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Isolated Diplopia as the Sole Manifestation of Solitary Clival Plasmacytoma

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Abstract

Background: Plasmacytomas are monoclonal proliferations of plasma cells occurring in bone or soft tissue without systemic involvement. Solitary bone plasmacytoma (SBP) is a localized tumor without other skeletal lesions or bone marrow infiltration. Clival involvement is extremely rare, with few cases reported. *Case Report*: A 47-year-old male presented with a 3-month history of diplopia. MRI revealed a T1- and T2-isointense clival lesion with bone destruction. Endoscopic transnasal transsphenoidal biopsy confirmed plasmacytoma. Systemic workup ruled out multiple myeloma. The patient received 50 Gy radiotherapy in 25 fractions. At 14-month follow-up, he showed complete clinical recovery and reduction in tumor size. *Conclusion*: Solitary clival plasmacytoma is a rare but treatable cause of isolated diplopia. It should be considered in the differential diagnosis of clival lesions, with radiotherapy offering favourable outcomes.

Keywords: Chemotherapy, Diplopia, Plasmacytoma, Radiotherapy, Tumor.

Introduction

Plasma cells, derived from B-lymphocytes, are essential for immune defence through immunoglobulin production. Plasma cell neoplasms arise from abnormal monoclonal plasma cell proliferation and can present as either diffuse diseases like multiple myeloma (MM) or localized forms such as plasmacytomas [1]. Plasmacytomas represent an intermediate stage between monoclonal gammopathy of undetermined significance (MGUS) and MM, and may be classified as solitary bone plasmacytoma (SBP) or extramedullary plasmacytoma (EMP), based on their site of origin [1,2]. SBPs are localized bone lesions without systemic features of MM and typically show low marrow involvement (<10%) [3]. They most commonly affect the vertebrae and skull, with rare occurrences at the skull base [4]. Among these, clival involvement is exceedingly rare, with only a few reported cases in the literature [5]. Notably, SBPs have a higher risk of progression to MM (up to 67%) compared to EMPs, leading to poorer overall prognosis [6,7].

This report presents a rare case of solitary clival plasmacytoma manifesting solely as isolated diplopia, without other neurological or systemic symptoms. The tumor showed excellent response to standard radiotherapy (50 Gy), highlighting the importance of including SBP in the differential diagnosis of clival lesions presenting with cranial nerve involvement. Early recognition is crucial for appropriate management and prognosis.

Case Report

A 47-year-old male presented with a 3-month history of diplopia. He denied associated symptoms such as headache, vomiting, seizures, or limb weakness. Neurological examination revealed isolated left sixth cranial nerve palsy with visual field defects, while the remaining cranial nerves and systemic examinations were normal. MRI brain revealed a 4.5×3.3 cm T1 and T2 isointense soft tissue mass in the clival region with homogeneous contrast enhancement and osteolytic destruction, bilaterally encasing the internal carotid arteries without sellar invasion **[Fig.1]**. Whole-body PET

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scan identified a solitary metabolically active lesion at the clivus, with no other abnormalities. Given the location, a provisional diagnosis of chordoma was considered. The lesion was partially resected via a transnasal transsphenoidal approach, and biopsy was obtained. Histopathological analysis revealed sheets of mature plasma cells with eccentric nuclei, abundant eosinophilic cytoplasm, and distinct borders. Immunohistochemistry was positive for CD138 and MUM1, with a Ki-67 index <5% **[Fig.2]**. No evidence of amyloid or epithelial malignancy was seen, and the lesion was diagnosed as clival plasmacytoma. Further evaluation to exclude multiple myeloma including skeletal survey, serum and urine electrophoresis, CBC, serum calcium, and bone marrow biopsy yielded normal results. A diagnosis of solitary clival plasmacytoma was thus confirmed.

The post-operative course was uneventful, with gradual improvement in sixth nerve palsy. The patient received 50 Gy radiotherapy in 25 fractions **[Fig.3]**. At 14-month follow-up, diplopia had fully resolved. Follow-up MRI and PET scans showed significant tumor regression and no new lesions **[Fig.4a,4b]**. The patient remains clinically stable and under surveillance.

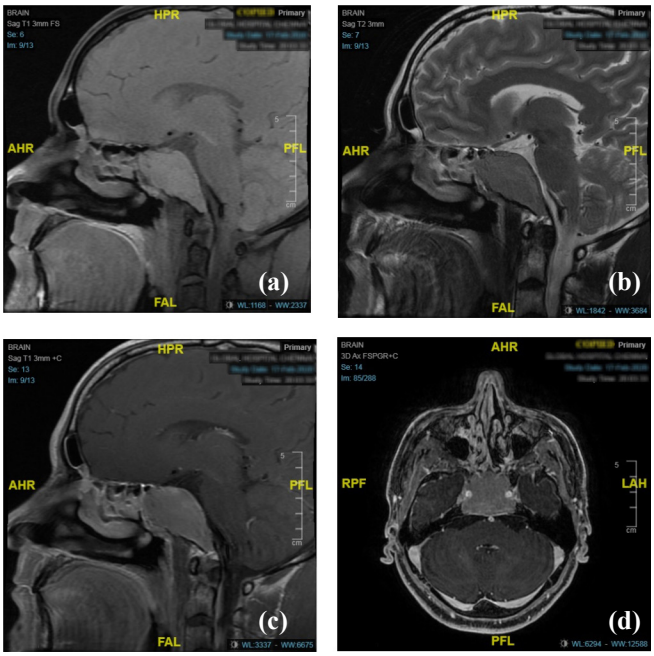

Fig.1: *Magnetic resonance imaging* **(a):** *Sagittal T1 weighted image showing isointense lesion of size 4.5×3.3 cm in the clival region with destruction of clivus;* **(b):** *Sagittal T2 weighted image showing isointense lesion;* **(c):** *Sagittal T1 contrast image showing homogeneous contrast enhancement;* **(d):** *Axial contrast enhancement image showing bilateral ICA encased by the lesion.*

Discussion

Solitary bone plasmacytoma (SBP) and multiple myeloma lie at opposite ends of the spectrum of plasma cell dyscrasias. SBP is a localized collection of monoclonal plasma cells, commonly affecting vertebrae and skull bones. Although vertebral plasmacytomas are frequently reported, intracranial plasmacytomas, especially those at the skull base, remain rare. The clivus is an uncommon site, with only a few isolated cases reported in literature, typically presenting with cranial nerve deficits due to its critical anatomical location.

To date, only nine cases of solitary clival plasmacytoma have been described in the literature from 1980 onwards. Most patients were middle-aged to elderly (5th-6th decades), with a male

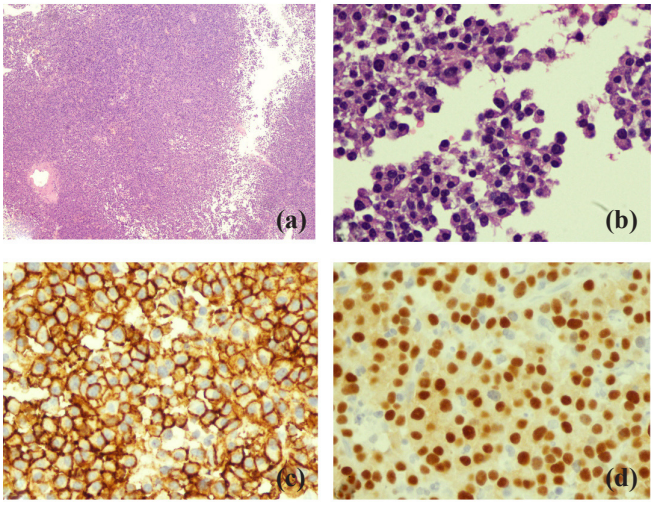

Fig.2: *Histologic features of solitary clival plasmacytoma* **(a):** *Low power (×100) haematoxylin and eosin stain showing sheets and aggregates of mature plasma cells;* **(b):** *High power magnification (×1000) demonstrating abundant eosinophilic cytoplasm, eccentric nuclei and clock face like chromatin;* **(c):** *Immunohistochemical staining positive for CD138;* **(d):** *Immunohistochemical staining positive for MUM1.*

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predominance. Headache and diplopia were the most frequent presenting symptoms. Sixth cranial nerve involvement was most commonly reported, followed by the second, fifth, seventh, and eighth nerves [8]. One rare case also presented with lateral medullary and bulbar syndrome due to vascular compression [9]. All reported cases were managed with biopsy or subtotal resection, followed by radiotherapy. Only one case underwent complete excision but resulted in early mortality while in other patient, gamma knife surgery was required after initial treatment [10,11].

Radiologically, clival plasmacytomas appear as osseous-destructive lesions on CT and demonstrate iso to hypointense signals on T1-weighted and iso to hyperintense signals on T2-weighted MRI, with uniform enhancement [12]. Histologically, these tumors are composed of sheets of plasma cells showing varying degrees of cytologic atypia. Hallmark features include eccentric nuclei with clock-face chromatin and expression of plasma cell markers like CD138 and CD38. Light chain restriction confirms clonality [13]. Diagnosis of SBP requires the exclusion of systemic disease. Criteria include a solitary lesion, normal skeletal survey, bone marrow with <5% plasma cells, serum monoclonal protein <2g/dL, no Bence-Jones proteinuria, and absence of anemia, hypercalcemia, or renal impairment [14,15]. Whole-body PET scan plays a crucial role in excluding multiple myeloma.

Surgery is primarily reserved for biopsy or decompression in symptomatic cases. Its role in definitive treatment remains unclear [12]. Radiotherapy is the mainstay of treatment, with SBP being highly radiosensitive. Doses of 40-50 Gy are generally recommended. Higher control rates (up to 94%) have been reported with doses exceeding 40 Gy [16,17]. The role of chemotherapy in SBP remains uncertain. While some studies suggest improved outcomes with adjunct chemotherapy such as melphalan and prednisolone [18], newer agents like thalidomide and proteasome inhibitors

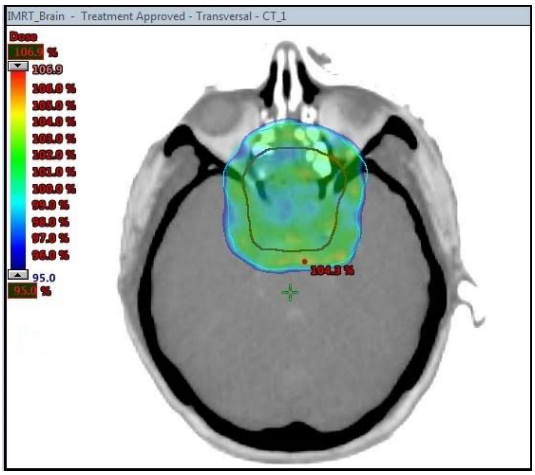

Fig.3: *Axial planning computed tomography slice showing typical dose colour wash and dose distribution of intensity modulated radiotherapy.*

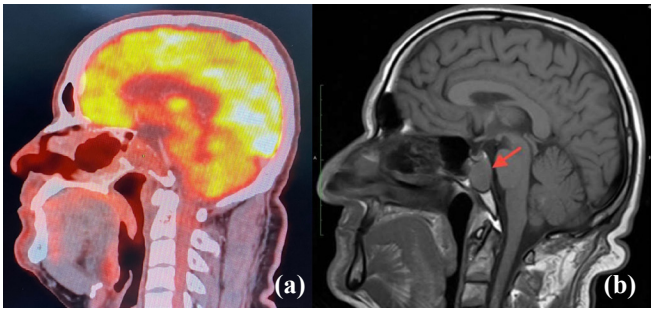

Fig.4: (a): *Follow up MRI after 12 months showing reduction in size of the tumor;* **(b):** *Follow up PET scan showing reduction in the size of lesion and metabolic activity.*

are under investigation [19]. Despite appropriate treatment, 50-60% of SBP cases may eventually progress to multiple myeloma, often within 2 years [20,21]. Larger lesion size (>5 cm) and older age are associated with higher progression risk. Five-year overall survival stands at approximately 70%, with disease-free survival around 46% [22].

Conclusion

Solitary clival plasmacytoma is a rare entity and can mimic other skull base lesions like chordoma [23,24]. Radiotherapy offers excellent local control, while surgery plays a diagnostic and symptomatic role. Given the high risk of progression to multiple myeloma, long-term surveillance with imaging

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and laboratory tests is essential. Clinicians should consider clival plasmacytoma in the differential diagnosis of isolated diplopia, as early recognition and radiotherapy can achieve good outcomes while avoiding unnecessary surgical morbidity.

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