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Extramedullary Plasmacytoma Of Forehead – A Rare Entity.

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ABSTRACT

Plasmacytomas are tumors linked to multiple myeloma and are classified as either intramedullary or extramedullary based on their site of origin. Extramedullary plasmacytomas, which occur outside the bone marrow, account for less than 5% of plasma cell malignancies and are most commonly found in the head and neck region - such as the nasopharynx, nasal cavity, sinuses and tonsils. Involvement of the frontal bone is rare, with only a few cases documented in the literature. We present the case of a 65-year-old woman who presented with a swelling on her forehead, which was diagnosed as a plasmacytoma on fine needle aspiration cytology (FNAC). The diagnosis was further confirmed using cell block analysis and immunohistochemical staining.

Keywords: Plasmacytoma, FNAC, myeloma.

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INTRODUCTION

Plasma cell tumors can present as one of three distinct pathological entities: multiple myeloma, extramedullary plasmacytoma and solitary bone plasmacytoma. While multiple myeloma is a systemic condition, both extramedullary and solitary plasmacytomas are localized neoplasms without the features of end organ damage i.e. CRAB. These tumors are marked by the abnormal monoclonal proliferation of mature B-cells, which produce monoclonal immunoglobulins through clonal expansion. Extramedullary plasmacytomas arise in extraosseous locations and typically involve upper respiratory tract including nasal cavity, paranasal sinuses and nasopharynx often presenting with a mass effect, localized pain, and invasive behavior [1]. The exact cause of plasmacytoma remains uncertain, although factors such as viral infections and physical trauma have been implicated. In particular, Hepatitis C and Epstein-Barr virus have been suggested as potential viral contributors. Surgical resection is considered an effective primary treatment for solitary craniocerebral plasmacytoma [2].

CASE REPORT

A 65-year-old female presented to the Surgery Outpatient Department with a complaint of forehead swelling for one month (Photograph 1). On clinical examination, the swelling was soft to firm in consistency, located over the glabella and measured approximately 3–4 cm in size, extending toward the right medial canthus. The swelling was not associated with facial pain, and there were nasal obstruction and orbital symptoms. There was no history of fever, loss of weight or loss of appetite.

Photograph (1a and 1b) of patient showing forehead swelling extending to right medial canthus

Ultrasonography of the lesion revealed an irregular, ill-defined, heterogeneously hypoechoic mass measuring 4.3 × 2.3 cm, demonstrating internal vascularity on color doppler imaging. The lesion appeared to extend into the frontal sinus. A subsequent CT scan identified a hyperdense mass lesion measuring 6.5 × 3.0 cm in the frontal region, seemingly arising from the frontal sinus. The mass was associated with bony destruction of the frontal bone and the medial walls of both orbits, with extension into the nasal cavity (Photograph 2). There was no evidence of infarct or intracranial hemorrhage. The bilateral ventricular system and sulcal spaces were within normal limits, with no significant midline shift. The posterior cranial fossa structures also appeared normal.

Fine Needle Aspiration Cytology (FNAC) of the mass revealed highly cellular smears composed of numerous mature and immature plasmacytoid cells, dispersed singly and in clusters against a hemorrhagic background. Binucleate and multinucleate plasma cells along with occasional plasmablasts were also noted. On high-power microscopy, the plasma cells exhibited characteristic round to oval eccentrically placed nuclei with abundant basophilic cytoplasm (Photomicrograph 1).

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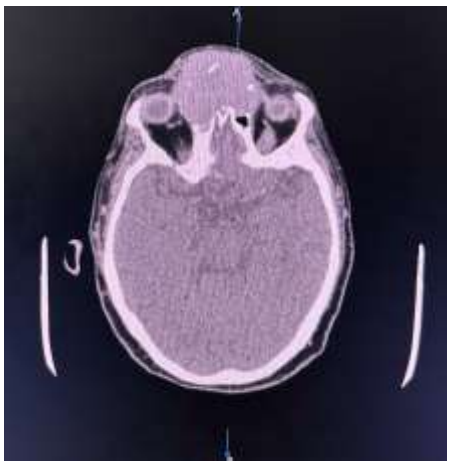
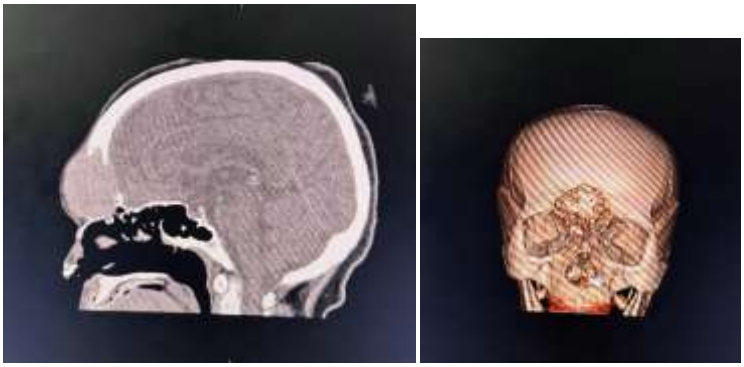
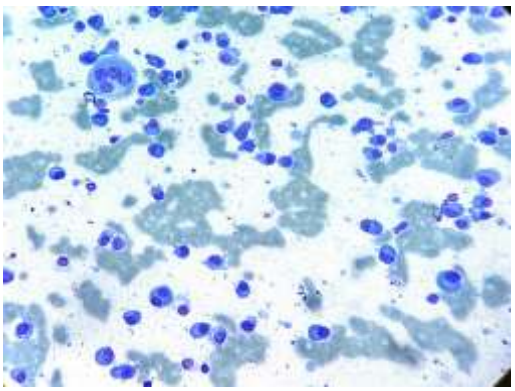
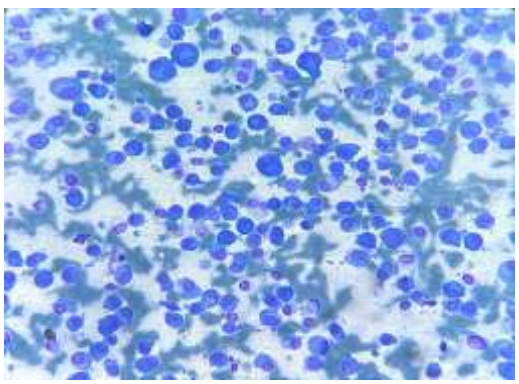




Photograph (2a, 2b and 2c) showing CT scan of the mass arising from the frontal sinus. The mass was associated with bony destruction of the frontal bone and the medial walls of both orbits, with extension into the nasal cavity.

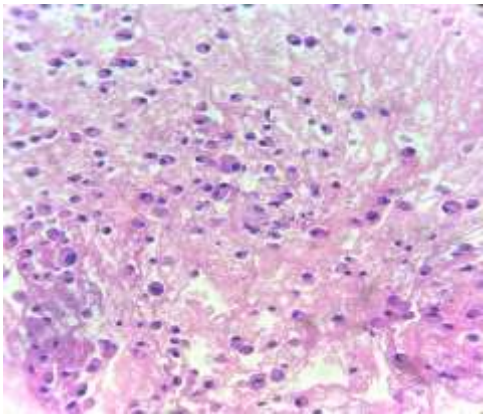




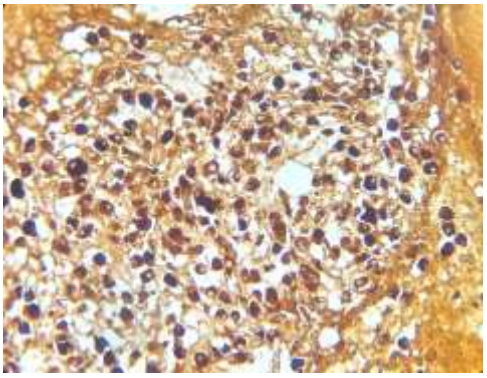
Photomicrograph 1 (Leishman Stain; 400x) showing numerous mature and immature plasma cells dispersed singly and in small clusters. Numerous binucleate forms are also seen.

Based on cytological findings, a diagnosis of plasmacytoma was made. A thorough evaluation was conducted to rule out multiple myeloma, including a skeletal survey, bone marrow biopsy, serum protein electrophoresis (SPEP), quantitative serum immunoglobulin levels, serum calcium, serum albumin, renal function tests, and beta-2 microglobulin assessment. All results were within normal limits. Additionally, Bence Jones protein was also not detected in the 24- hour urine sample.

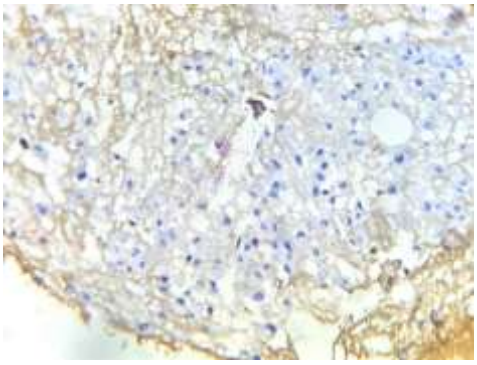
A cell block was prepared (Photomicrograph 2), and immunohistochemical (IHC) analysis confirmed the diagnosis of Kappa-restricted extramedullary plasmacytoma. On IHC, tumor cells showed positivity for CD138 and Kappa light chains, while Lambda was negative.



Photomicrograph 2 (H&E Stain; 400x) showing numerous plasma cells in cell block



Photomicrograph 3 (IHC Stain; 400x) showing kappa positivity on cell block



Photomicrograph 4 (IHC Stain; 400x) showing lambda negativity

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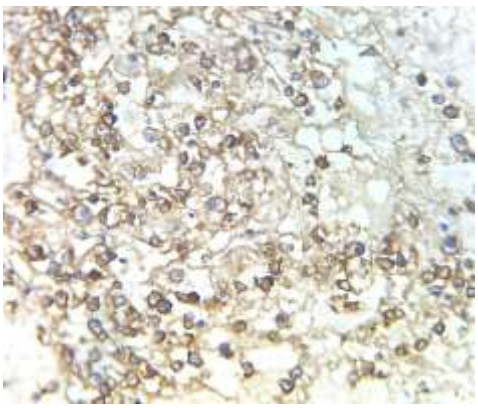

Photomicrograph 5 (IHC Stain; 400x) showing CD138 positivity

DISCUSSION

Extramedullary plasmacytomas (EMPs) are plasma cell tumors that develop outside the bone marrow. These most commonly occur in the head and neck region, followed by involvement of the gastrointestinal tract, urinary bladder, central nervous system, thyroid, breast, testes, parotid gland, lymph nodes, and skin [3].

Although EMPs can present at any age, they are most frequently diagnosed in individuals in their fifth or sixth decade of life. Initial investigations should include a complete blood count, metabolic profile and urinalysis all of which are typically within normal limits in isolated EMP. Clinical signs and symptoms vary significantly and are often non-specific, largely depending on the tumor's anatomical location. In cases where the neoplasm originates from the skull or dura, neurological manifestations—such as headache, nausea, vomiting, and visual disturbances—are usually attributed to elevated intracranial pressure. Due to these non-specific features, EMPs are often misdiagnosed prior to surgery. The differential diagnosis for cranial EMPs includes meningioma, metastatic tumors, lymphoma, osteochondroma, infectious meningitis, and sarcoma [4, 5].

Histologically, EMP must be distinguished from plasma cell granuloma, which is immunohistochemically characterized by a polyclonal plasma cell population, unlike the monoclonal pattern observed in EMP [6]. Surgical excision is considered an effective primary treatment strategy. However, higher cure rates have been reported when radiotherapy - either preoperative or postoperative - is combined with surgical intervention [7, 8].

Despite treatment, EMP carries a risk of dissemination or progression to multiple myeloma (MM), necessitating close post-treatment surveillance. Follow-up should include clinical assessment, skeletal surveys, serum protein electrophoresis (SPEP), and bone marrow evaluation to monitor for signs of disease progression. EMPs involving the skull base have a particularly high risk of transformation into MM, with progression rates exceeding 50% and a 10-year survival rate of only 16% [9].

CONCLUSION

Extramedullary plasmacytomas are uncommon neoplasms and rarely present as masses in the forehead region. This case underscores the diagnostic value of fine needle aspiration cytology (FNAC) in evaluating forehead swellings, as it plays a crucial role in guiding appropriate treatment strategies. Radiotherapy, often followed by surgical resection of any residual tumor can yield excellent clinical and radiological outcomes. Importantly, FNAC can help avoid the need for extensive surgical procedures in this anatomically sensitive area [10].

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