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Case Report

# Giant forehead plasmacytoma as a growing lump in a patient with multiple myeloma ☆

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ABSTRACT

Extramedullary plasmacytoma represents less than 5% of plasma cell malignancies, 85% corresponding to head and neck masses. Symptoms are related to compressive effects according to location, aesthetics issues and can be misleading associated with soft tissue disorders. In this case report, we discuss a 70-year-old woman who presented with a 3-month history of a growing painless forehead lump and confusion, for which she had an emergent simple head computed tomography scan. The images revealed a well-defined mass eroding the frontal bone with multiple lytic lesions that were also found along with long bones radiography. The mass biopsy showed a monomorphic plasmatic cell infiltrate, bone marrow studies confirmed the diagnosis of a light chain secreting multiple myeloma. Extramedullary plasmacytoma is a very unusual first presentation form of multiple myeloma and represents a clinical and radiological challenge. A systematic approach of lytic bone lesions along with the differential diagnosis of head masses are skills the clinician should develop to promptly recognize this condition considering further complications of delayed treatment. In this case, the histopathological confirmation allowed the patient to avoid neurosurgery and the early start of systemic chemotherapeutic treatment.



## Introduction

Multiple myeloma (MM) represents 10% of hematological neoplasms [1], it is slightly more frequent in men than in women with a median presentation age of 65 years at diagnosis [2]. Diagnosis criteria of MM include evidence of more than one MM defining events plus clonal bone marrow plasma cells ≥10% or confirmation of plasmacytoma in biopsy [1].

Plasmacytomas are divided into 2 groups depending on their relationship with bone structures: paraskeletal plasmacytomas or extramedullary plasmacytomas [3].

☆ **Contributors**: We were all involved in the diagnosis, management, and treatment of the patient. We were all involved in writing the manuscript. Written consent for publication was obtained from the patient.

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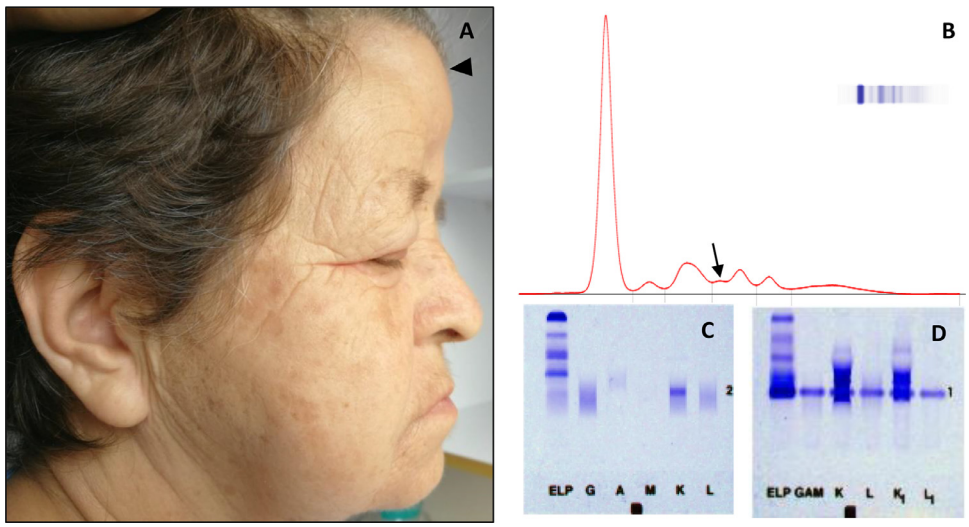


**Fig. 1 – Forehead visible lump (A - arrowhead). Serum protein electrophoresis (B) with hypogammaglobulinemia and α2-β1 bridge (arrow). Serum (C) and urine (D) immunofixation studies both positive for kappa free light chain, Bence Jones protein was detected in urine.**

Extramedullary involvement is unusual and represents 1% or 2% of all cases at diagnosis and 8% of patients can present it in the course of the disease [4].

We discuss the case of a woman with multiple myeloma whose first manifestation was a frontal plasmacytoma as an extramedullary disease. We describe the diagnostic process and clinical follow-up.

## Case presentation

A 70-year-old woman presented to our hospital with a 3-month history of a growing painless frontal lump on her scalp. Her chief complaint was confusion, she was unable to recognize her relatives. She was otherwise healthy.

On examination vital signs were normal, she had a frontal soft painless mass of 8 × 6 cm and a smaller temporal mass with no inflammatory changes (Fig. 1). Her breast, neck, lungs, skin, abdomen, and neurological examination had no other relevant findings. Initial blood tests showed moderate hypercalcemia 12.4 mg/dL (normal range 9.5-10.5 mg/dL), acute kidney injury with serum creatinine 4.42 mg/dL (previous 1.52 mg/dL, normal range 0.8-1 mg/dL) and mild anemia – hemoglobin 11.7 g/dL (normal range 12-13 g/dL).

Computed tomography (CT) of the head showed a well-defined mass eroding the frontal bone with peripheral bone pieces and multiple smaller skull lesions (Fig. 2). The mass measured 8.3 × 6.5 cm and there were multiple lytic intradiploic lesions, the middle line shift was absent. According to this presentation, we suspected a plasmatic cell neoplasm and x rays of large bone revealed multiple lytic lesions (Fig. 3). There was no hypergammaglobulinemia but serum and urine immunofixation were positive for kappa free light chains with urinary Bence Jones protein (Fig. 1). A biopsy of the frontal mass showed a monomorphic plasmatic cell infiltrate (Fig. 4). High-level kappa light chains were measured in serum and urine. A diagnosis of a light chain secreting multiple myeloma with a gigantic plasmacytoma of the skull was made.

A chemotherapy regimen with cyclophosphamide, bortezomib, and dexamethasone (CyBorD) was prescribed. After dexamethasone, the lesion size was rapidly reduced (Fig. 5) and the renal function progressively recovered. On her follow up, after the fourth chemotherapy cycle, a complete response was documented by normal serum-free light chains and no further brain images (CT or magnetic resonance imaging) were performed.

## Discussion

One of the challenges a physician must overcome is making a proper and prompt diagnosis. This is especially true in the Oncology field, where new technologies and investigations have provided different treatment options.

Clinical practice is usually based on finding the main symptom and rationally thinks select possible differential diagnoses, considering epidemiology, typical presentation, clue symptoms, physical exam, and laboratory findings. Atypical presentation of common diseases represents a real challenge in daily practice.

MM clinical presentation is characterized by osteolytic bone lesions and it is often associated with hypercalcemia, renal insufficiency, anemia, and bone lesions documented by radiography, CT, or PET-CT (formerly known as CRAB) meaning myeloma defining events. Diagnostic criteria were recently updated and are based on 2 main features that must be present: >= 10% of plasma cells in bone marrow biopsy or a tissue demonstrated plasmacytoma plus any one or more of myeloma defining events, bone marrow biopsy with ≥60% of plasmatic cells, high serum light chains (>100 mg/L), and more than one focal magnetic resonance imaging lesions of 5 mm [5]. MM typically presents with increased monoclonal

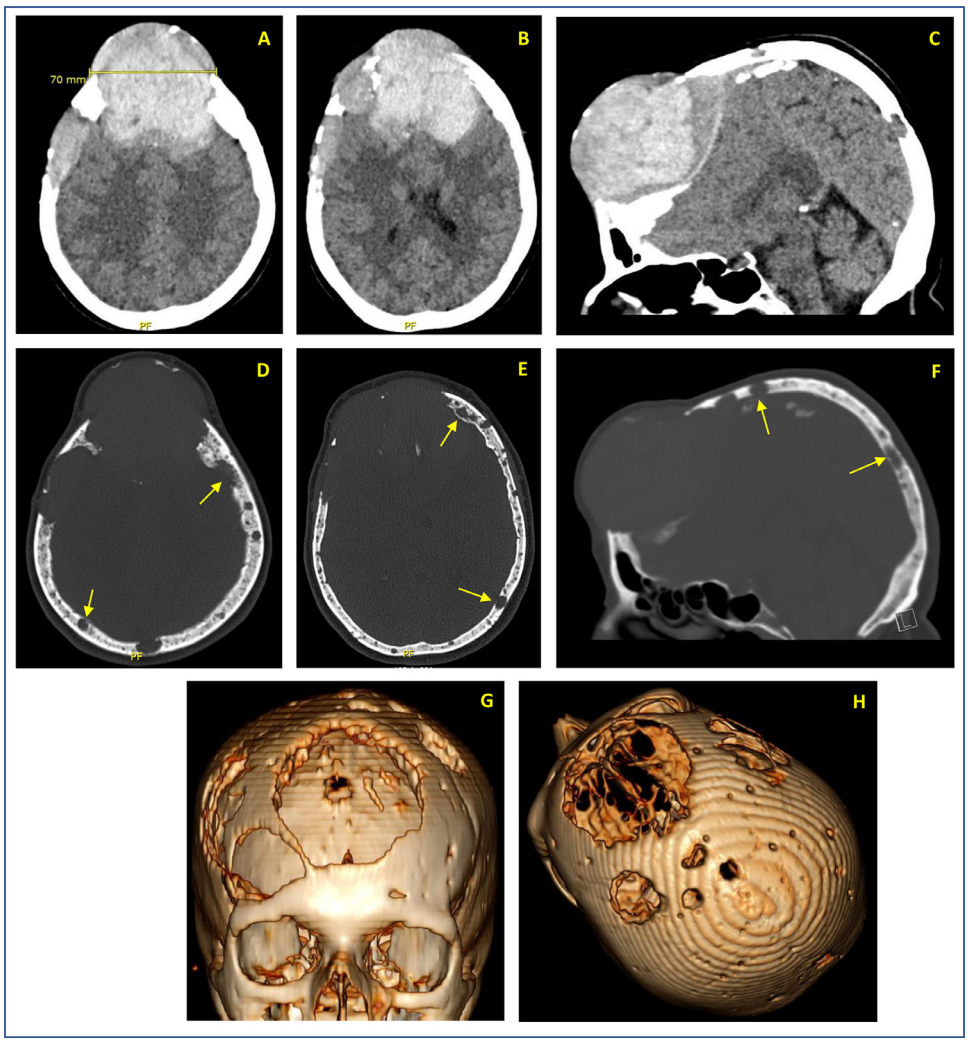


**Fig. 2 – CT head axial and sagittal brain (A, B, C) and bone window (D, E, F) of a mass eroding the frontal bone with bony fragments (arrowhead) and lytic lesions in skull (arrows). 3D rebuilding from head CT series: frontal view with complete loss of frontal bone below the mass (G), upper view with multiple erosive bone areas corresponding with lytic bone lesions (H).**

antibodies mainly IgG or IgA, called M protein in secretory disease. Nevertheless, 2% of all MM is associated with light chain production kappa or lambda [1]. In opposition to MM, solitary plasmacytoma requires only histopathological demonstration of plasm cells in the mass in the absence of bone marrow involvement by plasmacytes and other MM criteria previously exposed [5].

Central nervous system involvement is more frequent in other hematologic malignancies such as lymphomas or leukemias [6] and, plasmacytomas are rarely seen in central nervous system, which made this case remarkable.

On one hand, the CRAB component was evident and turned MM as a likely diagnosis but was not the cardinal symptom. On the other hand, the forehead mass did not fit with the MM classical picture and was the main patient complaint. The patient was programmed to neurosurgical mass removal for tissue study, but kidney failure halts the process. In the clinical approach of nontraumatic scalp masses, there are 3 main groups: round and well-defined masses (trichilemmal cyst, dermoid cyst, epidermoid cyst, lipoma, slow-flow vascular malformation, and sinus pericranii), soft-tissue infiltrating (plexiform neurofibroma, basal cell carcinoma, and squamous cell carcinoma), and associated with bone lesions (Langerhans cell histiocytosis, intraosseous hemangioma, atypical and malignant meningioma, lymphoma, and metastases) [7,8]. Finally, the tissue study was safely performed with fine needle mass biopsy reporting forehead plasmacytoma and pointing to MM as the principal diagnosis. As a limitation, no further brain image was performed and thus the evolution was formerly clinical.

The classic presentation of hypercalcemia, anemia, kidney injury, and bone lytic lesions is typical for multiple myeloma, nevertheless, the atypical plasmacytic frontal mass and the absence of monoclonal antibodies represented a clinical challenge to make a diagnosis. Concordance of both bone marrow and mass biopsies guided to a medical successful management avoiding surgical therapy in hematologic disease.

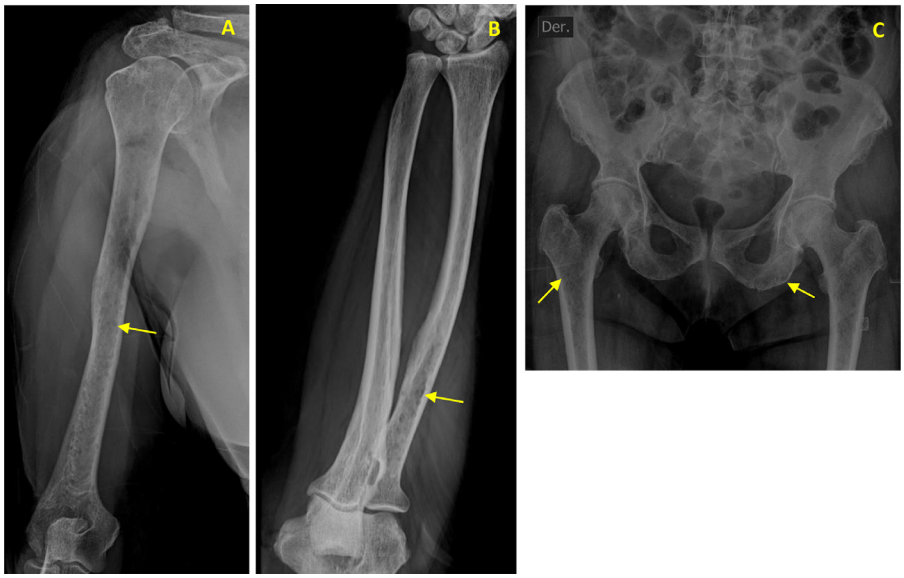


**Fig. 3 – Large bones radiographs with multiple lytic lesions (arrow): right shoulder and humerus (A), right forearm (B) and pelvis (C).**

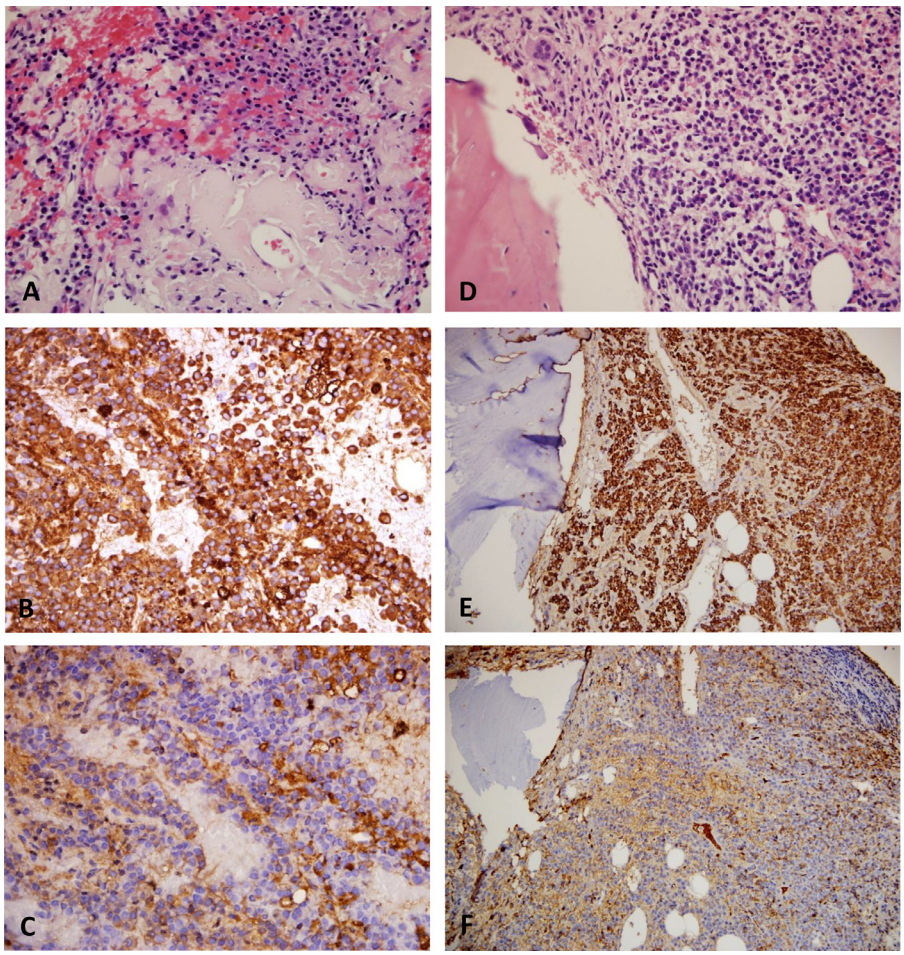


**Fig. 4 – Forehead mass biopsy (left) compared to bone marrow biopsy (right): Diffuse plasma cells infiltrate is observed (A, D) that are monoclonal for the expression of the Kappa light chain (B, E) and negative for the expression of the Lambda light chain (C, F). In bone marrow plasma cells correspond to approximately 80% of cellularity.**

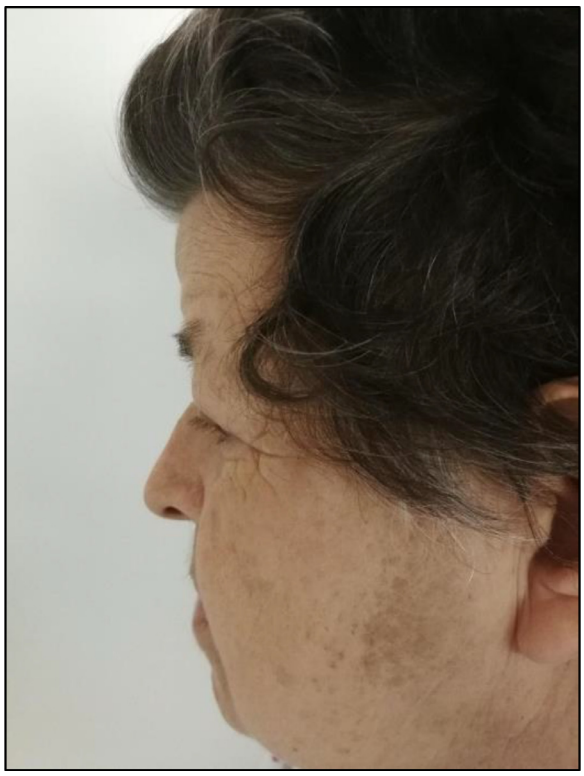

**Fig. 5 – Remarkable reduction of forehead mass one week later after CyBord treatment.**

## REFERENCES


[1] Rajkumar SV, Dimopoulos MA, Palumbo A, Blade J, Merlini G, Mateos MV, et al. International Myeloma Working Group updated criteria for the diagnosis of multiple myeloma. Lancet Oncol [Internet] 2014;15(12):e538–48. Available from: http://dx.doi.org/10.1016/S1470-2045(14)70442-5.

[2] Kyle RA, Gertz MA, Witzig TE, Lust JA, Lacy MQ, Dispenzieri A, et al. Review of 1027 patients with newly diagnosed multiple myeloma. Mayo Clin Proc 2003;78(1):21–33.

[3] Jiménez R, Rosinol LCM. Incidence and outcome of soft-tissue plasmacytomas in patients with multiple myeloma before and after the introduction of novel drugs. Blood [Internet] 2017:3140. Available from:http://www.bloodjournal.org/content/130/Suppl_1/3140.

[4] Short KD, Rajkumar S V, Larson D, Buadi F, Hayman S, Dispenzieri A, et al. Incidence of extramedullary disease in patients with multiple myeloma in the era of novel therapy, and the activity of pomalidomide on extramedullary myeloma. Leukemia 2011;25(6):906–8.

[5] Rajkumar SV. Multiple myeloma: 2020 update on diagnosis, risk-stratification and management. Am J Hematol. 2020;95(5):548–67.

[6] Cerase A, Tarantino A, Gozzetti A, Muccio CF, Gennari P, Monti L, et al. Intracranial involvement in plasmacytomas and multiple myeloma: a pictorial essay. Neuroradiology 2008;50(8):665–74.

[7] Carratalá RM, Cabezuelo MEC, Herrera IH, Azabarte PC, Tapias SD, Presa RMD La, et al. Nontraumatic lesions of the scalp: practical approach to imaging diagnosis. Radiographics 2017;37(3):999–1000.

[8] Leung LK. Differential diagnosis of soft scalp lumps. BMJ Case Rep 2011:4–7.