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Case Report

Clival Plasmacytoma: A Rare Tumor

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Abstract

Background: Plasmacytomas are formed by monoclonal proliferation of plasma cells. This tumor can originate from bone or soft tissue. The skull base is a rare region for plasmacytomas and there are few cases reported in the literature.

Case Presentation: thirty-eight-year-old male with steadily aggravating headache that has been going on for several months and diplopia. Computerized brain tomography (CBT) was performed. Soft tissue mass causing destruction in the clivus was detected in CBT. After that, brain and pituitary magnetic resonance imaging (MRI) was performed. A mass was detected in MRI that which caused expansion in the clivus and enhancement. Erosion was detected in the sphenoid sinus posterior wall. As a result of bone marrow biopsy, 40-50% plasma cell infiltration was detected. The patient received 4 cycles of chemotherapy and then autologous stem cell transplantation therapy.

Conclusion: As a result of the 6-month follow-up, it was observed that the mass seen at the base of the skull regressed significantly and the patient was stable.

Keywords
Plasmacytoma, clivus, skull base, multiple myeloma, MRI

Introduction

Solitary plasmacytoma (SP) is the local proliferation of neoplastic monoclonal plasma cells that occur in bone or soft tissue. It accounts for less than 10% of all plasma cell neoplasms [1]. Plasmacytomas (PI) has been classified by the World Health Organization into two different types: solitary bone plasmacytoma (SBP) and extramedullary plasmacytoma (EMP) [2]. The vertebra and skull are the bones where SBP is most common [1]. EMP is commonly caused by the head and neck region, nasal cavity, and nasopharynx [1]. The skull base is a rare region for Pl and the most commonly involved bones are the orbit, sphenoid sinus, and dorsum sella [3]. The clivus is rich in bone marrow, and clival plasmacytomas (CP) are thought to be intramedullary [4]. There are very few cases in the literature where clivus involvement has been reported [5]. We present a case of clivus Pl in a 38-year-old male patient.

Case Presentation

A 38-year-old male with a steadily aggravating headache that has been going on for several months and diplopia. There was no history of nausea, vomiting, and seizures. There was no history of hypertension, diabetes, cardiovascular disease, or stroke. He had no complaints such as blurring, deviation in the jaw, facial asymmetry, hearing loss, or tinnitus. On examination, his general condition was good, conscious, and cooperative and mental functions were normal. Visual acuity and visual field examination were normal. Other system examinations were normal.

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Computerized brain tomography (CBT) was performed. Soft tissue mass causing destruction in the clivus was detected in CBT **(Figure 1).** After that, a brain and pituitary MRI was performed. In cranial and pituitary MRI: affecting the central skull base and clivus, causing pronounced expansion and erosion in the clivus, partially enclosing both ICA cavernous segments, isointense in T1 weighted images (WI), hyperintense in T2WI **(Figure 2),** contrast enhancement mass lesion of 37x32x22 mm was observed. Erosion was present in the sphenoid sinus posterior wall. The mass extended into the sphenoid sinus. It was observed that the mass was close to the adenohypophysis posterior, but the pituitary was natural **(Figure 3).** Hyperprolactinemia (107.22 ng / mL [normal, 2.64-13.13 ng / mL]) was seen in the endocrine evaluation, but there were no signs of hypopituitarism. Upon detecting a progressive increase in the patient's creatine values (serum creatinine 11.33 mg / dL [normal, 0.84-1.25 mg / dL]) and simultaneous hypercalcemia (serum Ca 12.78 mg / dL [normal, 8.8-10.76 mg / dL]) received. After that, as a result of a bone marrow biopsy performed with multiple myeloma (MM) pre-diagnosis, 40-50% plasma cell infiltration was detected. The patient received 4 cycles of chemotherapy and then autologous stem cell transplantation therapy. As a result of the 6-month follow-up, it was observed that the mass seen at the base of the skull regressed significantly and the patient was stable. Informed consent form was obtained from the patient.

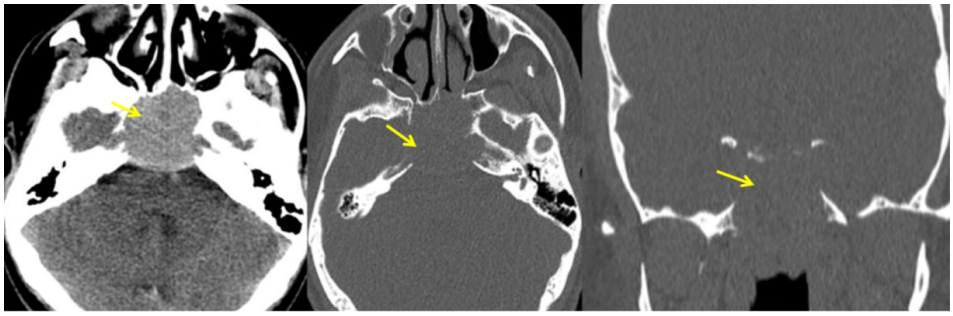

Figure 1: Soft tissue mass causing destruction in the clivus was detected in CBT.

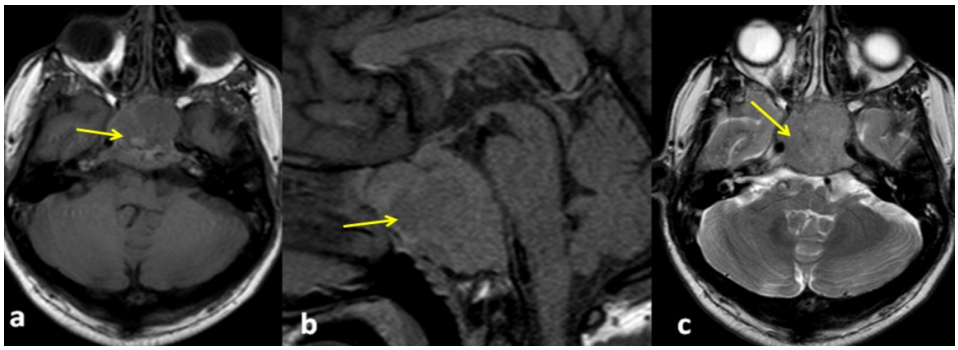

Figure 2: Affecting the central skull base and clivus, causing pronounced expansion and erosion in the clivus, partially enclosing both ICA cavernous segments, isointense in T1WI **(a, b),** hyperintense in T2WI **(c),** lesion was observed.

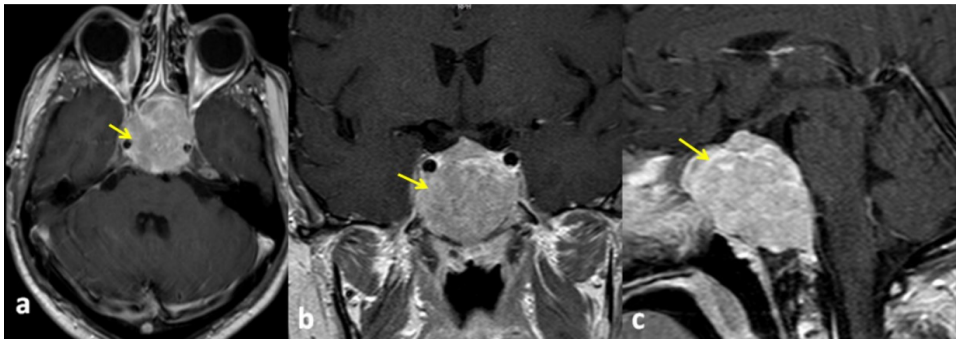

Figure 3: a, b, c. Affecting the central skull base and clivus, causing pronounced expansion and erosion in the clivus, contrast enhancement mass lesion was observed.

Discussion

Solitary intracranial plasmacytomas (SIP) are extremely rare. The main location in the intracranial area is the frontal bone. The skull base is rarely affected. Orbital apex, sphenoid sinus, and dorsum sella are preferred localizations [6]. In reported cases, CP usually occurs in the sixth decade, and female dominance (female/male ratio: 2: 1) [3,6]. The most common symptoms are headache, diplopia, vision problems, and endocrinological disorders due to seller invasion [3,6]. Direct compression or involvement of the cranial nerves causes neurological symptoms. The sixth cranial nerve (abducens nerve palsy) is most commonly involved, followed by the second, fifth, seventh, and eighth cranial nerves [6]. According to clinical data in cases where clival Pl has been reported, headache, double vision, and third and sixth cranial nerve disorders are the most common symptoms. Endocrinological disorders have been observed in 25% of cases due to sella invasion [6].

The CP can be seen as SBP, or in our case, as EMP by holding the submucosa of the sphenoid sinus. By definition, there is no evidence of underlying systemic disease at the time of diagnosis in patients with SP [2]. In MRI, intracranial Pl is observed in the isointense signal on T1WI and T2WI and enhancement. Lesions are osseo-destructive and enhanced in CT examination too [6]. In the radiological differential diagnosis of skull base Pl, nasopharyngeal carcinoma, chordoma, meningioma, osteosarcoma, lymphoma, pituitary adenoma, metastatic carcinoma, eosinophilic granuloma, and MM should be considered [6].

Trans-sphenoidal surgery is required to reduce tumor size in patients with neurological deficits and for treatment in recurrent / residual lesions [6]. SP of the skull base is a radiosensitive tumor, so radiotherapy is a preferred treatment [6]. Patient follow-up is required to detect the progression of the SIP to MM. If MM is diagnosed, chemotherapy followed by autologous stem cell transplantation is the treatment of choice in appropriate patients [6]. The average survival is 3 years in MM disease. Patients with solitary lesions have a better prognosis. <30% of EMP progresses to MM and 70% do not have disease in 10-year follow-up. However, the rate of SBP's progression to MM is more than 50%, and the 10-year disease-free period is only 16% [7].

Conclusion

Pl should be considered in the differential diagnosis of solitary clival lesions. All SP patients should be evaluated to rule out the underlying MM and should be carefully followed up to take into account the high risk of progress to MM.

Author contribution statement

All authors (HK, VK) participated in the planning, writing, editing, and review of this manuscript.

Declaration of patient consent

Informed consent was obtained from the patient for the case to be presented.

Conflicts of interest

None Declared.

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