

Two proatlas manifestations (processus condylicus posterior and condylus occipitalis tertius) in a historical male skull – a case report

Sven Schumann, Silke D. Storsberg

Institute of Anatomy, Brandenburg Medical School, D-16816, Neuruppin, Germany

SUMMARY

Anatomical variations of skull base and craniocervical junction are of high clinical interest. Here, we describe the rare case of a combination of an anterior and a posterior proatlas manifestation in a West-European male. Posterolateral to the left occipital condyle we observed an irregular shaped posterior condylar process (processus condylicus posterior). Additionally, a median (or third) occipital condyle (condylus occipitalis tertius) at the anterior circumference of the foramen magnum was present. Clinically, proatlas manifestations may mimic fractures or tumors of the occipital bone, can lead to compression of surrounding structures, and might alter biomechanical properties of the craniocervical junction, potentially leading to neck pain in patients.

Key words: Anatomical variation – Occipital bone – Atlantooccipital joint – Chondrocranium – Occipital vertebra

INTRODUCTION

In the past 200 years, there has been great effort to describe and explain the manifestations of additional osseous structures at the craniocervical junction in man leading to a nomenclature of the numerous variants, which is often confusing and obscure (e.g., precondylar tubercle, precondylar pit, postcondylar tubercle, paramastoid process, hypocondylar arch).

Nevertheless, anatomical variations of the skull base and the craniocervical junction are of high clinical interest, as they may affect both the central nervous system and the locomotor apparatus function. Osseous alterations might result in significant impairment leading to potentially life-threatening conditions when the spinal cord and brain are involved (Muthukumar, 2016).

Craniocervical junction variation may lead to foramen magnum narrowness, and subsequently to compression of the cerebellum, brainstem or vertebral artery. Such constrictions are often associated with Chiari type I malformation,

Corresponding author:

Sven Schumann, Institute of Anatomy, Brandenburg Medical School, Fehrbelliner Straße 38, D-16816, Neuruppin, Germany. Phone: 01726723900. E-mail: sven.schumann@mhb-fontane.de

Submitted: June 10, 2024. Accepted: September 8, 2024

<https://doi.org/10.52083/ROHF5104>

Conradi or Goldenhar syndrome, for instance. Furthermore, osseous alterations of the cranio-cervical junctions often result in restriction of mobility (e.g., Klippel-Feil syndrome), as well as in cranio-cervical junction instability (e.g., Morquio syndrome) (Bams-Mengerink et al., 2006; Bayraktar et al., 2005; Boyles et al., 2006; Kagawa et al., 2006).

CASE REPORT

Here, we describe the coincidence of two proatlases manifestations in the macerated dry skull of a young male from the anatomical teaching collection of the corresponding author. It was handed over to the author by an anatomical dissector, who had received this skull in the 1990s from a private person. At this point in time, the skull had already been processed for medical education (see below). The previous owner had related that the man was of West-European origin and died at an age of around 24. The year of death of the individual, 1884, was inked into the internal surface of the calvaria (right parietal bone). We lack any further clinical information. A calliper was used for measurements.

General aspects

The skull shows typical morphological features of a male skull according to Buikstra and Ubelaker (1994) (nuchal crest, mastoid process, supra-orbital margin, supraorbital ridge/glabella, and mental eminence). The skullcap has been opened with a transversal saw cut about 1 cm above the orbital ridges and about 2 cm above the external occipital protuberance. The skullcap is held in place with two metal pins and can be fixated with two hooks. The mandible is mounted with two metal springs. A bone block was sawn out of the mandibular right retromolar region. The surface shows a spotted brown patina, so the skull was not bleached.

The great sutures of the calvaria (coronal, sagittal and lambdoid suture) are mobile. The wisdom teeth are present but retained in the upper jaw and missing in the lower jaw. The skull is intact except for the missing tip of the right styloid process, fractured teeth 21, 22, 32, and missing teeth 11, 24, 33, and 41, the alveoli are open and do not show signs of osseous modelling, an indication of postmortal loss of teeth.

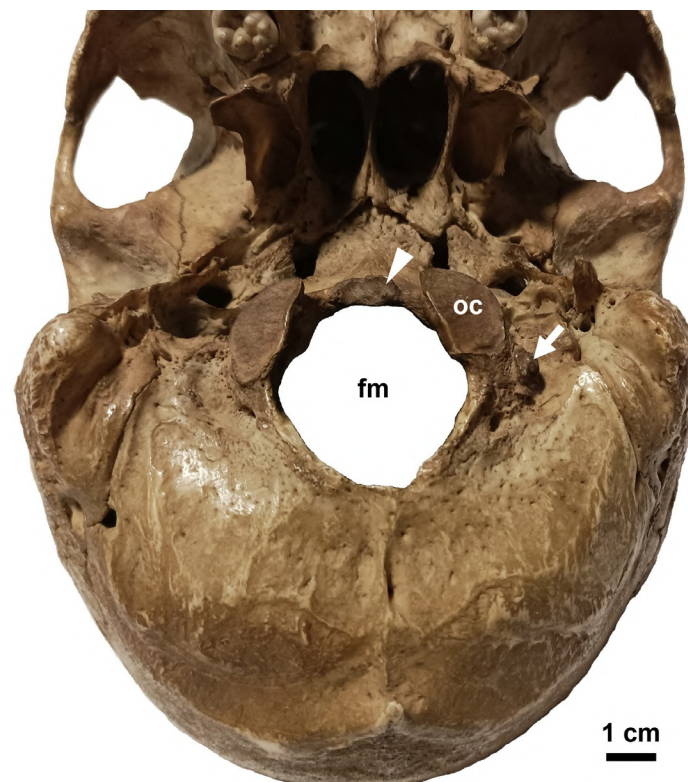


Fig. 1.- External view of cranial base with a processus condylicus posterior (white arrow) next to the left occipital condyle (oc) and a condylus occipitalis tertius (arrowhead) at the anterior circumference of the foramen magnum (fm).

The above information regarding ancestry and age at death is in accordance with the morphology of the skull. Since we did not expect further relevant information for this case report, we did not perform invasive procedures such as isotope or DNA analysis.

Anatomical variation

Posterolateral to the left occipital condyle (oc), we observed an irregular shaped posterior condylar process (processus condylicus posterior) with 0.7 cm in length, 0.4 cm in width and 0.3 cm in height (Fig. 1, marked with an arrow).

Additionally, a median or third occipital condyle (condylus occipitalis tertius) (Fig. 1, marked with an arrowhead) at the anterior circumference of the foramen magnum (fm) was visible. The third occipital condyle was 1.2 cm in width, 0.5 cm in length and 0.2 cm in height.

Besides a septal deviation to the right side, we did not observe further anatomical variations or pathologies of the cranium.

DISCUSSION

The human skull base develops from both the prechordal and the spinal skull base primordium. The axial sclerotomes of the first four somites are incorporated in the spinal part. An increased tendency of assimilation leads to an assimilation of the (pro)atlas, whilst a decreased tendency facilitates the formation of rudimentary occipital vertebrae.

During gestation, the somites are developed and each of the 42 somites differentiates into dermatome, myotome, and sclerotome. The first five sclerotomes further evolve into osseous structures of the skull. The sclerotomes below form the osseous part of the vertebrae. However, the fourth somite plays a pivotal role in the separation of occiput and vertebrae, and in the formation of the craniovertebral junction. Embryological malformation can generate craniocervical junction disorders, and patients with this condition experience neck pain, often combined with headache that starts at the occiput. Movement of the head usually increases these symptoms. Additionally, coughing and/or bending forward trigger the pain

and may lead to spreading into the upper extremities. These symptoms might start after a slight neck injury or for no apparent reason.

The proatlas

During embryological development the fourth sclerotome forms the so called proatlas. This vertebra is consistent in reptiles and lies between the occiput and atlas (Gadow, 1933; Hayek 1924). In humans, this structure further develops inter alia into the occipital condyles, parts of the clivus, parts of the dens axis and vertebral ligaments (Menezes, 2008). In rare cases, the human proatlas or parts of it persist and induce symptoms. These persisting proportions are considered as occipital vertebra and appear as osseous arches, condylar processes, or whole additional condyles. However, aplasia and fusion anomalies are also possible. Accountable genes have been detected as members of the Hox and Pax family (Kessel et al., 1990).

Processus condylicus posterior

The posterior condylar process (processus condylicus posterior) was initially described by Bolk in 1922 (Bolk, 1922) and interpreted as a remnant of the posterior part of the dorsal proatlas arch by Hayek in 1927 (Hayek, 1927). Unfortunately, there are no valid studies about the prevalence of the posterior condylar process, so far (Wolf-Vollenbröcker and Prescher, 2021). It can be associated with a ligamentous structure, the so-called posterior condylar ligament. Free ossicles within this ligament might represent posterior condylar processes unfused with the occipital bone (Wolf-Vollenbröcker and Prescher, 2021).

Condylus tertius

Johann Friedrich Meckel Jr. described the median or third occipital condyle (condylus occipitalis tertius) in 1815 for the first time. It arises from the anterior circumference of the foramen magnum and can articulate with the anterior arch of the atlas or the dens of the axis. The third occipital condyle itself might limit flexion-extension of the atlanto-occipital joints (Rao, 2002). Furthermore, it has been associated with cervical canal stenosis, hypoplasia of the dens, transverse ligament laxity, and atlanto-axial instability.

The prevalence of a third occipital condyle ranges between 0.5% (in a Caucasian population) (Kale et al., 2009) and 2.86% (in an Indian population) (Saralaya et al., 2012). Further investigation is needed, since it is not clear whether high prevalence rates are due to confusion with other variant structures located at the anterior circumference of the foramen magnum (e.g., basilar process). In the present case, the visible articular surface made it easy to distinguish between a third occipital condyle and a basilar process (Kale et al., 2009). The third occipital condyle results from the incomplete regression of the midline part of the hypochordal arch of the embryonic proatlas, with normal regression of the lateral parts. There seems to be no sexual dimorphism in occurrence of this trait (Hauser and De Stefano, 1989).

Our case shows the combined presence of an anterior (third occipital condyle) and posterior (posterior condylar process) proatlas manifestation. Proatlas manifestations may mimic fractures or tumors of the occipital bone. Additionally, they can lead to compression of surrounding neurovascular structures and might alter biomechanical properties of the craniocervical junction. Due to the additional adhesion of the atlas both on the anterior and posterior aspect of the foramen magnum, the combination of a third occipital condyle and a posterior condylar process might lead to impaired mobility of the craniocervical junction.

Taken together, it could be of scientific interest to investigate specimen in anatomical teaching collections. Additionally, specimens of anatomical variations serve as unique learning opportunities for students.

ACKNOWLEDGEMENTS

We thank Prof. Andreas Winkelmann for proofreading of the manuscript.

Ethical approval: The study was conducted according to the guidelines of the Declaration of Helsinki. Since this study was performed on a historical specimen, no further approval by an ethical committee was needed.

REFERENCES

BAMS-MENGERINK AM, MAJOIE CB, DURAN M, WANDERS RJ, VAN HOVE J, SCHEURER CD, BARTH PG, POLL-THE BT (2006) MRI

of the brain and cervical spinal cord in rhizomelic chondrodysplasia punctata. *Neurology*, 66: 798-803.

BAYRAKTAR S, BAYRAKTAR ST, ATAOLU E, AYAZ A, ELEVLİ M (2005) Goldenhar's syndrome associated with multiple congenital abnormalities. *J Trop Pediatr*, 51: 377-379.

BOLK L (1922) Über unvollständig assimilierte letzte Occipitalwirbel beim Menschen. *Anat Anz*, 55: 156-162.

BOYLES AL, ENTERLINE DS, HAMMOCK PH, SIEGEL DG, SLIFER SH, MEHLTRETT L, GILBERT JR, HU-LINCE D, STEPHAN D, BATZDORF U, BENZEL E, ELLENBOGEN R, GREEN BA, KULA R, MENEZES AH, MUELLER D, ORO JJ, ISKANDAR BJ, GEORGE TM, MILHORAT TH, SPEER MC (2006) Phenotypic definition of Chiari type I malformation coupled with high-density SNP genome screen shows significant evidence for linkage to regions on chromosomes 9 and 15. *Am J Med Genet A*, 140: 2776-2785.

BUIKSTRA JE, UBELAKER DH (1994) Standards for data collection from human skeletal remains. Proceedings of a seminar at the Field Museum of Natural History organized by Jonathan Haas. Arkansas Archeological Service, Research Series, 44, Fayetteville/Arkansas.

GADOW FH (1933) The evolution of the vertebral column: a contribution to the study of vertebrate phylogeny. Cambridge University Press, London, pp 93-94.

HAUSER G, DE STEFANO GF (1989) Epigenetic Variants of the Human Skull. E. Schweizerbart'sche Verlagsbuchhandlung (Nägele u. Obermiller), Stuttgart, pp 134-136.

HAYEK H (1924) Über das Schicksal des Proatlas und über die Entwicklung der Kopfelenke bei Reptilien und Vögeln. *Jahrbuch für Morphologie und mikroskopische Anatomie*. 53: 137-163.

HAYEK H (1927) Untersuchungen über Epistropheus, Atlas und Hinterhauptsbein. *Morphologisches Jahrbuch*, 58: 269-347.

KAGAWA M, JINNAI T, MATSUMOTO Y, KAWAI N, KUNISHIO K, TAMIYA T, NAGAO S (2006) Chiari I malformation accompanied by assimilation of the atlas, Klippel-Feil syndrome, and syringomyelia: case report. *Surg Neurol*, 65: 497-502.

KALE A, OZTURK A, AKSO F, GURSES IA, GAYRETLI O, BAYRAKTAR B, ZEYBEK FG, TASKARA N, ARI Z, SAHINOGLU K (2009) Bony variations of the craniovertebral region. *Neurosciences*, 14(3): 296-297.

KESSEL M, BALLING R, GRUSS P (1990) Variations of cervical vertebrae after expression of a Hox-1.1 transgene in mice. *Cell*, 61: 301-308.

MENEZES AH (2008) Craniocervical developmental anatomy and its implications. *Childs Nerv Syst*, 24: 1109-1122.

MUTHUKUMAR N (2016) Proatlas segmentation anomalies: Surgical management of five cases and review of the literature. *J Pediatr Neurosci*, 11(1): 14-19.

PRESCHER A, BRORS D, ADAM G (1996) Anatomic and radiologic appearance of several variants of the craniocervical junction. *Skull Base*, 6(2): 83-94.

RAO PVVP (2002) Median (third) occipital condyle. *Clin Anat*, 15: 148-151.

SARALAYA V, MURLIMANJU B, VADERAV R, TONSE M, PRAMEELE M, JJI P (2012) Occipital condyle morphometry and incidence of condylus tertius: phylogenetic and clinical significance. *Clin Ter*, 163(6): 479-482.

WOLF-VOLLENBROEKER M, PRESCHER A (2021) The ligamentum condylicum posterius as a precursor structure of the Processus condylicus posterior, another Proatlas-Manifestation of the human occipital bone. *J Anat*, 239(3): 611-621.