Eur Spine J
DOI 10.1007/s00586-016-4389-4

CASE REPORT

A case study of occipital outgrowth: a rare suboccipital abnormality

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Received: 29 September 2015 / Revised: 11 January 2016 / Accepted: 15 January 2016

Abstract
Purpose To describe the clinical and radiological characteristics of uncommon upper cervical spine abnormality in children.
Methods Clinical and diagnostic characteristics of three patients aged 6–12 years with a similar uncommon type of occipital anomaly are described. The patients were admitted in 2007, 2009, and 2014, respectively.
Results All patients were clinically and radiologically examined. In each case the massive, additional unilateral outgrowth of the occipital bone (os occipitale) was visualized. The signs and symptoms included torticollis, acute brain ischemia, and limited head motion. Two of the three patients underwent surgical treatment: an occipital–cervical fusion was performed in the first patient, and the outgrowth was removed in the second patient. After 1 year of follow-up the results were estimated as good for both patients, with better functional outcome for the second patient. The parents of the third patient did not consent for the surgical treatment.
Conclusions The unique features of this abnormality distinguish it from previous descriptions of the manifestation of pro-atlas, atlas, or atlanto–occipital synostosis. The presented abnormality had different manifestation of various severity in each case, from torticollis to acute vascular disorder.
Study design Clinical case series.
Level of evidence IV.

Keywords Bone torticollis · Occipital–atlas bone · Upper cervical spine abnormalities · Surgical treatment · Cervical spine · Suboccipital abnormality · Atlas manifestation

Introduction

The anatomy of the craniocervical junction is unique due to the specific division of the human skeleton. The function of the craniocervical junction is to provide support to the head and to mobilize and protect the central nervous system and vascular elements [1].

The occipito-atlanto-axial structure (C0–C1–C2) is formed from six sclerotomes, which in turn are developed from 11 to 14 foci of organogenesis [2]. This extremely complex and highly vulnerable embryological process is accompanied by numerous malformations, in most cases asymptomatic or of minimal clinical importance [3, 4]. This may explain why the majority of scientific papers on cranial–vertebral dysplasia are limited to the radiological description of anatomical features. The signs and symptoms in most symptomatic cases include vertebral artery insufficiency or neurological deficit resulting form atlanto-occipital or atlanto-axial displacement [5–7].

We observed three patients with an uncommon suboccipital abnormality, which had different clinical manifestations despite the similar radiological findings. No previous descriptions of this particular abnormality were

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Published online: 28 January 2016

found in the literature. The management of these patients granted us remarkable clinical experience, and we believe that our case study would be valuable for the future observations.

Materials and methods

Functional 3D computer tomography (3D CT) with angiography and functional selected angiography were the best methods to evaluate the locus and cause of compression of a. vertebralis. The procedure was performed in the presence of a spine surgeon who controlled the position of the patient depending on clinical signs and contributed to the interpretation of the findings. 3D CT was also used in the pre-operative planning. The same functional approach was also utilized postoperatively.

Case report 1 (primary examination in 2007)

An 11-year-old boy without the previous history of disease was admitted to the out-patient clinic with torticollis and facial asymmetry. Initially the C1 subluxation was diagnosed and traction, physiotherapy, and a support collar were recommended.

After 1 year of follow-up no clinical improvement was observed (Fig. 1a). In a 3D-CT scan, an additional ossicle between the left part of the occipital bone and the posterior arch of C1 causing an oblique skull position was clearly visible (Fig. 1b). Selective angiography with head turned to the right showed the signs of right vertebral artery compression (Fig. 1c).

The oblique C1 position was thought to compress the vertebral artery on the contralateral side with the full occlusion during head-turning test. The risk of surgical removal in the vicinity of the left vertebral artery was considered very high. Therefore occipital–cervical posterior fusion with autograft and wiring preceded by an asymmetric HALO-traction was performed, which resulted in a symmetric skull position for C0–C1. Short- and long-term (1 and 2 years) follow-up visits showed satisfactory skull positioning and solid fusion. The patient was asymptomatic with clinically insignificant limitation of head movement.

After the retrospective analysis of the angiograms, we acknowledge that it was the pseudo-compression of the vertebral artery (see arrow in Fig. 1c) due to summary visual effect of the 3D-artery position.

Case report 2 (primary examination in 2009)

A 6-year-old girl was admitted to the intensive care unit with an acute loss of consciousness. According to the parents, she experienced transient vertigo, episodes of syncope, and sudden muscle hypotonia with complete spontaneous recovery in 1–3 h. These symptoms were admittedly caused by a sharp turn of the head and a bend of the skull to the left. Previously performed MRI scan showed moderate enlargement of the lateral and third ventricles without sings of CSF obstruction.

The patient attempted to open her eyes during verbal contact; pupils equal, round, sluggish in reaction to light; no signs of facial asymmetry, responsive for pain. Other signs included general muscular hypotonia, symmetrically diminished patellar and Achilles reflexes. No signs of meningitis were found. The lumbar puncture showed normal CSF pressure and normal cell count. The patient was diagnosed with acute brain ischemia and due to unavailability of emergency angiography, a vascular-oriented treatment was initiated, which resulted in complete recovery during the next few hours. A CT scan performed the following day found a bony prominence on the left side of the skull compressing the anterior arch of C1 (Fig. 2a). A functional X-ray angiogram revealed a normal diameter of both common and internal carotid arteries on both sides (Fig. 2b). Complete occlusion of the vertebral artery at the level of the atlanto–occipital membrane was visualized after head rotation to the left.

Despite the potential risks for the patient's health, the parents refused additional examination and insisted on the discharge from the hospital.

Case report 3 (primary examination in 2014)

A 12-year-old girl was admitted to the out-patient clinic with restricted head inclination to the left (Fig. 3a) cervical discomfort for 2 years. Non-significant asymmetry in head position was observed form the infancy. With the onset of puberty the patient expressed also esthetical concerns. The patient's concerns were also induced by the failure of the physical therapy.

The X-ray examination showed an oblique skull position, and CT scan data revealed a massive additional bone fragment adjacent to the mastoid process (Fig. 3b, c). The base of the additional bone was attached to the occipital bone, and the apex was connected the lateral part of the posterior C1 hemiarch. As clearly shown on the CT angiogram, the abnormal bone had direct contact with the vertebral artery (Fig. 3d, e). No compression or stenosis of the artery was detected, which was confirmed by Doppler ultrasound. Although there were no neurological or vascular complications, the decision to remove the pathologic bone with simultaneous decompression of the vertebral artery was made.

The bone fragment was excised through the small lateral posterior cervical approach (Fig. 4a). The vertebral artery

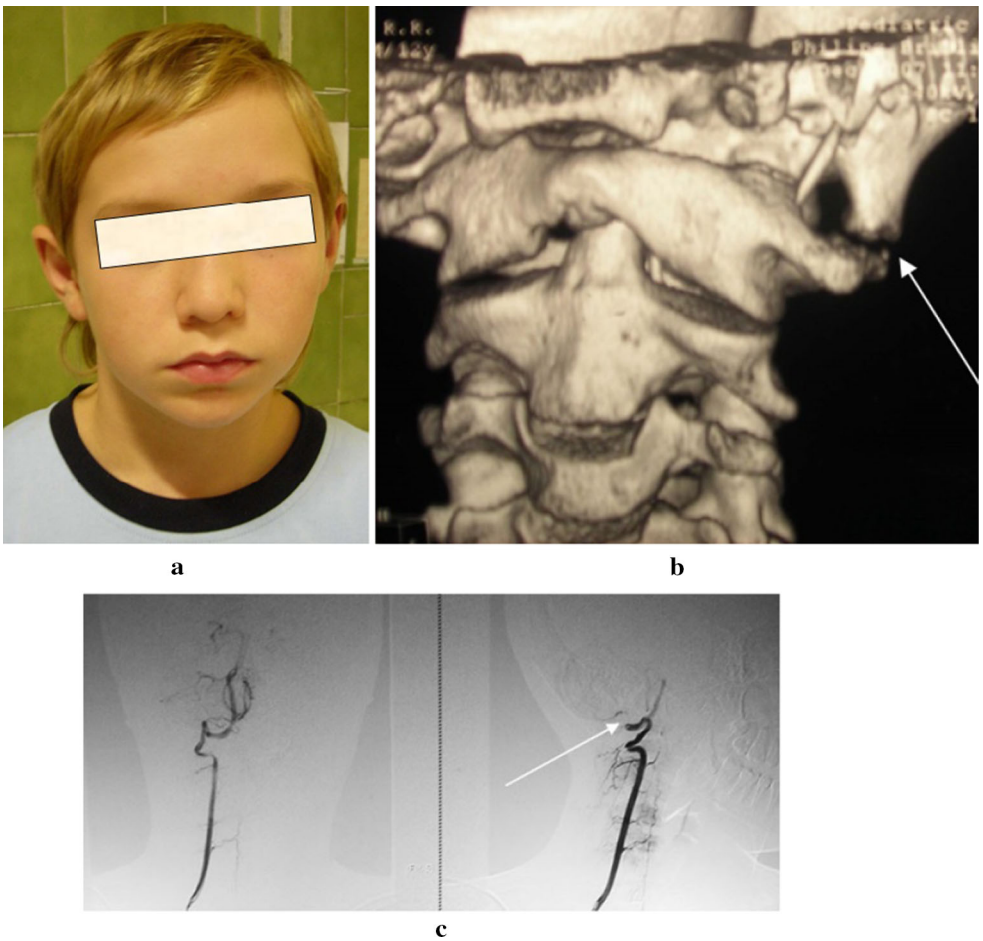

Fig. 1 The clinical photograph (**a**), CT scan (**b**), and angiogram (**c**) of the 11-year-old boy (case no. 1). **a** Facial asymmetry and torticollis; CT scan with an atlanto–occipital outgrowth (*arrow*); **c** X-ray angiogram showing compressed vertebral artery

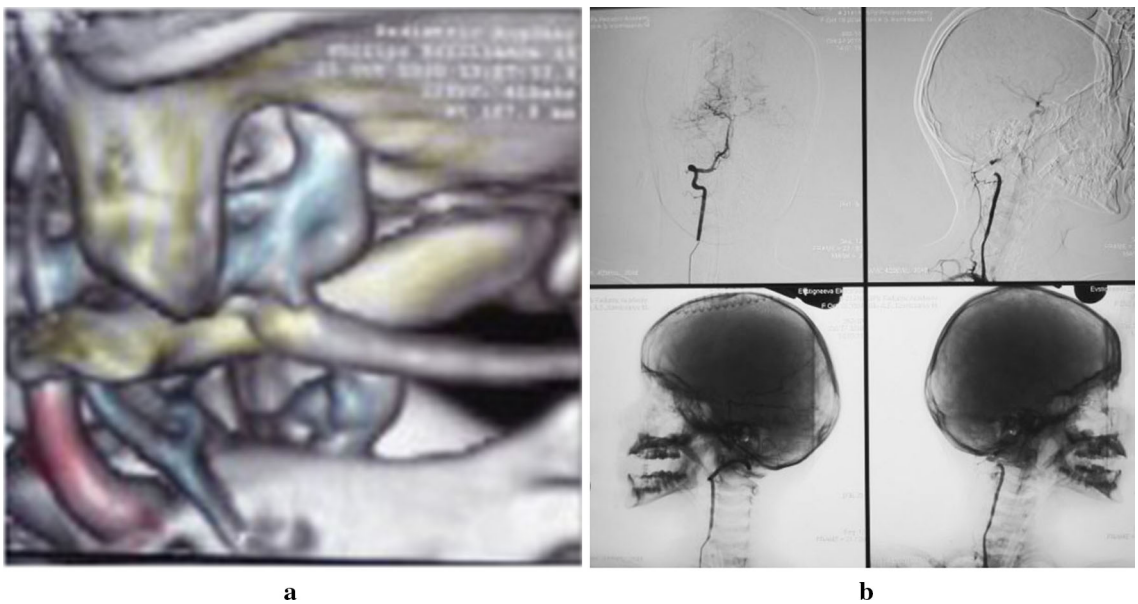

Fig. 2 Angio-CT scan (**a**) and X-ray angiogram (**b**) with a functional head-turning test in the 6-year-old girl (case no. 2). Compression of the right vertebral artery while turning the head to the left

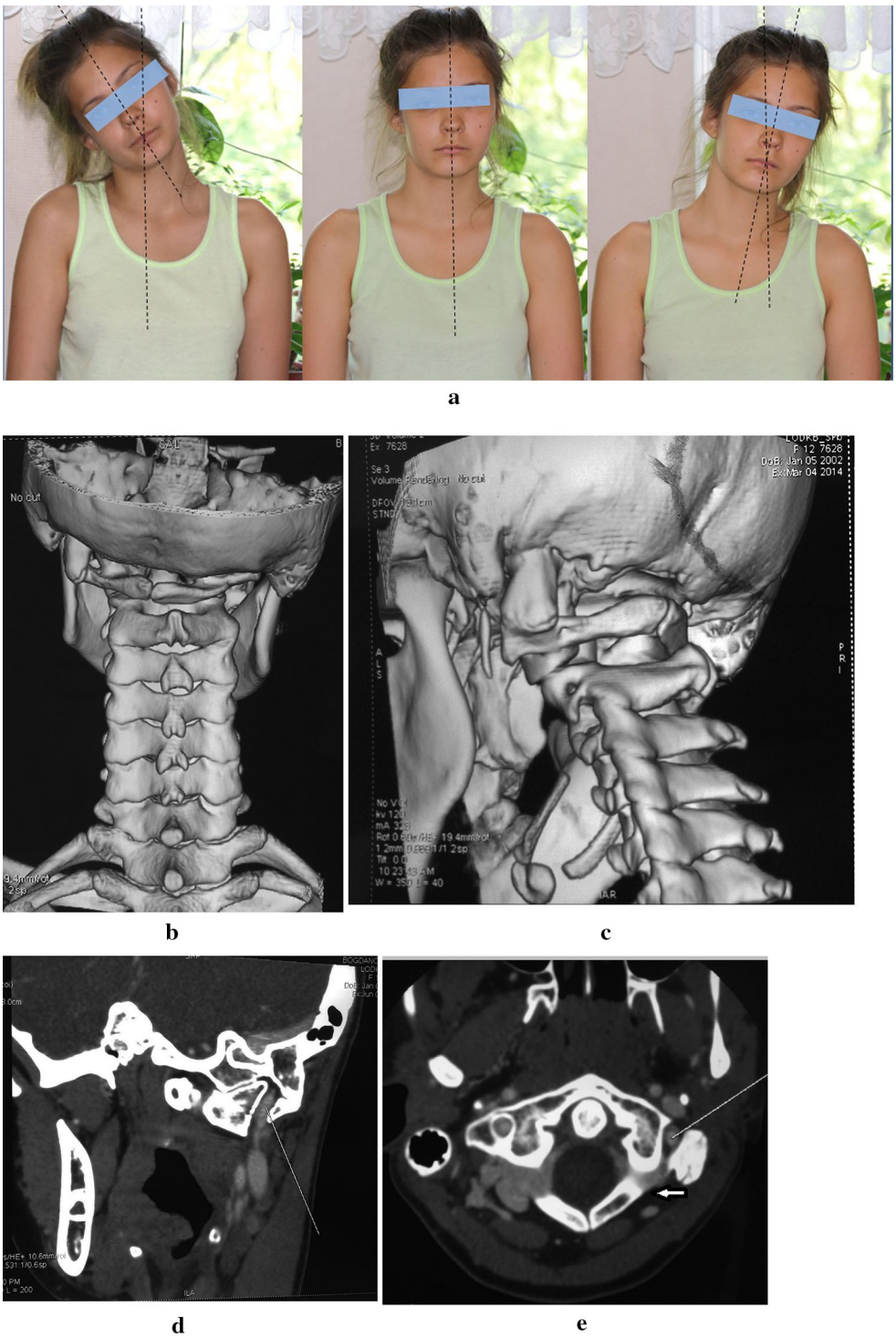

Fig. 3 Pre-op frontal photo-series (**a**), 3D-CT (**b**, **c**), and frontal (**d**) and axial (**e**) angio-CT scans of the 12-year-old girl (case no. 3). The pre-operative frontal plane maximal range of motion (ROM) is estimated as 42°d—0°—18°s. The additional bone is close to the vertebral artery

close to the medial side of the abnormal bone was identified and protected. The bone fragment was completely removed by a high-speed bur and Kerrison rongeurs to achieve a complete release of the artery (Fig. 4b). The bone wax on the base of the abnormal bone was applied.

A collar was used for immobilization and pain control for 3 days postoperatively. The head ROM was restored and the patient was complaint-free at 2, 6, 12, or 18 months postoperatively, despite mild residual asymmetry in head inclination (Fig. 4c).

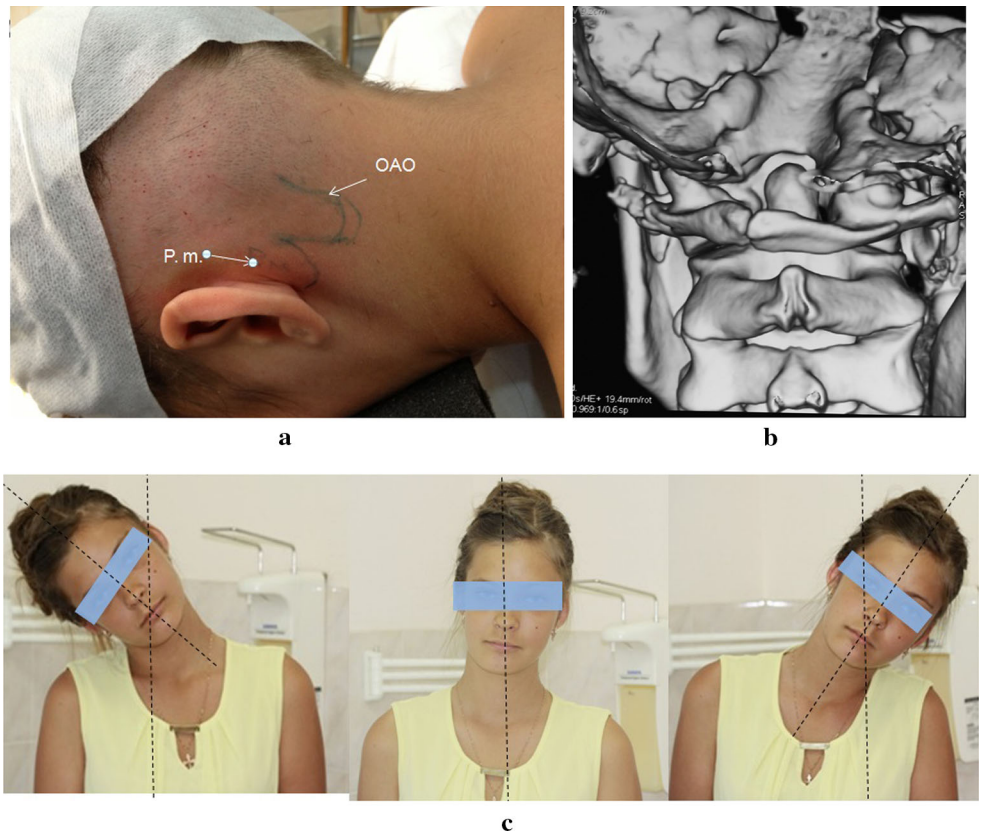

Fig. 4 Operative view (**a**), post-operative CT (**b**), frontal photo series (**c**), and 3D-CT (**b**, **c**) and frontal (**d**) and axial (**e**) angio-CT scans of the 12-year-old girl (case no. 3). ROM was significantly increased (47°d—0°—42°s) in comparison with the pre-operative data (Fig. 3). *p.m.* mastoid process, *OAO* occipital atlas outgrowth

Discussion

In the present case study, an abnormal bone formation originating from the occipital bone and compressing the atlas was detected in all three patients. In each case, the disease presented with a different main clinical sign: late torticollis in case nos. 1 and 3, acute transient cerebral ischemia in case no. 2, and the restriction of head motion and neck discomfort (case no. 3). Different causes can lead to the clinical manifestation of disease; for example, sharp movements of the head in case no. 2, active pre-pubertal growth in case nos. 1 and 3, and rehabilitation treatment recommended by doctors in case no. 3.

This pathology could be called the "atlas–occipital" bone as an analogue to the abnormal omovertebral bone, which connects the scapula and the spine in Sprengel's disease [8–10]. However, a more precise term may be to call it an "occipital–vertebral outgrowth". The unique features of this abnormality, especially its size and position, distinguish it from previous descriptions of the manifestation of pro-atlas, atlas, or atlanto–occipital synostosis [2–4], although they are likely to share a single cause in embryogenesis. The presented abnormality had different manifestation of various severity in each case, from torticollis to acute vascular disorder.

We believe that the surgical indications were the most justified in the third clinical case. However, the decision for surgical treatment should be based on a full radiological examination to evaluate the relationship between the bony, vascular, and brain structures (for example, CT angiography, including 3D-reconstruction and functional tests) as well as on the availability of minimally invasive instrumentation [11–13].

Compliance with ethical standards

Conflict of interest None of the authors has any potential conflict of interest.

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