

# Atypical Presentation of Plasmacytoma

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**Abstract** Plasmacytoma is a tumor seen with multiple myeloma and its localized forms are solitary bone plasmacytoma and extramedullary plasmacytoma. While the most frequent locations are vertebrae, sternum, nasopharynx, paranasal sinuses, rarely mediastinum, kidney, testis and ovary may be involved. In this case, relapsed multiple myeloma with atypical extramedullary plasmacytoma located in the frontal region is presented.

**Keywords:** Plasmacytoma, multiple myeloma

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## 1. Introduction

Plasmacytoma is a tumor originating from plasma cells. Kahler disease or Morbus Kahler statements were used firstly for plasmacytoma in 1889 by Bozzolo. V.RUS TITZKY used the term "multiple myeloma" in 1873 [1]. It has two types: solitary plasmacytomas of bone (SPB) and extramedullary plasmacytomas (EMP). Extramedullary plasmacytomas are plasma cell tumors that arise outside of the bone marrow. They are solitary lesions, and are most often located in the head and neck region, mainly in the upper aerodigestive tract, but may also occur in the gastrointestinal tract, urinary bladder, central nervous system, thyroid, breast, testes, parotid gland, lymph nodes, and skin.

Multiple myeloma (MM) is characterized by the neoplastic proliferation of a single clone of plasma cells producing a monoclonal immunoglobulin. This clone of plasma cells proliferates in the bone marrow and often results in extensive skeletal destruction with osteolytic lesions, osteopenia, and/or pathologic fractures. It accounts for approximately 1 percent of all cancers and slightly more than 10 percent of hematologic malignancies in the United States (US) [2]. Multiple myeloma, extramedullary plasmacytoma and solitary bone plasmacytoma constitute sub-groups of plasma cell tumors [3,4,5].

Here, we present a case of plasmacytoma atypically placed on the forehead of a patient with MM.

## 2. Case

A-59 year old female patient presented to hematology outpatient clinics with complaints of rapidly progressing

scalp swelling on her forehead for the last 3 months. We found a tense globular swelling over left frontal area, with a size of 5x4 cm (Figure 1).



Figure 1.

She was anemic without organomegaly or lymphadenopathy. Physical examination was normal other than that. Her past history revealed diagnosis of multiple myeloma 4 years ago and she had been treated with 8 cycles of VAD (Vincristine, Adriamycin, Dexametason). Two years ago, autologous bone marrow transplantation had been performed. Laboratory values were as follows: leukocyte count 6500/microL, hemoglobin 10,2 g/dl, platelet count 86000/microL, total protein 12,8 g/dl, albumin 3,3 g/dl. Skull X-ray (Figure 2), contrast computed tomography of brain (Figure 3) and contrast magnetic resonance imaging (MRI) of the brain were suggestive of an osteolytic skull lesion. At her brain MR, a mass with a size of 19x12 mm on her left frontal area was seen. Fine-needle aspiration was performed. Monoclonal plasma cell infiltration with a rate of 95% was detected.



Figure 2.



Figure 3.

### 3. Discussion

Plasma cell neoplasms are divided into multiple myelomas, solitary plasmacytomas of bone and extramedullary plasmacytomas. Multiple myelomas represent with systemic disease, where solitary plasmacytomas of bone and extramedullary plasmacytomas represent with local forms of plasma cell neoplasm.

Solitary plasmacytomas of bone are defined as clonal proliferations of plasma cells which are identical to those of plasma cell myeloma, manifesting as a localized osseous growth. Localized solitary plasmacytoma of bone is a rare disease and it is characterized by one or two isolated bone lesions, with no evidence of disease dissemination and it has been considered to be curable with radiotherapy and surgical resection.

The incidence of solitary plasmacytomas of bone has been reported to be approximately 3/1000000 annually [6]. MM is also slightly more frequent in men than in women (approximately 1,4:1). MM is a disease of older adults. Plasma cell myeloma mostly occurs in the elderly over 40 years of age; the median age of individuals with the disease in the United States is 62 years old [7]. Bone destruction due to myeloma may occur in any area of the body. Its incidence is as follows: spine 49%,skull 35%, pelvis 34%,ribs 33%,humerus 22%,femur 13%, mandible 10% [7]. Tumors which occur in the skull are called cranial myelomas or cranial plasma tumors. Single tumors are rarely seen in the clinical setting [8]. In our case, plasmacytoma was at the left frontal area.

At cases presenting with solitary osteolytic skull lesions, workup for differential diagnosis of plasmacytoma and multiple myeloma is essential. Cases of plasmacytoma should be on lifetime regular follow up, as they have a tendency to progress into multiple myeloma.

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