Osteoblastoma Mimicking an Idiopathic Intracranial Hypertension Syndrome

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Osteoblastomas are rare, osteoid-forming tumors first described by Lichtenstein and Jaffe in 1956. They differ from osteoid osteomas because of the larger size and the higher vascularization. Twenty-six cases of osteoblastoma involving the temporal bone have been reported so far. Localized pain and swelling with occasional 7th and 8th cranial nerve compression are the main presenting clinical features. Raised intracranial pressure (ICP) caused by mass effect has been reported in one case, and in another case, 6th nerve palsy caused by compression of the cavernous sinus has been described. The Gold Standard for the treatment of this type of lesions consists in the complete surgical resection, which unfortunately is not possible in every occasion.

We report on the management of a small right temporal bone osteoblastoma causing compression of the right transverse-sigmoid sinus junction, which became clinically evident with isolated intracranial hypertension syndrome (IHS).

Case Report

A 7-year-old boy with a history of autism presented to a peripheral hospital complaining of headache and neck pain followed by visual disturbance. The ophthalmic examination documented a bilateral loss of visual acuity (2/10), fixed bilateral mydriasis and a Grade IV bilateral papilledema. The noncontrast-enhanced computed tomography (CT) scan of the head was unremarkable. The patient was transferred to the pediatric ward of our hospital with a provisional diagnosis of pseudotumor cerebri.

The patient underwent a magnetic resonance imaging (MRI) scan of the brain with Gadolinium which showed signs in keeping with longstanding raised ICP, including bilateral kinking of the optic nerves within the orbit, enlargement of subarachnoid spaces, flattening of the posterior sclera, and the empty sella sign [Figure 1b]. The same investigation documented a single, 16 mm, oval, T1 and T2 isointense, contrast-enhancing lesion arising in the right mastoid region, and narrowing the transverse-sigmoid sinus junction [Figure 2b].

A cerebral angiography digital subtraction angiography documented the compression of the

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Case Report
right transverse-sigmoid junction distal to the vein of Labbé, within a picture of mild right side dominance of the venous sinuses circulation in the posterior fossa [Figure 2a]. A diagnosis of raised ICP related to impaired cerebrospinal fluid (CSF) absorption was supposed, considering the intracranial venous hypertension due to the lesion compressing the sinus. We decided to proceed with the surgical removal of the lesion.

The tumor was approached through a right retrosigmoid incision. Intraoperatively, an osteolytic lesion was found on the asterion with a moderately bleeding core underneath, extending in the right mastoid. The lesion did not involve the dura and was indenting and stretching the right transverse sinus. We also considered the possibility to measure intraoperatively the hydrostatic pressure in the transverse sinus proximally to the lesion, but the potential risks related to a maneuver involving a puncture of the sinus outweighed the benefits. However, noticeably the previous proximal turgid sinus collapsed after the tumor excision.

The removal was macroscopically complete, with immediate evidence of sinus decompression. The histological examination was in keeping with a diagnosis of osteoblastoma [Figure 3].

On the postoperative time, a right mastoiditis occurred, which was successfully treated.

The postoperative course was characterized by a prompt resolution of a headache and a progressive improvement of the visual acuity over the following weeks. Indeed, a 4-week follow-up ophthalmic exam documented a visual acuity of 7/10 in the right eye and 4/10 in the left, associated with a complete resolution of the papilledema and some degree of residual left optic atrophy.

A follow-up MRI after 4 months showed the normalization of the diameter of the right transverse-sigmoid junction [Figure 1a], and a significant regression of the signs of increased ICP [Figure 1c].

**DISCUSSION**

The morphological features of the venous sinuses in the general population are extremely variable: A symmetrical venous drainage in both transverse sinuses is reported in no more than 65% of cases and a prevalence of the right dominance account for 40%–60%.[8] Venous flow impairment through the transverse-sigmoid sinus has been often observed as a potential IHS-triggering mechanism, in particular, if associated with contralateral sinus hypoplasia. Several different conditions can account for venous sinus obstruction, including thrombosis and tumor compression.[9,10]

A recent retrospective study on a pediatric series 145 MR-venograms of patients diagnosed with idiopathic intracranial hypertension, was reviewed to assess the potential role of venous outflow impairment. A dominant-sided sinus obstruction was found in 52% of cases. Furthermore, obstruction of a dominant sinus
was significantly associated with an increased opening pressure after lumbar puncture (median value 34 cm H$_2$O and highest value 65 cm H$_2$O). The presence of dominant-sided venous outflow obstruction was associated with pathological collateral venous circulation in 68% of patients. Nevertheless, Maiuri et al. recently observed that a nondominant sinus compression may lead to raised ICP, whereas obstruction of a dominant sinus may remain without clinical consequences. Such findings, contrasting with the intuitive common sense, highlight the complexity of the venous sinuses as an anatomic-functional system, where an active role is also played by the development of an adequate collateral circulation. The temporal bone is the site where the transverse and sigmoid sinuses are located and so the development of an even small mass in this position may cause a compression of these vascular structures. The histopathological diagnosis in our case was in keeping with osteoblastoma, an osteoid-forming tumor that usually develops in the long bones or in the posterior vertebral arches. It differs from osteoid osteoma for the larger size and higher vascularity. The rising from the temporal bone is extremely unusual, with only 26 cases described in the literature. Only two other cases were reported with symptoms and signs similar to those found in our patient, but the mechanism responsible for that clinical condition was different. The first case presented with IHS due to the prevalent mass effect of a huge osteoblastoma (10 cm × 6 cm × 6 cm) located in the left temporal bone; the second case presented with headache and diplopia due to an osteoblastoma invading the apex of the cavernous sinus and causing 6th cranial nerve’s palsy. In our case, we postulate that a decrease in CSF reabsorption due to venous sinus obstruction was the underlying cause of the IHS. It should be noted that our patient suffered from learning disabilities, and this may have led to a diagnostic delay and the evidence of left optic atrophy suggests a longstanding raise in ICP. Noncontrast-enhanced CT scan does not help the diagnosis as in our case, whereas the MRI was essential to show the lesion and to document the characteristic signs of raised ICP. A cerebral angiography was performed to assess the extent of sinus obstruction, as in our case the MRI-angiogram proved to be of low accuracy to such purpose. The management of symptomatic patients with compression of a cerebral venous sinus entails restoring the normal venous drainage. When surgery is deemed unfeasible, interventional radiology represents a valid alternative. Temporal bone osteoblastomas are benign tumors that may recur. Therefore the gold standard of treatment is complete surgical resection. Radiotherapy, which has been used in the past, has shown to be ineffective or even detrimental because it can promote malignant degeneration. Chemotherapy, on the other hand, does not have a role in the treatment of the primary singular benign bone tumor. 

**CONCLUSION**

We report the novel finding of IHS caused by a small temporal osteoblastoma obstructing the transverse-sigmoid junction in a pediatric patient. Complete resection of the lesion restored the venous drainage, leading to prompt clinical improvement. In the management of patients with a venous sinus compression, restoration of venous drainage should be a priority.

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**Conflicts of interest**

There are no conflicts of interest.

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