Case Reports

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Solitary occipital bone plasmacytoma with unusual ocular signs

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Solitary occipital bone plasmacytoma at the level of the confluence of sinuses is a rare condition. We report a case of an unusual combination of occipital bone plasmacytoma, ocular motility abnormalities, and papilledema. To the best of our knowledge, no case has been reported in the literature on such a combination of nosology, location and clinical course.

Keywords:

solitary bone plasmacytoma, ocular motility abnormalities, papilledema

Introduction

Solitary plasmacytoma (SP) is characterized by a single mass of clonal plasma cells, with no or minimal brain marrow plasmacytosis. It can present either as extramedullary (extraosseous) plasmacytoma (EMP), i.e., in soft tissues, or as solitary bone plasmacytoma (SBP) [1]. SP is a rare condition with a cumulative incidence of 0.15/100.000 [2]. A recent Swedish population study showed a similar distribution of patients, with a global incidence of 0.191/100.000 for male and 0.090/100.000 for female patients [3]. SBP occurs primarily in red marrow-containing bones such as vertebrae, femurs, pelvis, and ribs. EMP can involve any site or organ, with the most frequent being the head and neck region (sinuses, naso- and oropharynx), gastrointestinal tract, and lungs. SBP comprises 70% of all SP cases [4, 5]. Patients presenting with SBP, especially those cases with minimal bone marrow (BM) plasmacytosis, have a higher risk of developing symptomatic multiple myeloma (MM): approximately 50% of patients with SBP and 30% of patients with EMP develop MM within 10 years after the initial diagnosis [6]. When compared to EMP, SBP has a worse prognosis with increased progression rates to MM, although the differences do not always translate into a significant difference in overall survival [1, 3]. MM is a pathological description for a plasma cell malignancy with bone marrow involvement which is associated with a wide spectrum of clinical, laboratory, and radiological findings [7].

Paralysis or neuropathy of cranial nerve (CN) III, IV, and/or VI may be caused by various processes like abnormal circulation, neoplasm, trauma or inflammation in the skull base or brainstem. The pathological process commonly develops in the cavernous sinus secondary to inflammation (Tolosa Hunt syndrome), tumor or aneurism of the internal carotid artery (cavernous sinus syndrome).

A nuclear lesion of the oculomotor nerve secondary to ischemia, inflammation, or trauma is commonly associated with abnormal conjugate eye movements (visual pareses). Isolated lesions of the oculomotor nerve nucleus are rare due to the proximity of nuclei to the horizontal and vertical gaze centers [8].

Papilledema is a clinical sign of increased intracranial pressure (ICP). Possible conditions causing high ICP and papilledema include intracerebral mass lesions, cerebral hemorrhage, head trauma, meningitis, hydrocephalus, spinal cord lesions, impairment of cerebral sinus drainage, anomalies of the cranium, and idiopathic intracranial hypertension (IIH) [9]. The speed of papilledema development depends more on the location of the neoplasm with regard to the cerebrospinal fluid system of the brain and venous collectors (cerebral sinuses) than on the size of the mass lesion. Tumors of the posterior fossa, fourth ventricle, pineal region, or aquaeductus cerebri cause a more rapid development of papilledema than tumors located in the frontal, temporal or occipital lobes [10].

However, in our young patient, papilledemas were associated with other causes.

Case description

A forty-four-year-old military service member presented to the Subtentorial Neurooncology Department with complaints of headache for a week. He reported that in recent days, the headache had been accompanied by progressive diplopia.

His initial neurological status was remarkable for a Glasgow score of 15 and cephalgia.

On examination, the pupils were equal in size and reactive to light, and the patient was noted to have left abducens nerve palsy with left gaze diplopia. Face was symmetric and swallowing and phonation normal. Tendon and periostal reflexes were bilaterally equal and normotonic. The finger-nose test was normal and the gait was steady with a negative Romberg test. The patient was able to control his pelvic organ function.

On eye examination, his uncorrected visual acuity OD was 0.6 and OS 0.6. Intraocular pressure (IOP) was OD 20 mmHg and OS 19 mmHg. No hemianopia was present. Eye movements were restricted on outward gaze bilaterally, and downward gaze was limited in the left eye (Fig. 1).

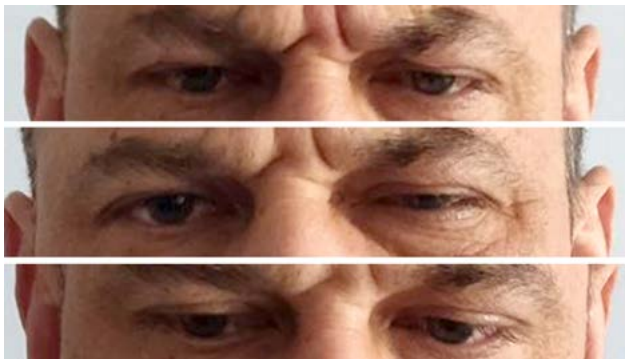

Fig. 1. Preoperative photographs

Fundocopic examination revealed hyperemic optic discs with blurred and edematous margins, no excavation and severely enlarged retinal veins. Magnetic resonance imaging (MRI) found a 15×34×55-mm posterior fossa mass lesion with occipital bone destruction (Fig. 2-3).

In the projection of the confluence of sinuses, multislice computed tomography (MSCT) of the brain showed a 15×34×55-mm neoplasm within the squama occipitalis, with significant loss of the occipital bone plate. The neoplasm exerted hardly any mass effect on the anatomical structures of the brain, with moderate compression on the cistern magna (Fig. 4-5).

On MSCT angiography (Toshiba Prime Aquilion), there was practically no contrast accumulation in the neoplasm, and the course of the major cerebral arteries was normal. MSCT venography showed the lack of filling of the confluence of sinuses and left transverse sinus.

Scheduled surgery was performed to remove a posterior fossa mass lesion. Intraoperatively, it was found that the tumor resulted in occipital bone destruction and thinning of the outer plate of the occipital bone. The tumor did not grow into, but caused compression of, the confluence of sinuses. There was postoperative MSCT evidence of total tumor removal (Fig. 6-7).

Postoperatively, there was an improvement in diplopia, palpebral fissures were equal in rest, and the ocular motility restored to normal (Fig. 8).

In addition, there was an improvement in papilledema, the disc margins became more clearly defined, and venous congestion disappeared.

Pathology of the excised tissue showed features of plasmacytoma.

At discharge, the diplopia and headache disappeared and the papilledema completely resolved.

Discussion

Solitary plasmacytoma of the occipital bone is a rather rare phenomenon. To the best of our knowledge,

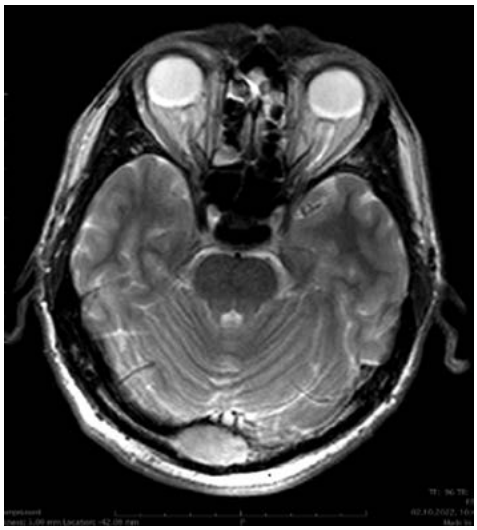

Fig. 2. Preoperative MRI, axial view

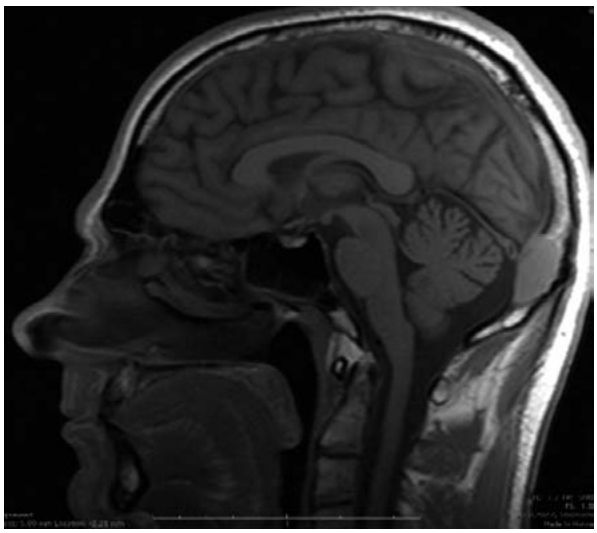

Fig. 3. Preoperative MRI, sagittal view

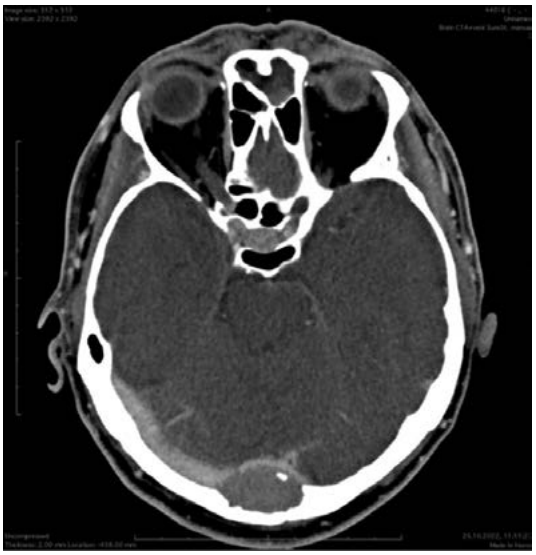

Fig. 4. Preoperative MSCT, axial view

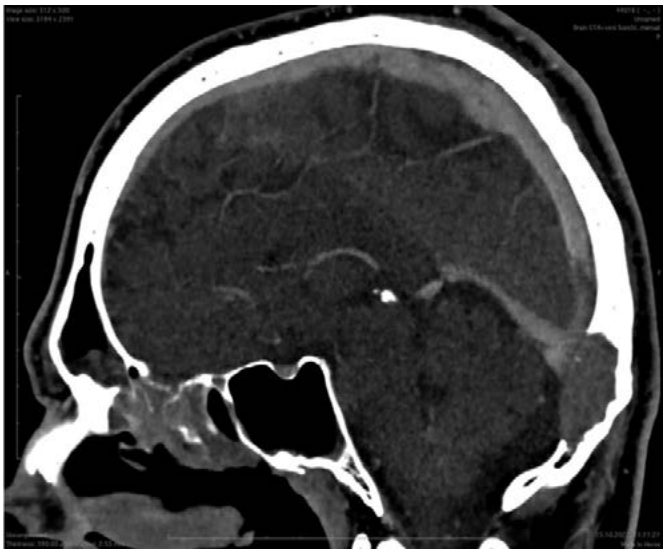

Fig. 5. Preoperative MSCT, sagittal view

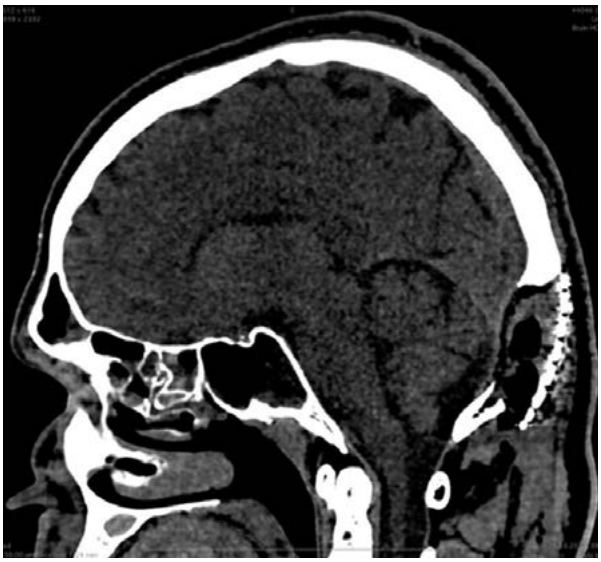

Fig. 6. Postoperative MSCT, sagittal view

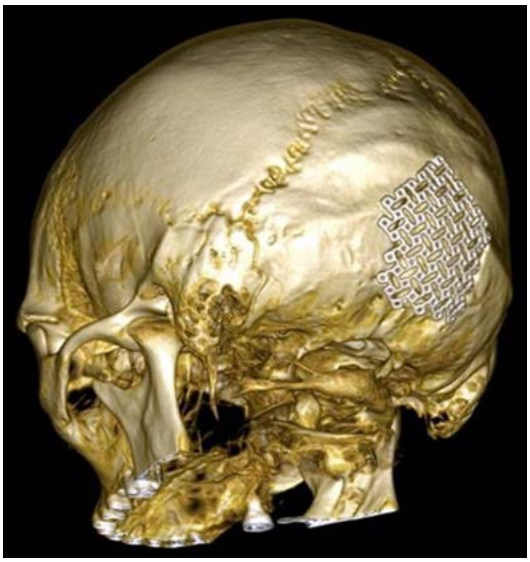

Fig. 7. Postoperative MSCT, 3D cranial reconstruction

only 11 cases with computed tomography (CT) or MRI evidence of solitary plasmacytoma of the occipital bone [11-19] were reported in PubMed until January 21, 2023. Of special rarity are reports on cases of plasmacytoma of the occipital bone with the following features seen in our case report: (1) papilledema (Okamoto et al (1997) [16] and Plant et al (1991) [18]); (2) compression of the confluence of sinuses (Jacubowski et al (1980) [19]); and (3) diplopia, but in the presence of normal ocular motility (Okamoto et al (1997) [16]). However, neither of these cases included the co-occurrence of all these three signs: diplopia, tumor compression of the confluence of sinuses and papilledema. However, this co-occurrence was mentioned in a report by Pagkou and colleagues [20] on a paramedian right occipital intradiploic epidermoid cyst at the level of the confluence of sinuses; others reported on optic disk swelling or hydrocephalus in similar cases of occipital intradiploic epidermoid cyst [21, 22, 23]. In addition, Plant and colleagues [18] reported a case of superior sagittal sinus occlusion by a Ewing's sarcoma at the level of the internal occipital protuberance, which caused papilledema and horizontal diplopia in the presence of bilateral weakness of the lateral recti. Friedman et al [24] reported on a number of cases of intracranial hypertension secondary to venous outflow abnormality - namely, brain sinus thrombosis. Since in some of these cases, symptoms were manifested not only by abducens nerve weakness, but also by weakness of other oculomotor nerves, it is reasonable to suppose that, in our case, insufficiency of the left oculomotor nerve could be secondary to increased ICP. Reid et al [25] noted that, aside from papilledema, CN VI palsy is the most common neurologic examination finding in pediatric IIH. This occurs because of elevated intracranial pressure (ICP), which results in a downward

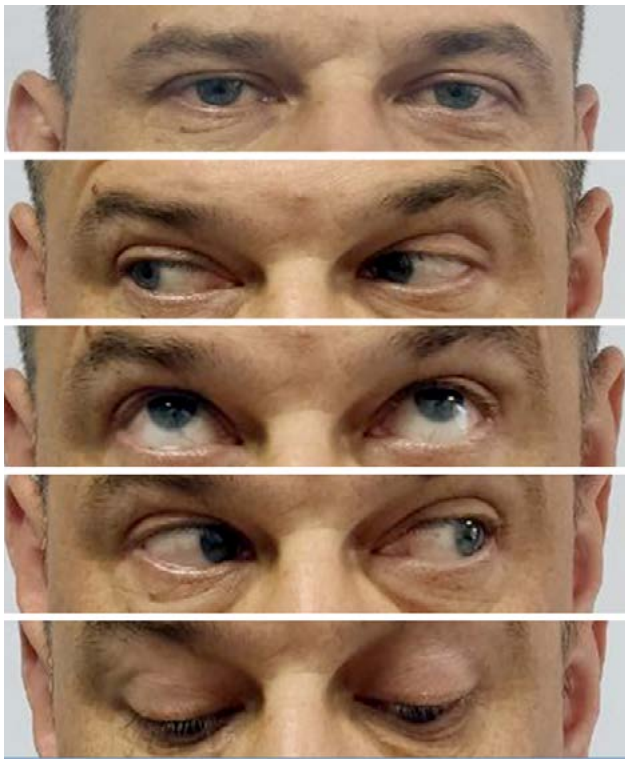

Fig. 8. Postoperative photographs

displacement of the brainstem that stretches CN VI as it crosses over the petrous ridge and enters Dorello's canal. However, this opinion is controversial, because ICP is diffusely elevated, and there is no focal source of elevated ICP that could result in a displacement of the brainstem [26]. Ding et al (2015) [27] described left transverse sinus stenting in a patient with right transverse sinus hypoplasia clinically defined by intracrainai hypertension, optic disc swelling and bilateral abducens nerve paresis. Based on a study by Lepore (2002) [26], they noted that abducens nerve paresis is the most common sign of the increased intracranial pressure. However, paresis pathophysiology differs from that described above, and it is likely that paresis develops due to the increased ICP and a long intracranial segment of the nerve. In the case reported by Ding et al [27], the bilateral abducens nerve paresis resolved within 8 hours after stenting.

Sixth nerve palsy is a common false localizing sign of raise ICP [27]; in other words, abducens dysfunction does not necessary indicate the presence of pathological process close to the nerve, and it cannot be excluded that the process is located at a distance from the lesion.

Solitary occipital bone plasmacytoma at the level of the confluence of sinuses is a rare and unusual combination of the nosology and location, which determines a special clinical course for the disease. The signs and symptoms are very similar to syndrome of brain pseudotumor (idiopathic intracranial hypertension) [28], but it is the presence of a particular cause of elevated ICP that makes the difference, and, although the cause (i.e., the process) is located extradurally, it plays a key role clinically.

Since indirect cause-effect relationships may take place among the location of the major focus and the symptoms caused, meticulous history collection, appropriate auxiliary investigations, and multispecialty approach to the diagnosis and selection of treatment strategy are required.

Conclusion

The case presented (1) improves our ideas about ocular signs of solitary plasmacytoma, (2) demonstrates that symptoms may develop at a distance from the primary site if the venous sinuses of the brain are involved in the process, and, correspondingly, (3) shows the need for appropriate auxiliary investigations in the absence of clear causes of the aforementioned ocular signs.

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Disclosures

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All authors reviewed the results and approved the final version of the manuscript.

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Ethics Statement: *The study involved human subjects, adhered to the tenets of the Declaration of Helsinki, and was approved by the Ethics Committee. Informed consent was obtained from all patients. This study was not conducted on animals.*

Abbreviations: *CN, cranial nerve; CT, computed tomography; MSCT, multislice computed tomography; MRI, magnetic resonance imaging*