PROCESSUS OCCIPITO-VERTEBRALIS: A RARE SUBOCCIPITAL ABNORMALITY WITH DIFFERENT CLINICAL SIGNS*

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The paper presents three cases of similar pediatric bony abnormality at the craniovertebral junction accompanied by torticollis and ischemic brain attacks. Two patients were operated on, and the outcomes are analyzed. Type of Publication: case series study. Level of Evidence – IV.

Key Words: torticollis, osteal torticollis, proatlas, atlantooccipital bone, cervical spine abnormalities, surgical treatment, neck, cranio-vertebral abnormalities.

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Due to the peculiar anatomy of the suboccipital region, which provides functional support of the head, allows a great deal of rotation and protects elements of the central nervous system and vessels, the craniovertebral junction is considered as the most unique part of the human skeleton.

The unusual structure of the Oc–C_1–C_2 complex is confirmed by the features of its embryologic development: the complex is formed from 6 sclerotomes that further differentiate into 14–16 ossification centers and after functional organization and sclerotomal resegmentation give rise to the cervical vertebrae [3]. It is no coincidence that this extremely complex and unstable process is accompanied by occurrence of multiple malformations, most of which, however, are asymptomatic or develop with minimal complaints. This may explain the fact that the majority of publications devoted to craniovertebral dysplasia is limited to the description of the anatomical features during their radiologic examination. The clinical manifestations of the disease generally refer to by a collective term "vertebrobasilar insufficiency" and are usually accompanied by signs of circulatory disorders in the basin of the vertebral arteries (so-called vertebral artery syndrome) or neurological disorders at dislocations of Oc–C_1 and C_1–C_2 segments. Orthopedic pathology, which is the leading sign of craniovertebral dysplasia, occurs quite rare, except for severe abnormalities.

In different years, we have observed three patients with the same type of craniovertebral abnormality who had similar radiological features and different clinical signs. We have not found publications devoted to this pathology. We gradually accumulated experience and technical facilities for the surgical treatment of neck diseases in children; hence we treated these patients in different ways. Our retrospective experience may be of interest to specialists.

Case description 1 (2007). A boy aged 11 was considered healthy. During rapid growth, the mother noted the appearance and progression of torticollis and facial asymmetry. At residence, he was diagnosed with "chronic subluxation of C_1". Cervical halter traction, correction with a Philadelphia cervical collar, and physiotherapy gave no success. The patient was examined 1 year after the onset of the disease (Fig. 1a). 3D-CT imaging revealed a bony process arising from the occipital bone on the left that formed nearthrosis with posterior arch of C_1 (Fig. 1b). Functional selective angiography in head rotation to the right revealed changes regarded as the compression of the right vertebral artery at the level of C_1 (Fig. 1c).

Supposing that the oblique position of the atlas decreases the Oc–C_1 distance on the right at head rotation and causes compression of *a. vertebralis dex.* and since we considered risky an intervention in the region of the left vertebral artery loop, we agreed on balancing posterior fusion Oc–C_1. Before the operation we visually corrected deformity of the cervical spine during 14 days by halo traction with asymmetric traction (load right 6 kg, load left 2 kg). Without removing traction, posterior occipitospondylodesis Oc–C_1 was made with a fragment of iliac crest autograft and wiring.

The postoperative period was without complications, the child achieved verticalization with a Philadelphia collar. Control functional angiography 2 weeks after surgery revealed no signs of compression of *a. vertebralis dex.* Examination after 1 year: torticollis removed, facial asymmetry preserved (Fig. 1d). The bone block is formed.