CASE REPORT

AN UNUSUAL CASE OF UNILATERAL ATLANTO-OCCIPITAL ASSIMILATION WITH SKULL ASYMMETRY

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ABSTRACT

A congenital fusion of the atlas to the base of occiput is defined as assimilation of Atlas. This condition is due to failure in segmentation and separation of the most caudal occipital sclerotome and first cervical sclerotome during the first week of fetal life. This is a case of unilateral fusion of left half of atlas with corresponding occipital bone and atlas is rotated and inclined. Skull showed asymmetries in various skull features on left side like change in size of neurovascular foramina, shifting of position of styloid process, reduction in size of middle cranial fossa along with corresponding petrous bone. Hence knowledge of position of various features of skull, beneficial to the clinicians, surgeons, neurologist and radiologist.

Key words: Assimilation; Atlas; Occipitalization

INTRODUCTION

A congenital fusion of the atlas to the base of occiput is defined as assimilation of Atlas. It is also known as occipitalization of atlas, occipitocervical synostosis and atlanto-occipital fusion. It is one of the most frequent osseous anomalies of Atlas and it is very important in clinical practice as it shows multiple variations and combinations. The atlas vertebra partially or totally fuses with the occipital bone. Complete fusion is more common than the incomplete while multiple variations of partial assimilation have been reported and may involve any aspect of atlanto-occipital articulation. The most probable cause of the occipitalisation is a congenital disorder. This condition is first, described by Rokitansky in 1844 and Schuller in 1911 demonstrated this anomaly on roentgen graphically and the incidence of this anomaly is ranges from 0.08-3% of the general population¹. We present a skull which shows unilateral fusion of left half of atlas with occipital bone along with asymmetries in various features of skull.

CASE REPORT

During the osteology demonstration class for undergraduate medical students, we noticed a skull with fused first cervical vertebra (Atlas). This skull was of adult as socket for upper third molars with synostosis of spheno-occipital synchondrosis, noted.

OBSERVATION AT ATLANTO-OCCIPITAL REGION

The entire left half of the atlas vertebra [anterior arch, articulating facet and posterior arch] was assimilated with the occipital bone. Right half was free from fusion but its anterior arch remained attached to left half of anterior arch and site of union between two parts seen with prominent anterior tubercle and two halves of the posterior arch of the atlas were discontinuous, an interval between them seen [Fig 1YA]. Left Superior articular facet was fused with occipital condyles, while inferior articular facet fused with tubercle (of transverse ligament) and looks like a fused mass protruding into the foramen magnum [FM], large facet of irregular shape noted on it [Fig 1 'a']. Both transverse processes were appearance, normal in but foramen transversarium [FT] was larger on left with thin costotransverse bar, and its tip was fused with jugular process [Fig 1BA] and a gap was created [between jugular process and costo-transverse bar] this probably gave exit for ventral ramus of 1st cervical nerve [Fig 2A RA]. The fused left atlas possessed five openings, three anterior and two posterior. Anteriorly, hypoglossal canal and two very small foramina were located between fused anterior arch and basilar part of occipital bone [Fig 2A]. Hypoglossal canal was separated from jugular foramen by thin plate of bone. Posteriorly, one canal for vertebral artery was located just behind the fused lateral mass and opens in to the FM and it was small in size as compare to FT. Posterior to it, a small foramen was noted, which probably gave exit for dorsal ramus of 1st cervical nerve [Fig.2B]. Anterior part of upper surface of left posterior arch was grooved and being continuous with canal for vertebral artery. Protruding mass on its superior aspect showed a groove, which runs forward from vertebral canal [Fig 3A YA], probably vertebral artery has taken this course to reach anterior. Such foramen for vertebral artery was absent on right side. Sagittal and transverse diameters of FM were 26mm and 27.5mm respectively. Protrusion reduced left half of FM. Irregular shaped tubercle, anterior to left transverse process was noted [Fig 3B WA], it extended_downward for few mm and was separated from occipital condyle by a well marked fossa can be called as fossa for vertebral artery [Fig 1BkA]. Bilateral condylar canals were absent.

Atlas appeared to be rotated to left in a vertical axis and hence anterior tubercle was in line with hamulus of left pterygoid lamina. This rotation shifted articular facet for odontoid process of axis on right [Fig 1 WA]. Unilateral fusion led to an inclination towards the left of 8.3mm. Instead of external occipital crest there was prominent tubercle just 10.3mm behind the FM noted [Fig 3B YA]; this could be due to the abnormal pull on Ligamentum Nuchae.



Fig 1:







Fig 1: Adult skull, shows two YA show fusion of anterior arch and gap in posterior arch, 'a' is irregular facet on inferior articular facet. BA indicates fusion of tip of transverse process and BkA shows the position of fossa for vertebral artery while WA shows the position of facet for odontoid process

Fig 2A: RA indicates gap between Costotransverse bar and jugular process & 3 YA show positions of hypoglossal foramen [thick] and two small foramina medial to it.

 ${\bf Fig}~{\bf 2B}{\rm :}$ Two BkA show position of vertebral canal [thick arrow] and small foramen posterior to it

Fig 3A: YA indicate the protruding mass in FM and groove on it

Fig 3B: WA is the irregular tubercle anterior to left transverse process; two yellow circles show the position of styloid process and RL show the comparative distance between transverse process and mastoid process. YA locates the position of tubercle instead of occipital crest. BkA indicates the left sided occipital bone depressed.

Fig 4: Interior of skull showing small and compromised left half and two BkL indicate comparative length of petrous bone and two BA show the positions of left sigmoid sinus groove and jugular fossa

Note: BA- blue arrow, BkA – black arrow, WA – white arrow, YA- yellow arrow, RA – red arrow, RL- red line, BkL- black line,

Fig 4:

OTHER OBSERVATIONS

Inclination, rotation and unilateral fusion were responsible for various asymmetries in features on left

side. Squamous part of occipital bone was depressed on left side [Fig 3B BkA]. Distance between transverse process and mastoid process was remarkably reduced [Fig 3B RL], the left transverse process was moved away from styloid process [Fig 3B]; this probably disturbed the anatomy of various structures of that region. Further we noted that small sized stylomastoid foramen and short carotid canal on left. On intracranial examination, the grooves for transverse and sigmoid sinuses and jugular fossa were larger on left side [Fig 4 BA]. The size of left middle cranial fossa was smaller along with short and wide Petrous bone [Fig 4 BkL]. All these observations suggest that patient might have left torticollis with comparatively less developed neck muscles and facial muscles, absent atlanto-occipital joint movements and compromised brain size on left side.

DISCUSSION

While going through the literature we have not come across exactly similar description of any two cases of assimilation of atlas, this may be because assimilation may involve any aspect of fusion. Our case shows unilateral atlanto-occipital fusion on left associated with asymmetries in features.

In the development of basilar occiput and atlas, the rostral half of the first cervical sclerotome combines with the caudal half of the last occipital sclerotome to form the base of the skull. While the caudal half of the first cervical sclerotome combines with the rostral half of second cervical sclerotome to form 1st cervical vertebra and odontoid process ². In small number of cases the disruption of this merging process may result in atlanto-occipital assimilation. In our case median vertical division of anterior arch seen, in such cases there has been either no nucleus in the anterior arch and that ossification has taken place by an inward extension from the lateral mass or anterior arch developed from double centers of ossifications 3,4. The standard dimensions for FM range between 28-38mm and 25-40mm for the sagittal and transverse diameters respectively. Spinal cord compressions never occur when the sagittal diameter is 18 mm or more 1,5. We presume that protruding mass in FM and border line reduced diameter might not have caused compression of Medulla Oblongata or spinal cord but reduced diameter might compress vertebral vessels, 1st cervical nerve and may give physiological symptoms like dizziness, seizures and syncope and neurological symptoms. This neurological compression is also due

to the odontoid projection into the FM and repeated flexion and extension of the neck leads to a gradual increasing degree of ligamentous laxity and instability with aging. The first neurological signs and symptoms usually occur after second decade of life 5,6 .

Transverse process is very important landmark for head and neck surgeons, when it is inclined and fused to occipital bone, there may be confusion in reaching various structures and also this led to asymmetry in structure and shape of apertures for the vessels and nerves around the FM ^{7,8}. Such condition is associated with some other skeletal malformations such as basilar invagination, occipital vertebra, spina bifida of atlas, fusion of the axis and third cervical vertebra and atlanto-axial subluxation ^{2,9}, but here we can not rule out the other associated conditions, except occipital vertebra [which is absent here], as no other details are available of the specimen.

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