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Atlanto-Occipital Assimilation with Multiple Accompanying Anomalies

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ABSTRACT

Introduction: fusion of the first cervical vertebra with occipital bone is defined as atlanto-occipital assimilation, which may reduce the dimensions of foramen magnum and leads to consequent neurologic symptoms. Also, assimilation may compress the vertebral arteries and may lead to vertebra-basilar insufficiency.

Case report: a skull with the atlanto-occipital assimilation was noticed during demonstration of the norma basalis at Hacettepe University, Faculty of Medicine, Department of Anatomy. In addition to the fusion, multiple malformations were noted.

Conclusion: the knowledge of the normal anatomy and the malformations of the skull base is essential for both anatomy education and surgeries.

Keywords: Atlanto-occipital assimilation; Atlas, Variation; Skull; Anatomy.

Introduction

The atlanto-occipital joint is located between the superior articular facets of the lateral mass of atlas and the occipital condyles[1]. Stability of the joint is provided with the capsule, ligaments, the atlanto-occipital membrane and the muscles on the posterior aspect of the neck.

Fusion of the first cervical vertebra with occipital bone is an uncommon and non-life threatening congenital malformation which was first described by Rokitansky in 1844 and known as occipitocervical synostosis, atlanto-occipital assimilation or occipitalization of the atlas[1,2]. It is estimated to be about 0.08-3% in the general population and most of the cases remain asymptomatic throughout life[2,3].

Occipitalization of the atlas may lead to the reduced atlanto-occipital joint movements yet it may remain unnoticed due to the flexibility of the cervical column [2]. Absolute immobility of atlantooccipital joint due to occipitalization results in compensatory hypermobility of atlanto-axial joint[4].

Although the occipitocervical fusion is accepted as congenital, rarely it may occur as a result of some pathologic conditions like osteomyelitis, arthritis, syphilis or tuberculosis[3,5]. Clinical symptoms associated with assimilation of the atlas are short neck, restricted neck movements, headache, neck pain and torticollis. Assimilation may compress the vertebral arteries and may lead to vertebra-basilar insufficiency, dizziness and syncope[6].

Atlas arch anomalies can be found coincidentally and most of the anomalies affect the posterior arch[7]. The types of the posterior arch are: Type A defects are defined as failure of the midline fusion of two hemiarches; Type B consists of unilateral clefts, ranging from a small defect to a complete absence of one hemiarch; Type C, bilateral clefts; Type D, the complete absence of the posterior arch with a preserved posterior tubercle; and Type E, the complete absence of both the posterior arch and posterior tubercle[7].

Case Presentation

A skull with the atlanto-occipital assimilation with accompanying malformations was noted during demonstration of norma basalis at laboratory session to undergraduate students.

The dimensions of the foramen magnum were 34 mm anteroposterior and 28 mm transversely. Both of the occipital condyles were completely fused with the lateral masses of atlas. The anterior arch of the atlas was strongly developed with a normal anterior tubercle and an irregular articular facet for the odontoid process was detected. A foramen was noted between the anterior arch and anterior border of the foramen magnum (Figure 1a). The posterior arch was bifid. The transverse processes were bilaterally devoid of costal elements and they were not fused with occipital bone. Transverse foramina were bilaterally underdeveloped (Figure 1b) and additional foramina were detected. The diameters of the channels were 7 mm (right) and 6 mm (left).

In addition some other malformations were noted as follows: left hypoglossal canal with a septum (Figure 2a), bilateral large oval foramen 10 mm on the left and 9 mm on the right (Figure 2b), large 5 mm mastoid foramina on the right (Figure 2b), right jugular foramen divided into three compartments with two septae (Figure 2c) and right styloid process 33 mm in length (Figure 2b).

All measurements were taken with a digital pachymeter from Digimess.

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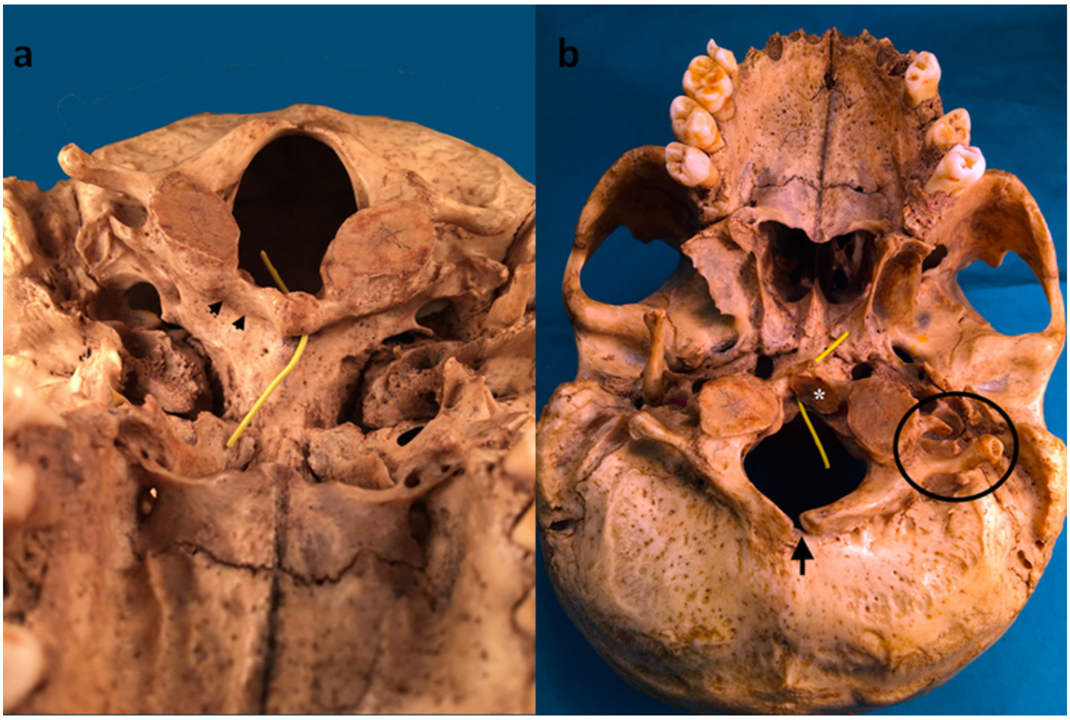

Figure 1. a) Fused anterior arch of the atlas was shown with arrow heads. Foramen between the anterior arch and anterior border of the foramen magnum was shown with yellow string. b) Bifid posterior arch was shown with an arrow and underdeveloped transverse foramina was shown with circle. Irregular articular facet for the odontoid process was shown with an asterix.

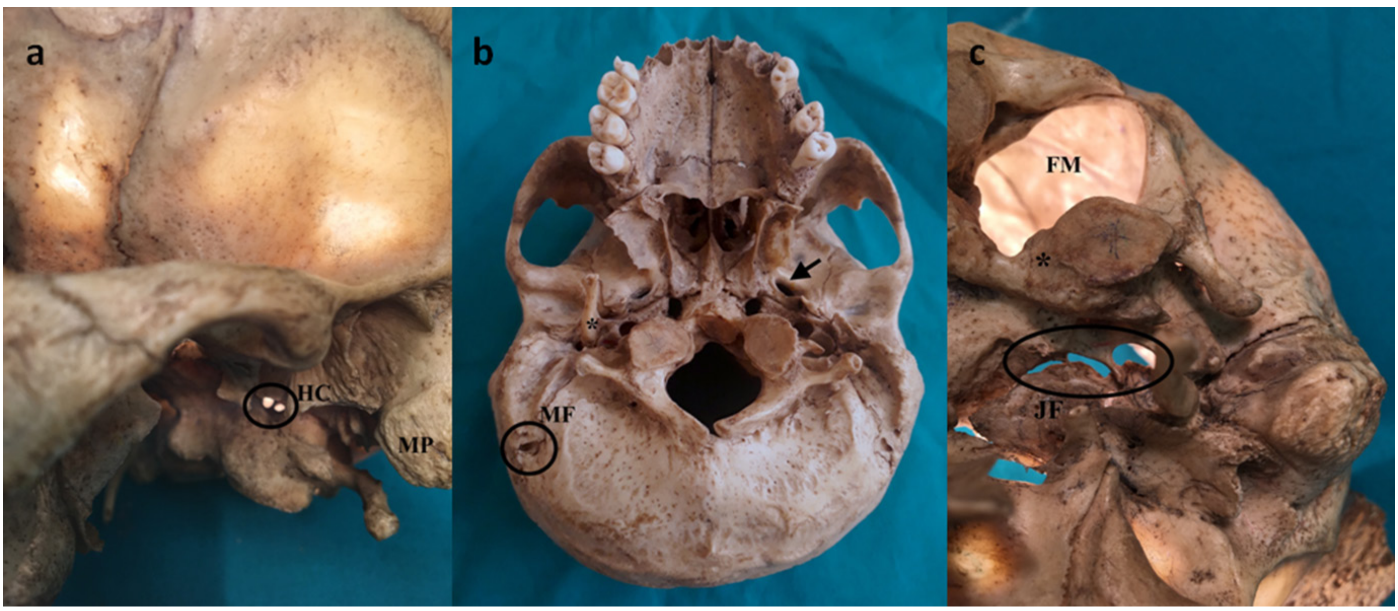

Figure 2. a) HC; hypoglossal canal with a septum, MP; mastoid process b) MF; mastoid foramina, long styloid process was shown with an asterix, large oval foramen was shown with an arrow c) JF; jugular foramen divided into three comportments with two septae, FM; foramen magnum.

Discussion

Atlanto-occipital assimilation was first described by Rokitansky in 1844[1]. There are 5 types of posterior arch defects that mostly effected[7].

Cases of partial and complete assimilation of atlas with the occipital bone were reported several times in the literature. Black (Mac Laughlin) *et al*. reported a case of unilateral fusion of the left occipital condyle to the superior articular facet of atlas[2]. In consequence of left side fusion, a lateral displacement of the atlas to the right side occurs. The anterior and posterior arches were normal and no impairment of the neurovascular structures was noted. Rajani *et al*. also found a case of unilateral assimilation of atlas[8]. The entire left half of the atlas was fused while the right half was free. Both transverse processes were normal and left foramen transversarium was larger. Left posterior arch was grooved and continuous with the canal of vertebral artery[8].

This case report, describes a Type A defect, a failure of complete fusion of the atlas with the occipital bone. There was a cleft on the fully developed posterior arch and there was a foramen between the fully developed anterior arch and anterior border of foramen magnum.

Assimilation can reduce the dimensions of the foramen magnum, leading to neurological symptoms due to the compression of the spinal cord and neurovascular structures[9]. The normal dimensions of the foramen magnum is about 30 mm transversely and 35 mm antero-posteriorly[9]. Aggarwal *et al.* reported a case of assimilation and the transverse and anteroposterior dimensions of the foramen magnum were measured as 30 mm and 32 mm, respectively[9]. Sharma *et al.* reported two cases with assimilation and measured anteroposterior and transvers diameters of the first case as 33 mm and 26 mm, respectively, while the second case's dimensions were noted as 25 mm and 19 mm, respectively[10]. Rajani *et al.* measured the sagittal and transverse diameters of the foramen magnum as 26 mm and 27.5 mm, respectively[8]. Kayhan *et al.* reported two cases with assimilation of atlas and the sagittal and transverse diameters of foramen magnum were 35.7±0.26 mm and 30.32±2.65 mm, respectively for the first case, 35.3±0.26 mm and 26.56±2.65 mm, respectively for the second case[1]. In this study the dimensions of foramen magnum were noted as 34 mm antero-posteriorly and 28 mm transversely.

Besides the assimilation, multiple accompanying malformations were noted as follows; hypoglossal canal bridging, large foramen ovale (which dimension/transverse) 10 mm on the left and 9 mm on the right side, large mastoid foramina and jugular foramen with a septum on the right side.

In the literature, the divisions of the hypoglossal canal by connective or bony tissue were reported with various classifications[11,12]. Eroglu investigated bridging of the hypoglossal canal on 324 ancient skulls belonging to 10 different ancient Anatolian population and classified according to their bridging types[11]. Anomalies of the hypoglossal canal should be kept in mind while researching the unknown etiology of the hypoglossal nerve palsy[12]. In this case complete osseous bridging of hypoglossal canal on the left side was recorded.

One of the important structures of the sphenoid bone, the foramen ovale, transmits the mandibular nerve, accessory meningeal artery, emissary vein and the lesser petrosal nerve[13]. The mean length of the foramen ovale is about 3.85 mm in newborn and 7.2 mm in adults[14]. In the present study the lengths of the foramen ovale were measured as 12 mm on the left side and 12 mm on the right side. Deasi *et al.* studied on 250 sides of the 125 skull and reported the mean lengths of the foramen ovale as 7.98±1.89 mm on the left and 8.14±1.42 mm on the right side[13]. The length of the foramen ovale was measured as 10 mm on the left side and 9 mm on the right side. As the foramen ovale enables access to the mandibular branch of trigeminal nerve, it is important in functional cranial anatomy and neurosurgeries[13].

Kim *et al.* studied the incidence, position and caliber of the mastoid emissary foramina on 106 cadaveric dry skulls[15]. They measured the mean diameter of the foramen as 1.64 mm and the largest specimen was measured as 7 mm. In this case, diameter of the mastoid emissary foramen on the right side was 5 mm. The emissary veins connect the posterior fossa sinuses with extracranial venous system so an increase in the caliber would be the reason of enlarged foramina[15].

Jugular foramen is another important structure of the skull base, transmitting the glossopharyngeal, vagus and accessory nerves and venous blood of the cranial cavity. The foramen varies in shape and size and compartmentation cases were described in the literature[16]. In this case, right jugular foramen was divided into three compartments by two occipital processes.

Styloid process belongs to the temporal bone and provides attachment to muscular and ligamentous structures[17]. Normal length of the process measures between 20-30 mm and any length measured more than this value is considered as elongated and the clinical signs are associated with eagle's syndrome[17]. The symptoms of Eagle's syndrome occur due to the compression of the surrounding trigeminal, facial and glossopharyngeal nerves by the elongated styloid process [17]. In this case only the left styloid process exist and the length of the process was measured 33 mm. According to the literature it is acceptable as elongated.

In conclusion, the knowledge of the normal anatomy and the varieties of the skull base is essential for both anatomy education and surgeries. To date, various types of atlanto-occipital assimilation cases and multiple malformations have been reported. Frequently most of the abnormalities cause no typical symptoms but if the patients have some undiagnosed neurologic symptoms or trauma history, assimilation should be kept in mind for differential diagnosis.

References

1. Kayhan B, İlhan P, Ertürk M, Şengül G. Occipitocervical synostosis: case report. Anatomy. 2015;9(2):104-7. doi: 10.2399/ana.15.017.
2. Black (MacLaughlin) S SL. Occipitalization of the Atlas with Reference to Its Embryological Development International Journal of Osteoarchaeology, . 1996;6:189-94. doi: https://doi.org/10.1002/(SICI)1099-1212(199603)6:2%3C189::AID-OA259%3E3.0.CO;2-D.
3. Khamanarong K. Occipitalization of the atlas: its incidence and clinical implications. Acta Medica Academica. 2013;42(1):41-

5. doi: 10.5644/ama2006-124.69.
4. Jayanthi V. K, R.; Kulkarni, R.N. Atlanto - Occipital Fusion – Report Of Two Cases
. J Anat Soc India 2003;52(1):71-3.
5. Tubbs RS, Salter EG, Oakes WJ. The intracranial entrance of the atlantal segment of the vertebral artery in crania with occipitalization of the atlas. J Neurosurg Spine. 2006;4(4):319-22. Epub 2006/04/20. doi: 10.3171/spi.2006.4.4.319. PubMed PMID: 16619679.
6. Shetty A.S. JR, Sirasanagandla S.R., Nelluri V. A Case Report of Atlanto-occipital Assimilation. Bangladesh Journal of Medical Science. 2018;17(01):169-71.
7. Senoglu M, Safavi-Abbasi S, Theodore N, Bambakidis NC, Crawford NR, Sonntag VK. The frequency and clinical significance of congenital defects of the posterior and anterior arch of the atlas. J Neurosurg Spine. 2007;7(4):399-402. Epub 2007/10/16. doi: 10.3171/SPI-07/10/399. PubMed PMID: 17933313.
8. Rajani SJ SI, Rajani JK. . An unusual case of unilateral atlanto-occipital assimilation with skull asymmetry. . National Journal of Medical Research 2012;2(2):238-40.
9. Aggarwal B, Gorea RK, Gorea A, Gorea A. Occipitalization and its clinical relevance: A case report. International Journal of Ethics, Trauma & Victimology. 2016;2(02). doi: 10.18099/ijetv.v2i02.6860.
10. Sharma DK, Sharma D, Sharma V. Atlantooccipital Fusion: Prevalence and its Developmental and Clinical Correlation. J Clin Diagn Res. 2017;11(6):AC01-AC3. Epub 2017/08/03. doi: 10.7860/JCDR/2017/26183.9999. PubMed PMID: 28764139; PMCID: PMC5535332.
11. Eroglu S. Variations in the form of the hypoglossal canal in ancient Anatolian populations: comparison of two recording methods. Homo. 2010;61(1):33-47. Epub 2010/01/26. doi: 10.1016/j.jchb.2010.01.002. PubMed PMID: 20097339.
12. Kanda T, Kiritoshi T, Osawa M, Toyoda K, Oba H, Kotoku J, Kitajima K, Furui S. The incidence of double hypoglossal canal in Japanese: evaluation with multislice computed tomography. PLoS One. 2015;10(2):e0118317. Epub 2015/02/24. doi: 10.1371/journal.pone.0118317. PubMed PMID: 25706378; PMCID: PMC4338081.
13. Desai S.D SHS, Shepur M.P, Thomas ST, Mavishettar GF, Haseena S. Morphometric analysis of Foramen ovale. SD Desai *et al* /J Pharm Sci & Res Vol4(7). 2012;4(7):1870-1.
14. Somesh MS, Sridevi HB, Prabhu LV, Swamy MS, Krishnamurthy A, Murlimanju BV, Chettiar GK. A morphometric study of foramen ovale. Turk Neurosurg. 2011;21(3):378-83. Epub 2011/08/17. doi: 10.5137/1019-5149.JTN.3927-10.2. PubMed PMID: 21845575.
15. Kim LK, Ahn CS, Fernandes AE. Mastoid emissary vein: anatomy and clinical relevance in plastic & reconstructive surgery. J Plast Reconstr Aesthet Surg. 2014;67(6):775-80. Epub 2014/04/10. doi: 10.1016/j.bjps.2014.03.002. PubMed PMID: 24713148.
16. Kapakin S. An Unusual Anatomic Variation of the Jugular Foramen with Doubled Posterior Condylar Canal. Int J Morphol. 2011;29(4):1186-8. doi: Doi 10.4067/S0717-95022011000400019. PubMed PMID: WOS:000302190500019.
17. Vadgaonkar R, Murlimanju BV, Prabhu LV, Rai R, Pai MM, Tonse M, Jiji PJ. Morphological study of styloid process of the temporal bone and its clinical implications. Anatomy & Cell Biology. 2015;48(3). doi: 10.5115/acb.2015.48.3.195.

Mini Curriculum and Author's Contribution

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