

Atlanto-Occipital Fusion with Mid Line Defect of Posterior Arch of Atlas

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Abstract: Congenital anomalies of posterior arch of atlas are very uncommon and usually asymptomatic. The abnormalities at the cranio-vertebral junction are present in approximately 1% of newborn infants, but they may not produce symptoms until adult life. The following are examples of such congenital anomalies: basilar invagination, assimilation of atlas, atlantoaxial dislocation, Arnold-Chiari malformation, and separate dens of the axis. Assimilation of atlas is due to non-segmentation at the junction of the atlas and occipital bone. During anthropometric study of 40 skulls in the Department of Anatomy, one skull was discovered, which exhibited assimilation of the atlas to the occipital bone. Knowledge of atlanto-occipital fusion may be important for forensic experts, radiologists, anaesthesiologist, orthopaedic and neurosurgeons. Skeletal abnormalities at the craniocervical junction may result in sudden unexpected death. It can also result in dysphagia, dysarthria or torticollis because of compression of cranial nerves.

Keywords: Atlanto-occipital fusion, posterior arch, foramen magnum.

1. Introduction

Congenital anomalies of the posterior arch of the atlas, though uncommon, are well documented. The abnormalities at the Cranio-vertebral junction are present in approximately 1% of newborn infants, but they may not produce symptoms until adult life¹. The following are examples of such congenital anomalies: basilar invagination, assimilation of atlas, atlantoaxial dislocation, Arnold-Chiari malformation, and separate dens. There may be partial or complete absence of the posterior arch. Clinical presentation in the patient with posterior arch anomaly is highly variable, and may range from being asymptomatic to quadriparetic.

Cranio-vertebral abnormalities have been recorded for many years in morphological and clinico-radiological studies. The most probable cause of the occipitalization is a congenital disorder. The atlanto-occipital fusion may lead to narrowing of the space for medulla oblongata, spinal cord and vertebral artery. This in turn may lead to many physiological symptoms. In patients with the atlanto-occipital fusion, the clinical findings suggest that the major neurological compression is due to the odontoid projection into the foramen magnum. The signs and symptoms of pyramidal tract, anterior bulbar and cranial nerve involvement may be present². Assimilation of atlas or occipitalization of atlas or atlanto-occipital fusion is one of the common skeletal abnormalities of upper cervical spine. Anomalies in the region of the foramen magnum were of

interest mainly to anatomist³, the fusion of occipital bone with the first cervical vertebra result in compression of first cervical nerve and vertebral artery because first cervical nerve and vertebral artery were on the superior surface of posterior arch of atlas. It should be noted that vertebral artery is an important artery grooving the superior surface of the posterior arch of atlas and the compression of the artery may compromise blood flow to the brain. Similarly, compression of first cervical nerve may cause neurological symptoms. Neurological presentations depend on type of defect of posterior arch of atlas. According to Currarino⁴, congenital anomalies of the posterior arch of the atlas were classified as follows:

Type A- A Failure of posterior midline fusion of the two hemi-arches

Type B Unilateral cleft

Type C Bilateral cleft

Type D Absence of the posterior arch with persistent posterior tubercle

Type E Absence of the entire arch including posterior tubercle

Type A anomaly was seen over the 90% of all posterior arch defects and present in 3% to 4% of individuals. In their study, they estimated that the Type B to E anomalies were seen in 0.69% of all population. It is important to determine the type of the defect of the posterior arch of atlas to understand the clinical significance and to prevent major neurological deficits.⁵ Standard text books of Anatomy do

not high light much on the fusion of occipital bone with the first cervical vertebra; hence the research reports are the only source of information. Defect of posterior arch of atlas is believed to occur due to failure of local chondrogenesis rather than ossification.

2. Materials and Methods

A total of 40 skulls in the Department Of Anatomy, were included in the study. The dried bones were studied in detail. We noted only one skull showing occipitalization of atlas. Appropriate measurements were taken and the specimen was photographed.

3. Observation and Results

Out of 40 adult human skulls studied, we observed atlanto-occipital assimilation (Fig .1) in one skull only amounting to approximately 2.5% .The skulls were randomly selected without considering age and sex. Each skull was carefully observed for variations. The skull showed incomplete fusion of a posterior arch of atlas and occipital bone in the midline. Anterior arch was complete and anterior tubercle was present (Fig.1). The right & left lateral mass and right posterior arch of atlas were completely fused with the occipital bone while right posterior arch of atlas was incompletely fused with the occipital bone. The skull showed foramen on the superior surface of posterior arch of atlas on both side (Fig.2 &3). The skull showed midline defect in posterior arch of atlas, measuring 0.7 cm because of failure of fusion of right and left halves of the posterior arch and absence of posterior tubercle (Fig.1). The left and right posterior arches extend 2.5 cm and 2.0 cm from the inferior articulating surface of left and right side, respectively. The superior articular facets of both sides were completely fused with the occipital condyles. The maximum transverse width of the inferior articulating surface measured 1.75 cm on left and right sides. The transverse processes of atlas were having complete foramen transversarium (Fig. 1). The antero-posterior diameter and transverse diameter of foramen magnum are 3.43 cm and 3.06 cm respectively which are within normal limit as some studies suggest^{6, 7 and 8}. The hypoglossal canals were present on both sides but shifted upward.

4. Discussion

The central regions of all four occipital sclerotomes contribute to the parachordal cartilage, which surrounds the notochord and extends as a flat plate on either side of it by the end of the seventh week, forming the basioccipital component of the occipital bone. The exoccipital components (derived from sclerotomes 3 and 4) chondrify soon afterwards; they border the rostra] half of the foramen magnum, forming the occipital arch, which is developmentally equivalent to the neural arch components of vertebrae.⁶

The embryological reasons for the occipitalization of atlas are due to failure to differentiate the fused caudal and cranial segments of the fourth occipital and first cervical sclerotomes and the lack of segmentation and separation between the loose and dense zones of the first cervical sclerotome.

In patients with the atlantooccipital fusion, the clinical findings suggest that the major neurological compression is due to the adontoid projection into the foramen magnum. The signs and symptoms of pyramidal tract, anterior bulbar and cranial nerve involvement may be present.⁷Spano and Darling⁸ suggest that any morphological and structural alteration of the cervical spine may lead to stenosis or sub-stenosis of the vertebral arterial circulation and hence to brain stem anoxia.

Although atlanto-occipital fusion is a congenital condition, many patients do not develop the symptoms until the second decade of life. Lopez et al.⁹ reported that three patients with atlanto-occipital fusion have had cervical pain and two patients had tonic or clonic convulsions. Iwata et al¹⁰ reported a case of atlanto-occipital fusion with unusual neurological symptoms. Sabuncuoglu H et al.¹¹ reported a case of hypoplasia of the posterior arch of the C1.They said that upper cervical anomalies can be misinterpreted as fractures, luxation, osteolysis or instability. Bose A and S Shrivastava¹² found in their study that there is partial fusion of the incomplete bony ring of the atlas to the base of occipital bone. Radhika Paramesh Mudaliar et al.¹³ showed occipitalization of atlas in 2 skulls out of 200 dry adult human skulls (1%).One of the skulls showed partial fusion (0.5%) while the other showed complete occipitalization(0.5 %). They concluded that knowledge of bony fusion between the cranial base and the first cervical vertebrae is important as such skeletal anomaly may result in sudden unexpected death due to compression of vital structures such as brain stem and vertebral arteries.

Shaikh V G et al. ¹⁴ found atlantooccipital assimilation in one skull out of 82(1.21%) while we found atlantooccipital assimilation in one skull out of 40(3.33 %).Khamanarong K et al ¹⁵ detected occipitalization of the atlas in 2 skulls (0.32%) out of studied 633 skulls. The first case was a male skull (54 years of age at decease), where the atlas was partially fused to the occipital bone. The second case was also a male skull (59 years of age at decease) showing complete fusion of the anterior arch of the atlas.

Congenital defects of posterior arch of atlas are rare and most of them are found incidentally. This anomaly is thought to develop due to a failure of chondrogenesis. These defects were classified into five types, depending on the presence of posterior tubercle. In our study we found type "A" anomaly according to "Currarino classification" of congenital anomalies of the posterior arch of the atlas. Because of the neurological presentation associated with the type of the defect, it is worthy to recognize and classify the exact type.

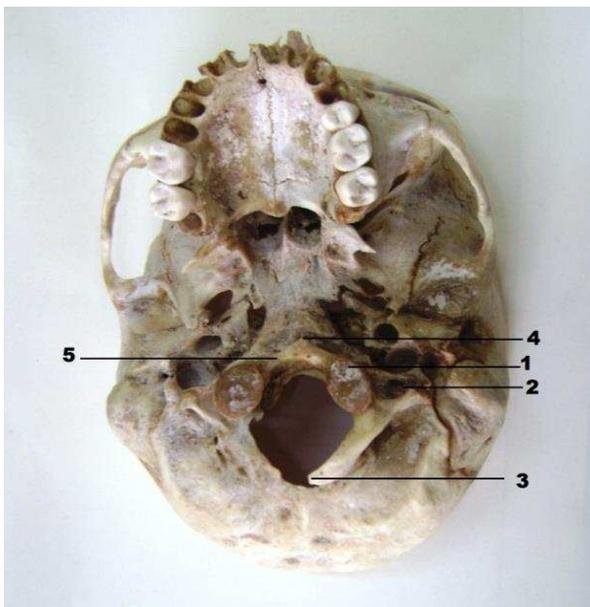


Figure 1: 1-Inferior Articular Facet, 2- Foramen Transversarium, 3-Incomplete Posterior Arch, 4-Anterior Tubercle, 5-Anterior Arch

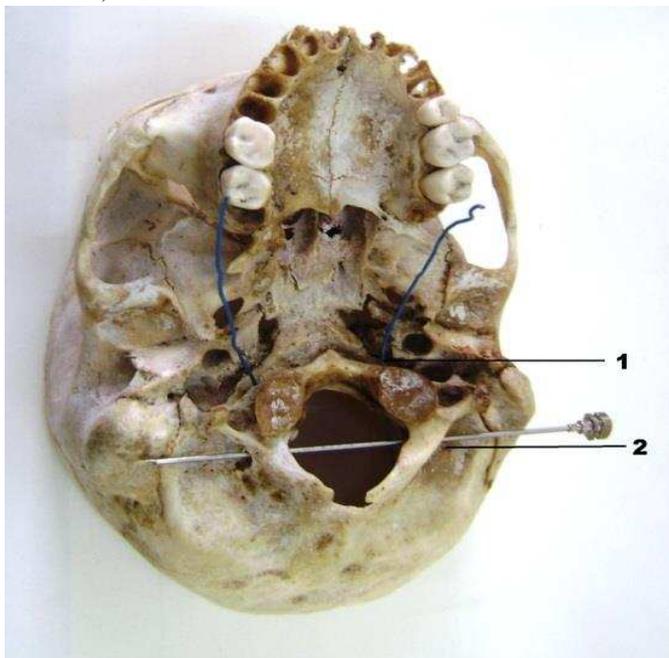


Figure 2: Basal View: 1- The Hypoglossal Canal, 2- Foramen On The Superior Surface Of Posterior Arch Of Atlas

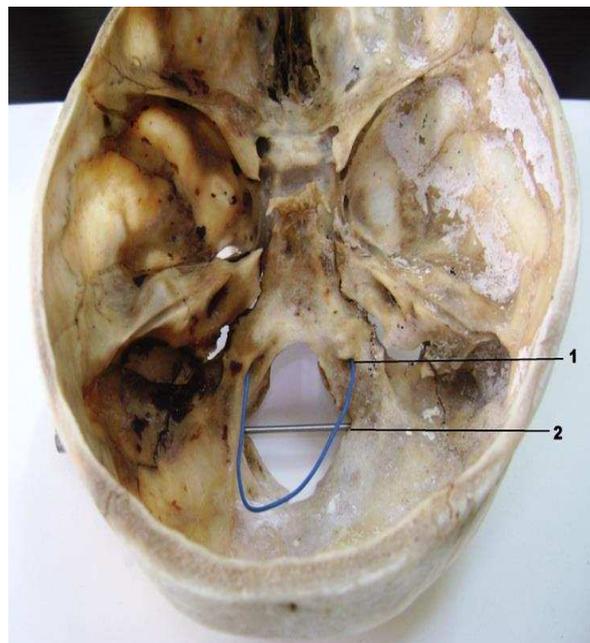


Figure 3: Inferior View: 1- The Hypoglossal Canal, 2- Foramen On The Superior Surface Of Posterior Arch Of Atlas

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