

diness. A CT study was also done for bony detail. Multiplaner and surface shaded display images were reconstructed from the source axial CT scan images for better appreciation.

OBSERVATIONS

The 1.5T MRI and CT scan revealed the following features – the clivus, which was hypoplastic (Figs. 1, 2, 3, 4). The proatlas developed into a separate vertebra – an occipital vertebra (Figs. 1, 2, 3, 4), which was incomplete and only present anteriorly. It fused posteriorly with the occipital bone (Fig. 1). The hypochordal bow of the proatlas was attached to the right side of atlas, resulting in partial occipitalization of the atlas (Figs. 2, 4, 5) and the failure of fusion of two secondary centres of the posterior arch of the atlas in midline, resulting in atlas posterior midline rachischisis (Fig. 1). This was not a fracture since the margins were smooth. This is consistent with a congenital anomaly.

DISCUSSION

The numerous variants of the CVJ can be understood only if the regional embryonic development of the region is known precisely. The evolutionary processes that result in the formation of both the base of the skull and the cervical spine are of outstanding importance. First, segmented vertebral material is included in the base of the skull to form the «spondylocranium» or «spinal skull segment». Second, material from the vertebral segments is reduced, displaced in a caudal direction, and added to the upper cervical vertebrae. The latter process is especially important for the proatlas, whose corpus forms the tip of the dens axis, called the ossiculum terminale Bergmann. The corpus of the atlas forms the dens axis and the base of the dens. These two processes provide a basis for understanding the variants of the CVJ. If too much material is included in the base of the skull, occipitalization of the atlas will result. If the amount is too small, especially the material coming from the proatlas, manifestation of the occipital vertebra will result (Kollmann, 1905; 1907). The frequency of such manifestations is about 0.1% to 0.5% for minor and 0.0001% to 0.001% for marked cases (Lang et al., 1979).

A mesenchymal strip is also involved, and is important for understanding variants in the

craniocervical junction. In fetuses between 12.5 and 21.0 mm in crown-rump length (Ingelmark, 1947), this strip is located at the ventral boundary of the occipital foramen magnum between the ventral end points of the occipital condyli. This structure is called the hypochordal arch of the proatlas. It recedes in a controlled way during individual development. First, the medial part vanishes so that the arch falls into two parts; these then disappear so that the arch disappears completely. However, if this hypochordal blastema persists, typical manifestations of occipital vertebrae will result.

Out of four occipital sclerotomes the first two form the basiocciput, the third Jugular tubercles and the fourth (Proatlas) form parts of foramen magnum, atlas and axis. Hypoplasia of the basiocciput may be mild or severe, depending on the number of occipital sclerotomes affected. This results in a shortening of the clivus. It is always associated with basilar invagination (Von Torklus and Gehle, 1972; Wackenheim, 1985). There may be a bow-string deformity of the cervicomedullary junction.

Anomalies and malformations of the caudal most part of the occipital sclerotomes are collectively termed “manifestations of occipital vertebrae” (Van Gilder et al., 1987). Four typical manifestations of occipital vertebrae are the basilar process, the condylus tertius, the paracondylar process, and the prebasioccipital arch. The basilar processes are hemisphere-shaped bone projections located at the front rim of the occipital foramen magnum. They occur most often as a continuation of the occipital condyli. They may be unilateral or bilateral or present as isolated and sphere-shaped bone elements. According to Ingelmark, the basilar processes are formed from the hypochordal arch of the proatlas such that the medial part of the hypochordal arch vanishes but both lateral parts persist. In contrast to the processus basilares, the condylus tertius is the medial residue of the hypochordal arch. In this variant, the lateral parts vanish but the medial part persists (Ingelmark, 1947). The condylus tertius, first described by Meckel in 1815, is a bony process in the median line located at the front rim of the occipital foramen magnum. It is always present in reptiles. It may be either a tightly attached or an isolated bone element (Putz, 1975) or it may be articulated with the tip of the dens axis or with the anterior atlantic arch. The paracondylar process (which was once incorrectly called the paramastoid or

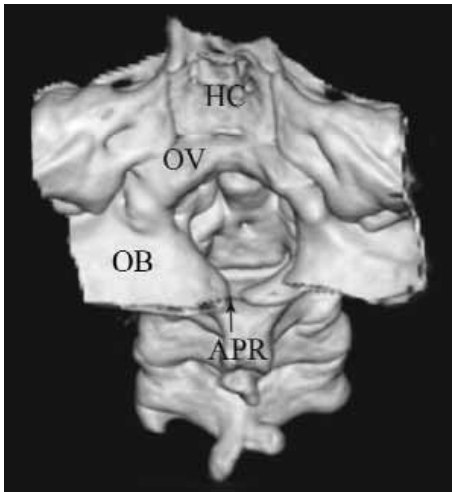


Figure 1. 3D reconstructed CT scan. HC: hypoplastic clivus; OV: occipital vertebra; OB: occipital bone; APR: atlas posterior midline rachischisis.

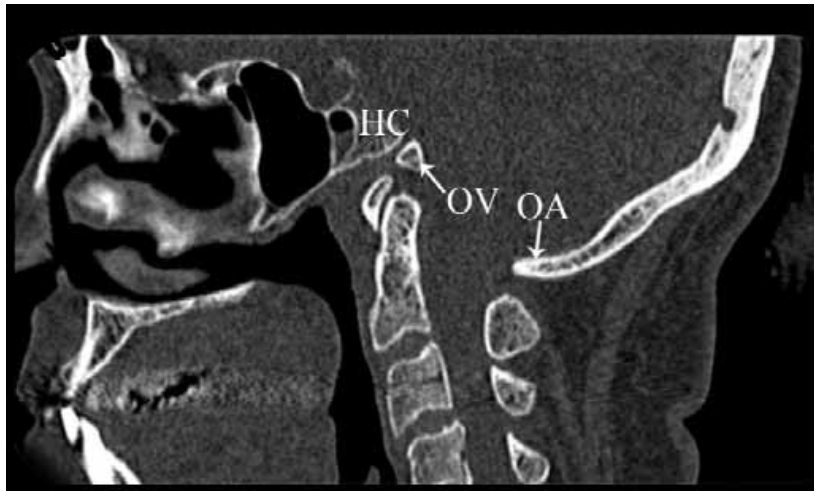


Figure 2. CT scan sagittal reformatted image. HC: hypoplastic clivus; OV: occipital vertebra; OA: occipitalization of atlas (posteriorly).



Figure 3. CT axial section. HC: hypoplastic clivus; OV: occipital vertebra (present only anteriorly).

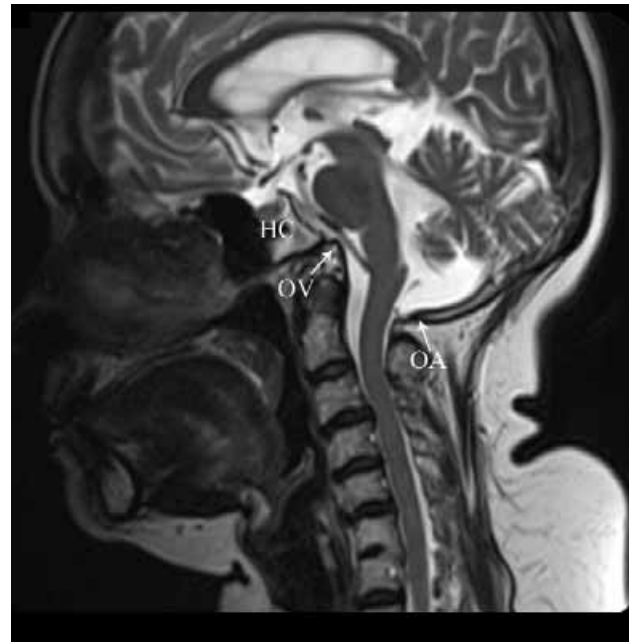


Figure 4. MRI sagittal section. HC: hypoplastic clivus; OV: occipital vertebra (present only anteriorly); OA: occipitalization of atlas (posteriorly).

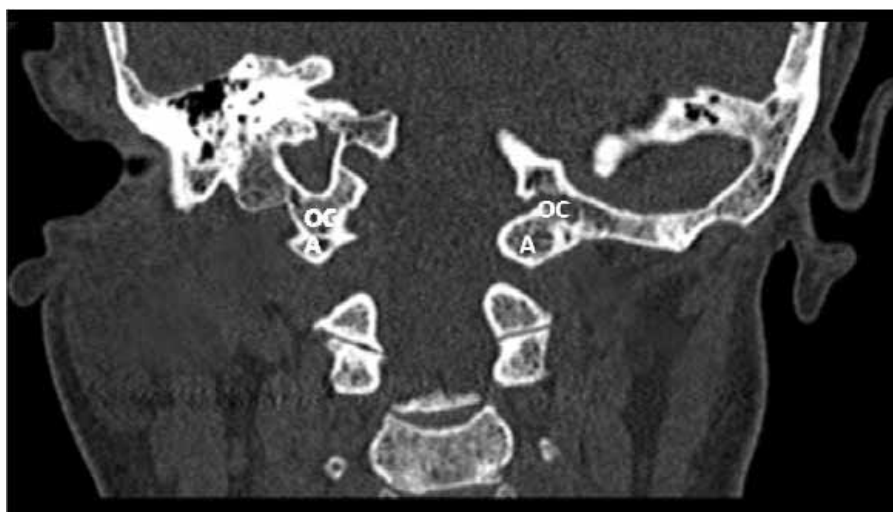


Figure 5. CT coronal section. OC: occipital condyle; A: atlas (showing partial occipitalization of atlas).

paraoccipital process) is a wide-based, bony cone that stems from the exo-occipitale beside the occipital condylus. It is directed toward the transverse atlantic process and may be connected with it by a joint. The paracondylar process develops if the material of the processus transverses of the proatlas is not integrated into the lateral parts of the foramen occipital magnum. Complete persistence results in the massa paracondylica, while minor degrees of persistence can form the paracondylar process or, by dislocation and assimilation to the processus transverses atlantis, an epitransverse process. The prebasiooccipital arch is a bony, bulging arch at the front rim of the occipital foramen magnum. In extremely rare cases, it can be isolated and mounted on the dens axis in a manner resembling a Napoleon hat while articulating with the anterior atlantic arch and the front rim of the occipital foramen magnum. The prebasiooccipital arch develops if the whole hypochordal arch of the proatlas persists and ossifies. The prebasiooccipital arch is in a sense the combination of bilateral processus basillares and a condylus tertius. The highest degree of persistence will be reached if the prebasiooccipital arch is established as an isolated bony arch. The osseous assimilation to the anterior margin of the foramen occipital magnum points to a regressive tendency. According to this, the present patient has a prebasiooccipital arch-type occipital vertebra (Figs. 1, 3).

Thus, when the hypochordal bow of the fourth occipital sclerotome (proatlas) persists or when the proatlas fails to integrate, an ossified remnant may be present at the distal end of the clivus, resulting in typical manifestations of occipital vertebrae (Van Gilder et al., 1987; Von Torklus and Gehle, 1972; Menezes and Van Gilder, 1990). Although typically single, multiple supranumerary ossicles may be present (Wackenheimer, 1985). They may form a joint or pseudojoint with the odontoid process or the anterior arch of atlas and may lead to limitation in the range of motion of the CVJ (Von Torklus and Gehle, 1972). There is an increased prevalence of os odontoides associated with this abnormality (Van Gilder et al., 1987).

The failure of segmentation between the skull and first cervical vertebra results in assimilation of the atlas. According to Yochum and Rowe (1987) 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 occipital bone (Tun et al., 2004). Patients with CVJ anomalies exhibit the first neurological signs and symptoms usually no sooner than the second decade of life (Kruffy, 1965). The assimilation may be complete or partial, and it invariably results in basilar invagination. There is an increased prevalence of associated fusion of the axis and third cervical vertebra in association with atlantooccipital assimilation (Mc Rae and Barnum, 1953). In patients with atlantooccipital fusion, the clinical findings suggest that the major neurological compression is due to odontoid projection into the foramen magnum. The signs and symptoms of pyramidal tract, anterior bulbar and cranial nerve involvement may be present (Hensinger, 1986). In some instances, atlantooccipital assimilation may be associated with sudden death (Vakili et al., 1985; Hensinger, 1986).

The usual reported incidence of posterior arch defects of C1 is 3% (Wysocki, 2003). They are mostly incidental findings, as in this case, and their clinical significance is not very clear. Normally, three ossification centres for the atlas appear during the embryonic period. The two centres for the lateral masses normally unite posteriorly by perichondral growth, giving rise to the posterior arch at 3-5 years of age. The anterior centre for the anterior tubercle and anterior arch usually unites with the two lateral centres at 5-9 years of age. An anatomical classification of the defects of the posterior atlas was proposed by Currarino et al. (1994) and modified by Von Torklus and Gehle (1975):

Type A: Failure of the posterior midline fusion of the two hemiarches.

Type B: Unilateral cleft.

Type C: Bilateral clefts.

Type D: Total absence of the posterior arch with a persistent posterior tubercle.

Type E: Total absence of the posterior arch with a missing posterior tubercle.

According to this classification, the present patient has a type A abnormality (Fig. 1).

Although the absence of the posterior arch of the atlas, when isolated, has been stated to be asymptomatic (Dalinka et al., 1972), reports of associated anterior atlantoaxial subluxation have been made (Schulze and Buurman, 1980). Bilateral atlantoaxial offset has been reported in association with both total and partial aplasias, simulating the Jefferson fracture (Gehweiler et al., 1983). In contrast to the aplasias and hypoplasias, clefts of the atlas

arches are much more commonly observed. Posterior rachischisis, the most common, is observed in 4% of adult autopsy specimens (Gehweiler et al., 1983). The vast majority of posterior atlas clefts (97%) are midline, whereas lateral clefts through the sulcus of the vertebral artery, account for the remaining 3%.

In patients with the myriad anomalies at CVJ, the natural history is not understood clearly; many patients are asymptomatic and probably remain so all their lives. Clinical manifestations develop only with advanced age or after a traumatic event. Symptoms of clinical importance (Godlewski and Dry, 1963; Godlewski, 1972; Von Torklus and Gehle, 1975; Mc Rae, 1953; Unterharnscheidt, 1992) are caused by four principal mechanisms: 1. mechanical compression of nervous structures 2. mechanical compression of vascular structures, especially of the vertebral arteries 3. marked instability or, in contrast, elevated rigidity 4. disturbance of the mechanical mobility of skull joints, for example, the three-legged head bearing with condylus tertius or paracondylar process. Patients seeking medical advice often complain of pain in the neck region or at the back of the head, or they suffer from vertigo or disturbances in balance. Several medical disciplines must be included in treatment, such as orthopedics, neurology, ENT, radiology, neurosurgery, and skull base surgery (Hirschmann, 1978). Mistaken diagnoses have led to delayed treatment and at times adverse results. A review of the anatomy and embryology of this region is essential to the understanding of the pathophysiology encountered.

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