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Case Report

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Pituitary extramedullary plasmacytoma without any systemic involvement: a rare case report

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ABSTRACT

Extramedullary plasmacytoma (EMP) is a rare soft tissue plasma cell disorder without systemic involvement like multiple myeloma (MM). It develops outside of the bone marrow. It happens seldom for a plasma cell neoplasm to manifest as an intracranial or cranial tumor, and it is much less common for it to resemble a pituitary adenoma. In patients with sellar area plasmacytomas, headaches, cranial nerve deficits, visual abnormalities, bloody nasal discharge, and discomfort in the eyes and craniofacial regions are common presentations. A biopsy is required to make the diagnosis. For parasellar plasmacytomas to be successfully managed, an accurate diagnosis is essential. Overall, patient survival is excellent, although it might be lowered if they go on to develop overt MM. Suppose there is no known history of MM. In that case, it is advised to do a thorough workup to identify any underlying MM or to closely monitor any MM development in the future. In this study, we aimed to report a rare case who presented with loss of vision without systemic involvement of MM and whose pituitary biopsy was a plasmacytoma.

Keywords: Pituitary plasmacytoma, multiple myeloma, radiotherapy, monoclonal gammopathy

INTRODUCTION

Extramedullary plasmacytoma (EMP) is a rare soft tissue plasma cell disorder without systemic involvement like multiple myeloma (MM). EMP usually affects the upper respiratory system, which includes the larynx, nasopharynx, sinuses, and nasal cavity.[1] In a few number of instances, the brain has been implicated. Approximately 4% of all plasma cell tumors are EMP.[2] It is less common for myelomatous illness to cause involvement of the central nervous system (CNS), and most of these individuals exhibited normal systemic symptoms of MM before developing brain involvement.[3] In patients with sellar area plasmacytomas, headaches, cranial nerve deficits, visual abnormalities, bloody nasal discharge, and discomfort in the eyes and craniofacial regions are common presentations.[4] A biopsy is required to make the diagnosis. It is necessary to follow up on these patients later. As per research, 25-45% of individuals receive a subsequent diagnosis of MM; hence, it is important to seek assessment for systemic disease upon diagnosis to further guide therapy.[5,6] In this study, we aimed to report a rare case who presented with loss of vision without systemic involvement of MM and whose pituitary mass biopsy was a plasmacytoma.

CASE

A 45-year-old female patient was admitted to the emergency department with acute loss of vision. There were no neurological or endocrinological clinical findings other than acute vision loss in the patient's history. In the physical examination performed in the emergency room, no pathological findings were found in the systemic and neurological examination, except for vision loss. In the patient's cranial magnetic resonance imaging (MRI), a 32x47 mm contrast-enhanced mass was detected in the pituitary gland, expanding the sella and extending to the clivus. The suprasellar cistern was narrowed, and there was slight pressure on the optic chiasm (**Figure 1 a-d**). The patient underwent microscopic transsphenoidal surgery via an endonasal approach. The tumor was gross-total resected. Histopathologically, the surgical specimen showed a monotonous plasmacytoid cell population with eccentric nuclei and abundant cytoplasm (**Figure 2a**). Immunohistochemical staining was positive for CD138, MUM1, (**Figure 2b, 2c**), and the kappa light chain, but not the lambda light chain (**Figure 2d, 2e**). Based on these findings, the tumor was diagnosed as a plasmacytoma.

The patient had no complaints other than acute vision loss. Neurological examination was normal except for visual impairment. After the excision of the pituitary mass, vision loss resolved. There was no comorbidity in the patient's history and no medications. There was no history of malignancy in his family history. Laboratory tests revealed a hemoglobin level of 11.5 g/dl, leukocyte 10.8x10^9/L, and platelet 391x10^9/L. No endocrinological anomaly was detected in the patient's

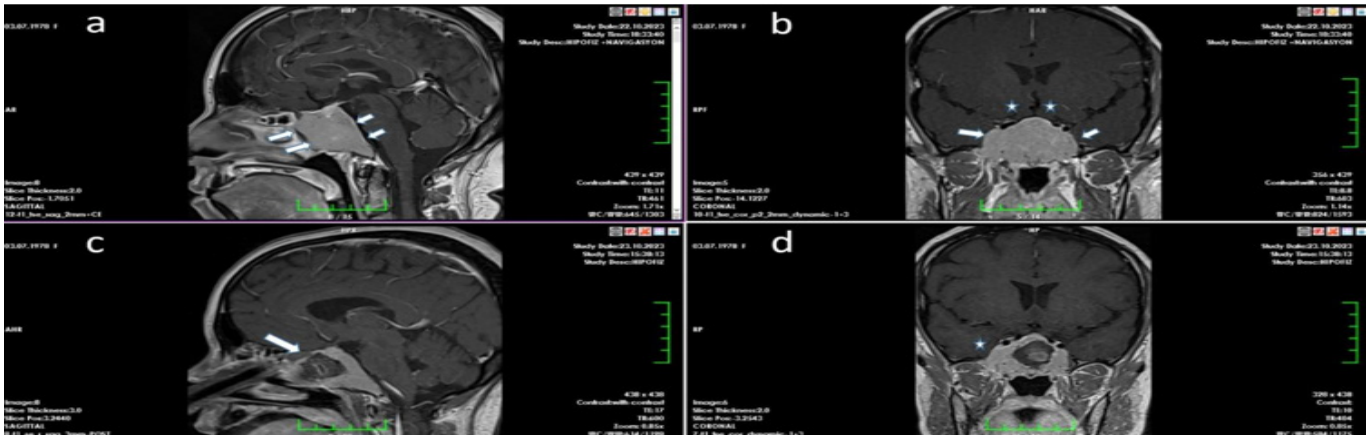

Figure 1. (**a,b**): Preoperative sagittal and coronal sections contrast-enhanced MRI, (**c,d**): postoperative sagittal and coronal sections contrast-enhanced MRI

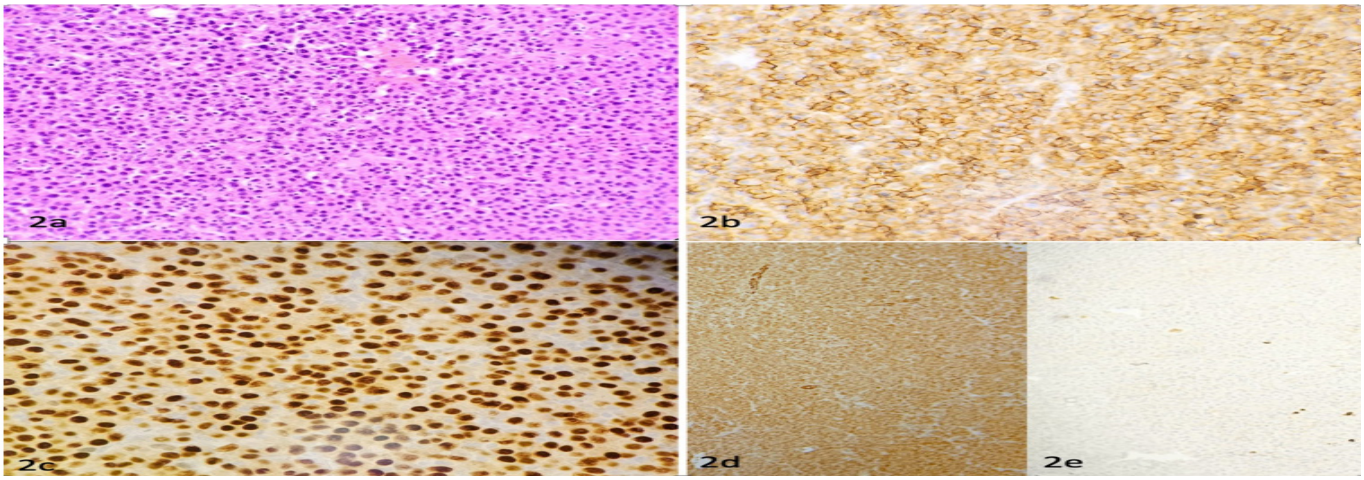

Figure 2. (**a**) Neoplastic plasmacytoid cells in diffuse sheets (HEX200), (**b**) Neoplastic cells with diffuse CD138 positivity (X200), (**c**) Neoplastic cells with diffuse MUM 1 positivity (X200), (**d**) monoclonal kappa light chain positivity (X100), (**e**) Lambda light chain immunonegative (X100)

laboratory findings. There was no evidence of renal dysfunction or hypercalcemia. No monoclonal gammopathy was detected in protein electrophoresis and serum, urine immunofixation. Serum immunoglobulin levels and serum-free light chains were found to be within the normal range. Serum beta-2 microglobulin was normal. The patient underwent bone marrow aspiration and biopsy. The plasma cell rate was observed to be around 8%. No increase in monoclonal plasma cells was detected in bone marrow biopsy. The patient was evaluated with positron emission tomography (PET). There was no involvement or any lytic lesion in the patient's PET findings. The EMP that was diagnosed was found to be a mass in the pituitary region with no systemic symptoms. The patient was consulted with radiation oncology for residual disease in the pituitary. The patient was given 46 Gy/23 FX radiotherapy to the pituitary region. The patient's vision loss was completely resolved. The patient, who had no active complaints, was kept under close clinical follow-up.

DISCUSSION

MM, solitary plasmacytoma of bone, and EMP single lesions with a microscopic appearance of plasma cell neoplasms without any clinical or radiological evidence of MM are among the clinicopathologic entities that are classified as plasma cell neoplasms.[7] It happens seldom for a plasma cell neoplasm to manifest as an intracranial or cranial tumor, it is much less common for it to resemble a pituitary adenoma.[8] Through a careful endocrine workup, neurological, radiological assessment, and sellar biopsy, most reported cases had no known diagnosis of plasma cell tumor until the sellar biopsy. This is because a plasmacytoma is one of many intrasellar masses that can mimic a pituitary adenoma.[9,10] The management of plasmacytoma and MM necessitates cooperation between subspecialists in hematology with a focus on stem cell transplants, radiation oncologists, and surgeons. Radiotherapy or surgery is usually recommended when a single plasmacytoma is detected, or both may be used together. There was no systemic involvement in the case we presented. There is a solitary plasmacytoma. She had the maximum safe excision of the plasmacytoma, which helped with debulking and alleviated her symptoms. Then, she got a total of 46 Gy/23 FX radiotherapy. Before a surgical pathologic evaluation, the tumor's rarity, clinical presentation, and imaging results that resemble those of other regional cancers sometimes lead to a misdiagnosis as pituitary adenomas, chordomas, or meningiomas. A misdiagnosis can postpone systemic therapy for the underlying MM and result in an incorrect surgical strategy. Lee et al.[11] reported a healthy 65-year-old male patient with no previous medical illness. Neurological examinations reported normal functioning of the cranial nerves, except for the patient's complaint of diplopia that occurred 2 weeks before presentation, similar to our case. Blood tests and pituitary hormone evaluations showed normal results, as in our case. It was reported that the patient developed MM 15 months later.[11] Another patient, a healthy 54-year-old male with no previous medical illness, was admitted to the hospital with intermittent headaches. Neurological examinations, including cranial

nerves, and ophthalmological tests showed normal results. Pituitary hormones were evaluated as normal. However, it was reported that the patient was given chemotherapy and radiotherapy because of pituitary plasmacytoma and involvement in the bone marrow biopsy.[11] Ferreira et al.[12] reported a 68-year-old male patient who first applied to the endocrinology clinic due to gynecomastia, decreased libido, and sexual impotence. Histological examination revealed plasmacytoma and MM was excluded. The patient was treated unsuccessfully with radiation therapy (no tumor shrinkage). Myeloma eventually developed and several similar lesions occurred in different locations. The patient was started on chemotherapy and underwent bone marrow transplantation.[12] Sidlo et al.[1] reported an extremely rare case of sudden death due to intrasellar EMP in a 24-year-old female patient with no previous clinical findings. The cause of death was determined to be CNS failure. Jin et al.[13] reported 5 cases of parasellar plasmacytoma. All patients underwent endonasal endoscopic surgery with adjuvant therapy. They reported complete remission after postoperative radiotherapy at a median follow-up of 41 months (range, 15–120).[13]

CONCLUSION

In conclusion, for parasellar plasmacytomas to be successfully managed, an accurate diagnosis is essential. Overall, patient survival is excellent, although it might be lowered if they go on to develop overt MM. Suppose there is no known history of MM. In that case, it is advised to do a thorough workup to identify any underlying MM or to closely monitor any MM development in the future. Because of all these, we followed our patient closely.

ETHICAL DECLARATIONS

Informed Consent

The patient signed and free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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