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Short communication

Atlanto-occipital Fusion: Embryological Basis and Clinical Implication

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Abstract: Bony anomalies of cranio-vertebral junction may cause compression of vital structures and neuro-vascular complications. The congenital atlanto-occipital fusion is a rare phenomenon occurring in 0.08 – 3.63% of subjects. It results from inappropriate segmentation of cervical sclerotome during first week of fetal life. Here we report a unique case of symmetric complete atlanto-occipital fusion in an adult male skull, among 50 skulls studied. The knowledge of such anomaly may be of significance to orthopaedists, physiotherapist, neurosurgeons, radiologists and anaesthesiologists dealing with this area.

Keywords: Atlas, Occipitalization, Assimilation, Foramen magnum, Fusion

INTRODUCTION

Anomalous synostosis of the 1st cervical vertebra (atlas) to the basiocciput is termed as atlanto-occipital fusion. It is also referred to as occipito-cervical synostosis, occipitalization and assimilation of atlas. It is characterized by complete or partial fusion of anterior and posterior arches of atlas to the margins of foramen magnum (FM) of occipital bone. The incidence of occipitalization ranges from 0.08% - 3.63% of the population with no gender predilections [1-5]. It may result in restricted neck movement with subsequent adjacent segment syndrome. It can cause a wide variety of neurological and vascular symptoms, which range from intermittent headaches to serious complication like vertebro-basilar insufficiency, spinal cord compression and atlanto-axial instability [1, 5, 6]. The major neurological compression symptoms are due to the projection of odontoid process into the anterior part of FM. However, the onset of symptoms is usually seen in the second decade, with the younger patients commonly asymptomatic. The anomaly often goes unnoticed but incidentally reveals its presence as an anatomical or radiological finding. [7].

In the current study, we present an anatomical variant of occipitalization and discuss it with prevailing literature.

MATERIALS AND METHODS

Fifty adult human skulls from Department of Anatomy, Dr V. R. K. Women's Medical College and Katuri Medical College were studied for evidence of atlanto-occipital fusion. All the skulls were examined carefully for event of occipitalization and any associated anomalies. Among 50 skulls examined, one skull showed complete fusion of atlas with occipital bone. The variant specimen was photographed and the following details were obtained:

1. Antero-posterior diameter of FM: Measured by an imaginary line drawn from inion to basion.
2. Maximum Transverse diameter of FM
3. Antero-posterior diameter of FM behind odontoid process: (Antero-posterior diameter of FM) – (AP distance between anterior tubercle of atlas and the midpoint taken as an imaginary line drawn between the transverse ligament of atlas attachment).
4. FM index: Antero-posterior diameter of FM / Maximum Transverse diameter of FM.
5. Vertical distance between the base of skull and vertebral artery groove on posterior arch of attached atlas (Rt and Lt).
6. Maximum diameters of right and left vertebral foraminae of atlas.

All the linear measurements were taken with a vernier calliper. The extent of fusion between skull base and atlas was studied.

RESULTS

Among 50 skulls examined, only one skull showed atlanto-occipital fusion, the incidence being

2%. The variant specimen belonged to 60-70 years aged male and showed complete fusion of atlas vertebrae with occipital bone. The lateral masses were completely fused with respective occipital condyles. The hypoglossal canal was evident on left side but narrowed on right side. The anterior arch of atlas was fused with anterior margin of the FM, leaving a gap of 0.2 cm in the midline. The posterior arch was fused with the posterior rim of FM in the midline extending to right, leaving a small foramen on left (0.15 cm) and hardly any gap on right (0.05cm), probably for the transmission of vertebral artery and 1st cervical nerve. The transverse processes were incompletely fused with the occipital bone (Fig. 1).

The AP diameter of FM was 2.5 cm, the maximum transverse diameter of FM measured 1.9 cm. The AP diameter of FM behind OP was 1.8 cm. The maximum AP and Transverse diameter of vertebral foramen of atlas was 0.4 and 0.3 cm on left side, 0.7 and 0.5cm on the right side respectively. The vertical distance between vertebral artery groove and skull base on posterior arch of atlas on right and left side were 0.04 cm and 0.11 cm respectively. The FM was oval in shape and was overlapped by inferior articular facets of atlas laterally reducing its transverse diameter. The anterior arch with articular facet for odontoid process was lying parallel to the anterior rim of FM. However the posterior arch of atlas was partially encroaching onto the posterior aspect of FM, reducing its AP diameter (Fig. 1). Due to encroachment of fused atlas, the functional size of FM was reduced.

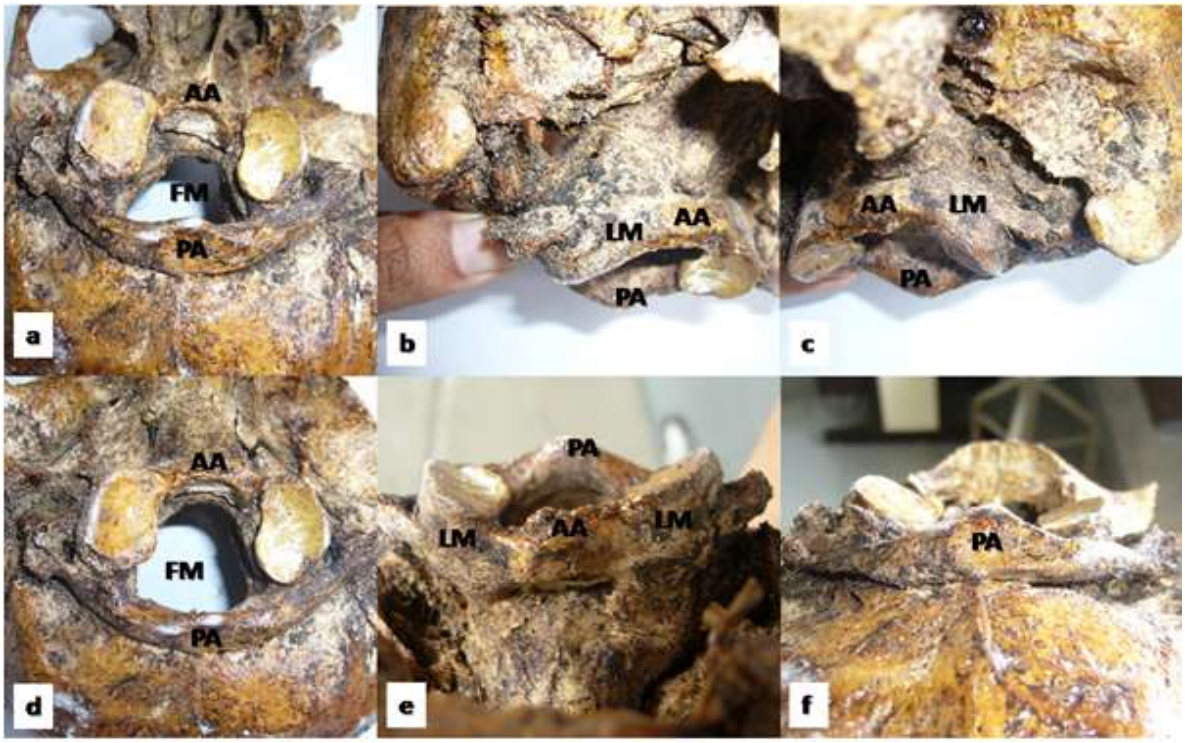

Fig. 1: Showing atlanto-occipital fusion from inferior view (a & d), right lateral view (b), left lateral view (c), anterior view (e) and posterior view (f). The lateral masses (LA) are completely fused with respective occipital condyles. The anterior arch (AA) of atlas is fused with anterior margin of the FM. The posterior arch (PA) is fused with the posterior rim of FM in the midline extending to the sides. The transverse processes are incompletely fused with the occipital bone.

DISCUSSION

Occipitalization is a congenital anomaly, sometimes seen associated with other malformations like, bony torticollis, spina bifida, basilar invaginations, platybasia, Klippel Feil Syndrome and Arnold Chiari I and cervical stenosis [8, 9]. It usually involves the anterior arch and lateral masses, or the entire atlas. It reduces the FM diameter, leading to subsequent neurological complications [7].

During development, the cranial half of 1st cervical sclerotome and caudal half of 4th occipital sclerotome fuse to form basiocciput, while the caudal half of 1st cervical sclerotome and rostral half of 2nd cervical sclerotome combine to form 1st cervical vertebra (atlas) and dens of axis. The inappropriate segmentation of the 1st cervical sclerotome destined to form basiocciput and 1st cervical vertebra (atlas) may result in occipitalization of the atlas [10]. Animal experiments have revealed that the inactivation of HOXD-3 gene expression in rats, results in anterior homeotic transformation at craniovertebral junction [11].

Absolute synarthrosis of atlanto-occipital joint in cases of occipitalization, results in compensatory hypermobility and instability at atlanto-axial joint, and develops concurrent subluxation in advanced age [12]. The increasing degree of ligamentous laxity caused due to repeated flexion and extension of neck, especially the transverse ligament may lead to cord compression or indentation of Medulla Oblongata. Usually, neurological symptoms of pyramidal tract, anterior bulbar and cranial nerve involvement may be present due to hypermobility of OP [10].

However, less frequently the involvement of posterior column may be seen in cases of compression due to posterior arch of atlas [7]. In the present report, the fused posterior arch of atlas was seen evidently overlapping the posterior rim of FM making it more liable to involve of posterior column. Cord compression in extremes of occipitalization, can lead to weakness/ataxia of lower extremities, numbness and pain of upper extremities [5]. The onset of clinical symptoms can be sudden in second decade or may precipitate by relative minor trauma [6].

Apart from SC compression, symptoms referable to vertebral artery compression such as dizziness, seizure, mental deterioration and syncope may also be evident [13]. Authors have suggested that any structural alteration of the cervical spine may lead to stenosis or substenosis of vertebral arterial circulation and brain stem anoxia [5]. The symptoms are progressive but usually intermittent depending on the position of the head [13]. In the current study, due to fusion between basiocciput and atlas, the vertebral artery groove on posterior arch of attached atlas was narrowed to 0.04 cm and 0.11 cm on right and left side respectively, making the vertebral artery more prone to compression.

The sagittal diameter of FM is an important landmark in symptomatic patients, it is accepted abnormal when less than 3 cm [14]. In the present case, we observed a sagittal diameter of 2.5cm, which is abnormal. However, according to Jadhav *et al*., the standard dimensions for FM range between 28-38 mm and 25-40 mm for sagittal and transverse diameters respectively [3]. According to Muthukumar *et al*., a FM index of more than 1.2 will require much more extensive bony resection than otherwise [15]. In the present study we observed a FM index of 1.3.

CONCLUSION

The awareness of the incidence and presentations of occipitalization is essential to surgeons and radiologists dealing with this area. Especially, the neurosurgeons operating on the cervical spine, in order to avoid complications during surgical intervention. Moreover, it is also of importance to anaesthetist performing cisternal puncture and physiotherapist dealing with the neck pains.

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