

Occipitalization of Atlas : A case report

Hussain Saheb S* , Mavishetter G F, Thomas S T, Prasanna L.C, Muralidhar P.

Department of Anatomy, JJM Medical College, Davangere, Karnataka, Pin – 577004.

Abstract:

Atlas is the first cervical vertebra. It does not have a body like other vertebrae. Its lateral masses forms ellipsoidal synovial joints with the condyles of the occipital bone. Rarely, it gets occipitalized, where the lateral masses of the atlas vertebra the condyles of the occipital bone. In this article, a total fusion of the atlas vertebra has been reported. The knowledge of such a fusion may be of importance for radiologists, anesthesiologists, orthopedic and neurosurgeons because skeletal abnormalities at the craniocervical junction may result in sudden unexpected death. It can result in dysphagia, dysarthria or torticollis because of compression of cranial nerves. Head and neck surgeons should be aware that such an anomaly may exist without any typical symptomatic presentation, and thus, serious consequences of upper cervical spinal manipulative therapy may arise when a complete and adequate clinical assessment is missed.

Key words: *Atlas, Basiocciput, Occipital bone, Atlanto-Occipital joint, Occipitalization.*

Introduction:

Craniovertebral abnormalities have been recorded for many years in morphological and clinicoradiological studies. Occipitalization of the atlas or atlanto-occipital fusion is one of the most common osseous anomalies of the craniovertebral junction. According to Yochum and Rowe[1] occipitalization represents the most cephalic ‘blocked’ vertebra encountered in the spine. It is characterized by complete or partial fusion of the bony ring of the atlas to the base of the occipital bone[2]. Hypoplasia of the basiocciput can be found in manifestations of the occipital vertebra, in atlas assimilation and in atlanto-occipital fusion. The patients with craniovertebral joint anomalies exhibit the first neurological signs and symptoms usually no sooner than the second decade [3].

Case report:

During pg dissertation work on 125 adult human skulls of Indian origin collected from Department of Anatomy, JJM Medical College, Davangere, Karnataka, India, one skull showing occipitalization of Atlas was observed the lateral and both anterior and posterior arches were completely fused with the occipital bone. In addition the transverse processes of the atlas were also fused to the occipital bone(Fig-1).

Discussion:

Fusion, either partial or complete, of the atlas with the occipital bone may occur in about 1% of cases so-called atlas assimilation[4]. Neurological symptoms

associated with occipitalization of the atlas are attributed to the laxity of the transverse ligament about the odontoid process caused by repeated flexion and extension of the neck leading to the compression of the spinal cord or actual indentation of the medulla oblongata. The occipital bone is derived from basioccipital, exoccipital and supraoccipital portions, all of which surround the foramen magnum [5]. The basiocciput goes on to develop into four occipital somites. The caudal portion of the fourth occipital somite goes on to fuse with the cranial portion of the first cervical somite to form the proatlas; the proatlas is assimilated into the occiput to form the articular condyles and the tip of the odontoid process. The caudal half of the first cervical somite along with the cranial part of the second cervical somite goes on to form the atlas and the odontoid process of the axis. A paracondylar process represents vestiges of the cranial half of the first cervical sclerotome. This formation is referred to as a caudal shifting where the occipital vertebra separates from the occiput[6].

Although atlanto-occipital fusion is a congenital condition, many patients do not develop the symptoms until the second decade of life. This may be due to a gradual increasing degree of ligamentous laxity and instability with aging.

According to Greenberg[7], spinal cord compression always occurs when the sagittal spinal canal diameter behind the odontoid process is less than or equal to 14 mm.

Table 1: Symptomatology of Atlanto-Occipital fusion.

Sl. No.	Author	Year	Symptoms
1	Kussmaul & Tenner	1859	Convulsions in all 3 reported cases
2	Englander	1934	Cervical pain
3	Hadlay	1948	Neurological Symptoms
4	Keller	1961	Seizure disorder
5	Lopez Zanon et al	1964	Cervical Pain in all 3 reported cases with tonic clonic convulsions in two out of them
6	Budin & Sondheimer	1966	Cervical pain following hyperextension and Onset of cervical pain while putting sweater
7	Albert & Castrischer	1968	Cervical Pain
8	Childers & Wilson	1971	Pain in the neck in some of 25 reported cases; others being either asymptomatic or having insufficient clinical data
9	Vakili et al	1985	Sudden death.

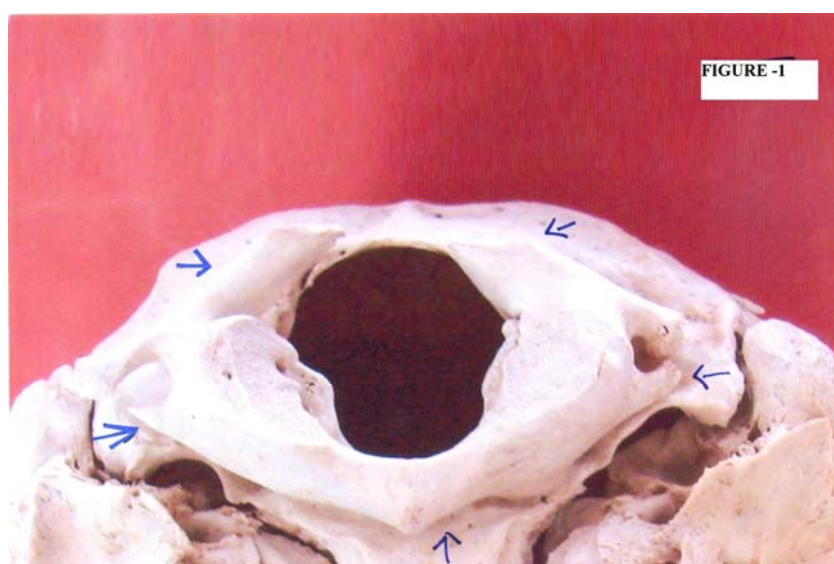


Figure 1: Case showing the lateral masses and both anterior and posterior arches of atlas completely fused with the occipital bone.

Cord compression is possible when the sagittal canal diameter is between 15 and 17 mm and it almost never occurs with a diameter of 18 mm or more.

With aging, the central nervous system may be less tolerant to repeated blows from the odontoid. McRae's & Barnon[8] studied in 25 patients and noticed fusion of C2 and C3 vertebrae in the cases. The patient will be probably asymptomatic if the odontoid process is located below the level of foramen magnum. This

relationship is best assessed through the use of McRae's & Barnon line which is drawn across the foramen magnum. Normally the odontoid process should not project above this line.

Hensinger RN[9] reported that the onset of clinical symptoms can be sudden and precipitated by relatively minor trauma, the most common course is a progressive but sudden onset or instant death. Lopez et al[10] reported that three patients with atlanto-occipital fusion have had cervical

pain and two patients had tonic or clonic convulsions. Iwata et al[11] reported a case of atlanto-occipital fusion with unusual neurological symptoms.

According to Mc Raes & Barnon, patients with occipitalization of the atlas may have the following physical features: low hairline, torticollis, restricted neck movements and / or abnormal short neck. In neurological examination of the atlanto-occipital fusion, patient may reveal the following clinical findings: headache, neck pain, numbness and pain in the limbs, weakness, abnormal head posture, posteriorly located dull aching headache. Cranial nerve findings associated with occipitalization of the atlas include tinnitus, visual disturbances and lower cranial nerve palsies leading to dysphagia and dysarthria. The neurological symptoms and signs of atlanto-occipital fusion can not be distinguished from those of the Arnold Chiari malformation as the pathophysiology of both is essentially the same. Fusion between atlas and occiput occurs anteriorly between the arch and rim of the foramen with some segment of the posterior arch of C1 present in some instances. This fragment can frequently constrict the spinal canal causing intermittent symptoms depending on the position of the head[12].

Conclusion:

Atlanto occipital fusion reduces the foramen magnum dimension leading to neurological complications due to compression of spinal cord.

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References:

- [1] Yochum TR, Rowe LJ. Essentials of skeletal radiology. Volume 1, 2nd ed. Baltimore, William and Wilkins.1987:p3.
- [2] Tun K, Okutan O, Kaptanoglu E, Gok B, Solaroglu I, Beskonakli E. Inverted hypertrophy of occipitalcondyles associated with atlantooccipital fusion and basilar invagination: a case report. *Neuroanatomy*.2004: 3: 43–45.
- [3] Kruff E. Occipital dysplasia in infancy. The early recognition of craniovertebral abnormalities.*Radiology*. 1965: 85: 501–507.
- [4] Bergman RA, Afifi AK, Miyauchi R. Compendium of human anatomical variations. Baltimore: Urban and Schwarzenberg. 1988:197.
- [5] Bopp A, Frauendorf E. Paracondylar process versus paramastoid process - a contribution to its nomenclature and topographic anatomy. *Homo*. 1996: 47: 163–176.
- [6] Black S, Scheuer L. Occipitalization of the atlas with reference to its embryological development. *Int. J. Osteoarch*. 1996: 6: 189–194.
- [7] Greenbery, A.D. Atlanto – axial Dislocation, *Brain*. 1968: 91:655.
- [8] Mc Rae DL, Barnon AS. Occipitalization of atlas. *American Journal of Roentgenology*. 1953: 70: 23 - 45.
- [9] Hensinger RN. Osseous anomalies of the craniovertebral junction. *Spine*. 1986: 11: 323–333.
- [10] Lopez ZA, Limousin LA, Mantz JR. Contribution a la sistematica dc las malformations del atlas a la luz de una neuva malformation inestable. *Rev. Clin. Espanola*. 1964: 94: 208–215.
- [11] Iwata A, Murata M, Nukina N, Kanazawa I. Foramen magnum syndrome caused by atlanto-occipital assimilation. *J. Neurol. Sci*. 1998: 154: 229–231.
- [12] Bailey RW, Sherk HH, Don EJ, Fielding JW, Martin Long D, Uno K, Fening L, Stauffer ES. The cervical spine : occipitalization of Atlas. J.B. Lippincott company. Ptd in U.S.A.1983: p.150.