence the extent of vertebral defect is a serious concern in the clinical evaluation of vague complaints, and in handling the case with apt medical/surgical management.

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