# Plasmacytoma skull: A rare entity

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## ABSTRACT

Plasmacytoma is a localized accumulation of abnormal plasma cells that grow within the soft-tissue or bony skeleton. Solitary plasmacytoma most commonly occurs in the bones of the axial skeleton, such as vertebra, skull, and others. Extramedullary type of plasmacytoma is generally observed in the head and neck and most frequently observed in the nasal cavity and nasopharynx. We want to enlighten the case of a 55-year-old hypertensive female patient who presented with a swelling on the left side of the forehead of size 7 cm × 4 cm for 2 years, which was associated with a constant headache. Her computed tomography scan of the brain with a three-dimensional reconstruction of the skull was suggestive of a diffuse swelling involving the left side of the forehead suggestive of a calcified lesion involving the skull with no involvement of the brain parenchyma. The patient was then subjected to elective craniectomy and was planned for cranioplasty after 6 weeks. Histopathology was suggestive of lymphoproliferative disease/plasmablastic lymphoma, after which the patient was subjected to radiotherapy.

**Key words:** *Brain, Plasma cells, Plasmacytoma, Skull*

Plasmacytoma is a rare disorder and its presentation in the skull is considered much rarer and less reported in the literature [1]. It is an infrequent form representing 5–10% of all plasma cell neoplasms [2]. Plasmacytoma, a form of plasma cell dyscrasia, is the proliferation of plasma cells, which can manifest as multiple myeloma, primary amyloidosis, or monoclonal gammopathy of unknown origin [3]. Plasmacytoma is divided as primary and secondary with multiple myeloma and may arise from osseous (medullary) or non-osseous (extramedullary) sites.

Primary extramedullary plasmacytoma can be solitary or multiple [4]. Solitary plasmacytoma is characterized by a localized accumulation of neoplastic monoclonal plasma origin cells and classified into two types according to the location as solitary plasmacytoma and extramedullary plasmacytoma [1]. Solitary plasmacytoma most commonly occurs in the bones of the axial skeleton, such as vertebra, skull, and others. Extramedullary type of plasmacytoma is generally observed in the head and neck and most frequently observed in the nasal cavity and nasopharynx also [5].

The rationale to report this case report is to enlighten more on the less engraved topic of the plasmacytoma skull and provides valuable information about the clinical presentation, radiological appearances, and management of patients of plasmacytoma skull.

## CASE REPORT

A 55-year-old female patient came to the department with chief complaints of swelling on the left side forehead of size 7 cm × 4 cm for 2 years (Fig. 1). She also complained of a constant headache, which was radiating to the whole of the head. She was also a known case of systemic hypertension for 15 years and was on antihypertensive medication. On examination, the vitals were within normal limits. The Glasgow Coma Scale was 15/15.

The patient was subjected to hematological examination such as complete blood picture, and renal and liver function tests which were within normal limits. Her overall nutritional status was assessed and was fit accordingly. The patient was then subjected to computed tomography (CT) scan brain with a three-dimensional reconstruction of the skull which was suggestive of a diffuse swelling involving the left side of the forehead suggestive of a calcified lesion involving the skull with no involvement of any part of the brain parenchyma (Fig. 2). Magnetic resonance imaging of the brain was suggestive of no parenchyma involvement.

The patient was then subjected to elective left frontal craniectomy, in which a major part of the left side frontal skull bone was excised and was then planned for cranioplasty after 6 weeks with titanium mesh plate of the skull. However, on histopathology, it was suggestive of lymphoproliferative disease/plasmablastic lymphoma plasmacytoma of the bone (Fig. 3). For further treatment, the patient then underwent post-operative radiotherapy and a good response to radiotherapy was observed on follow-up. After radiotherapy, the patient was then subjected to elective cranioplasty with titanium mesh plate after 6 months. At follow-up, the patient was healthy and living a good life.

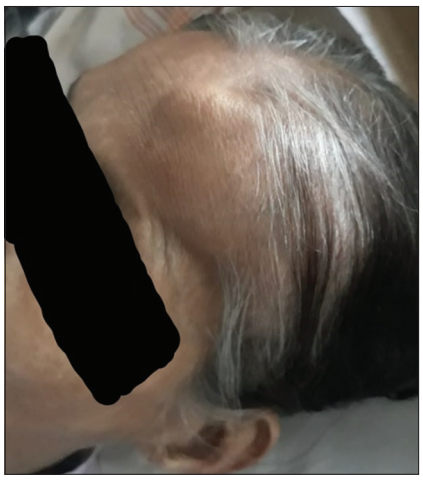

**Figure 1: Picture showing swelling over the forehead**

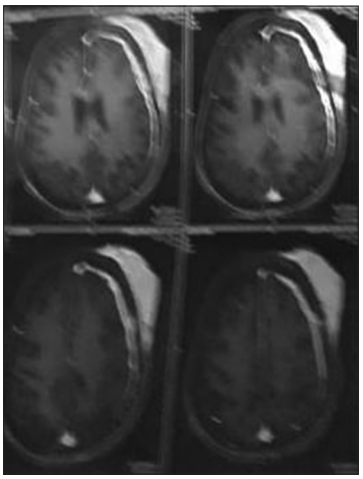

**Figure 2: Computed tomography scan showing calcified lesion over the skull**

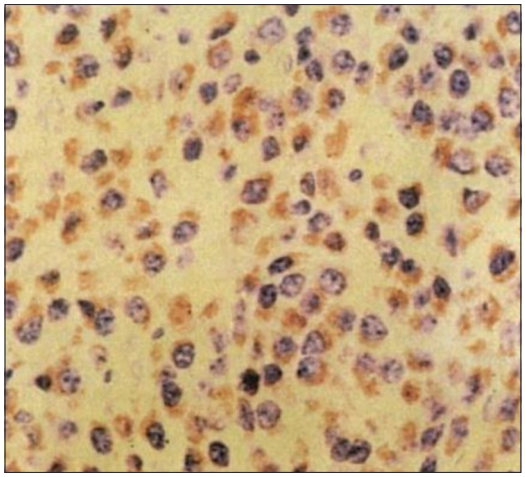

**Figure 3: Microscopy showing plasma proliferation in histology examination**

## DISCUSSION

A plasmacytoma is a unique solitary mass of neoplastic monoclonal plasma cells in either bone or soft-tissue (extramedullary). Solitary plasmacytoma is divided into two groups: Plasmacytoma of the skeletal system (solitary bone plasmacytoma) and extramedullary plasmacytoma [6]. The diagnosis of extramedullary plasmacytoma of the soft-tissue has been based on the following criteria: (a) Pathological tissue evidence of monoclonal plasma cells involving a single extramedullary site; (b) no bone marrow involvement; (c) no anemia, hypercalcemia or renal impairment caused by plasma cell dyscrasias; (d) negative skeletal survey results; and (e) low serum or urinary levels of monoclonal immunoglobulin [2].

The etiology of this disease remains unknown, but factors, such as chronic irritation from inhaled irritants or viral pathogenesis, have been previously indicated. The diagnosis of plasmacytoma can be missed with a differential diagnosis such as carcinoma nasopharynx, chordoma, meningioma, osteosarcoma, lymphoma, pituitary adenoma, metastatic carcinoma, eosinophilic granuloma, and multiple myeloma.

Primary plasmacytoma, whether osseous or non-osseous is distinguished from multiple myeloma by the absence of hypercalcemia, renal insufficiency and anemia, absence of bone marrow plasmacytosis, and serum or urinary protein level of <2 g/dL, is characteristic of plasmacytoma. The incidence rate of solitary plasmacytoma in the African is around 30% higher than the Cuscasian [7,8].

Radiotherapy remains the mainstay management of extramedullary plasmacytoma. As plasmacytoma skull is a radiosensitive tumour local control rate of 90 to 100% with associated conversion rate to multiple myeloma is 30%. Moderate dose radiotherapy of at least 40 Gy using limited radiation fields is recommended. Due to the high rate of recurrence and progression to multiple myeloma, follow-up radiological and electrophoresis assessment is required following treatment. The overall 10-year survival rate is ~70%. A literature search revealed no publications supporting the use of surgery alone to treat extramedullary plasmacytoma [1]. Surgical removal, followed by post-operative irradiation, is the treatment of choice because solitary plasmacytoma bone is very radiosensitive [9,10].

High local control rates are reported with radiotherapy, although the optimum dose and extent of radiotherapy portals remain controversial [11-13]. The local tumor recurrence rate is <10% by conventional radiotherapy [14]. The efficacy of chemotherapy for solitary plasmacytoma of bone is not certain. Shih *et al*. [15] and Delauche-Cavallier *et al*. [16] failed to find any benefit on the outcome for patients who had received chemotherapy.

## CONCLUSION

Solitary plasmacytoma of the skull is a rare presentation of a disease and is a challenge for neurosurgeons for diagnosis preoperatively. A proper diagnosis and management is the mainstay in the treatment of plasmacytoma of the skull. Primary treatment with surgery and post-operative radiotherapy gives a good response in patients of plasmacytoma.

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