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Solitary Bone Plasmacytoma in Skull, with Regards to a Case

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Summary

Plasmocytoma is a rare malignant tumor of differentiated B cells in the bone marrow. It is a solitary lesion without clinical, histological or radiological evidence of multiple myeloma. However, it tends to spread years later and turn into a multiple myeloma in 50% of patients.

The diagnosis is based on the radiological findings of a solitary bone lesion, plasma cells in the biopsy, < 5% of plasma cells in the bone marrow, < 2.0 g/dL of monoclonal protein (Protein M) in the serum when it is present, a negative urinary test for Bence Jones protein, without hyperglobulinemia and hyperkalemia data and absence of anemia.

We are presenting the case of a 57-year-old patient, who was apparently in good health and without comorbidities, but who later received a direct trauma to the occipital region, and then a lesion was incidentally discovered whose volume kept progressively increasing in the cranial occipital region.

Keywords: Solitary bone plasmacytoma; Myeloma; Occipital; Plasmacytoma

Introduction

Plasmocytoma is a rare malignant tumor of differentiated B cells in the bone marrow [1-4]. It represents 1% of all types of malignant tumors and a little more than 3-5% of plasma cell neoplasms. A plasma cell neoplasia refers to a monoclonal proliferation of B-cell lymphocytes. They are divided into three types. Solitary bone plasmacytoma, extramedullary plasmacytoma and its disseminated form also called multiple myeloma [1]. Primary cranioencephalic plasmacytoma is very rare and it may be a plasmacytoma of the skull (bone form) or a dural plasmacytoma (non-bone form) [3]. Solitary bone plasmacytoma tends to spread years later and turn into a multiple myeloma in 50% of patients [5]. The prognosis for solitary plasmacytoma of the cranial vault appears to be good when it is diagnosed with strict criteria, which is based on a radiologically solitary bone lesion, neoplastic plasma cells in the biopsy sample, < 5% plasma cells in the bone marrow, < 2.0 g/dl of monoclonal protein in the serum when present and negative urine test for Bence Jones protein (monoclonal light chain) [4,6].

Clinical Case

A 57-year-old male patient is being treated, with no relevant clinical record for his current condition. He began his clinical picture 6 months before his diagnosis, characterized by a fall from his own height and a direct contusion to the occipital region, after which a head trauma to a holocranial region followed, with pulsatile characteristics, which was exacerbated by movement, along with a feeling of nausea and it went into remission through rest and the use of pain medication. As the days went by, it kept increasing in subgaleal volume, in the contusion location (external occipital protrusion). There were no other symptoms.

The patient went in for his check-up at which point the lesion's volume had reached considerable dimensions (See below). In order to study it, a biopsy with fine needle aspiration was suggested, which was performed without success due to a hemorrhage at the puncture site, so the patient was sent to a specialized consultation. From a physical examination, a tumor in the occipital region was revealed with approximate dimensions of 15x7x3 cm, which had a soft consistency, regular edges, non-mobile pulsatile and with heartbeat transmission at the auscultation. The patient did not exhibit a neurological deficit. A multiplanar simple CT scan was performed with axial cuts which reported occipital irregular bone defect with a diameter greater than 15 cm. And extra axial lesion of regular edges of heterogeneous characteristics that displaces the underlying structures. Selective

angiography of the left external carotid artery was performed, whereby occipital branch involvement was evidenced, and it was embolized with platinum micro coils 1 coil axium 3D18 and 1 Coil Axium 3D 10x20 (Figure1).

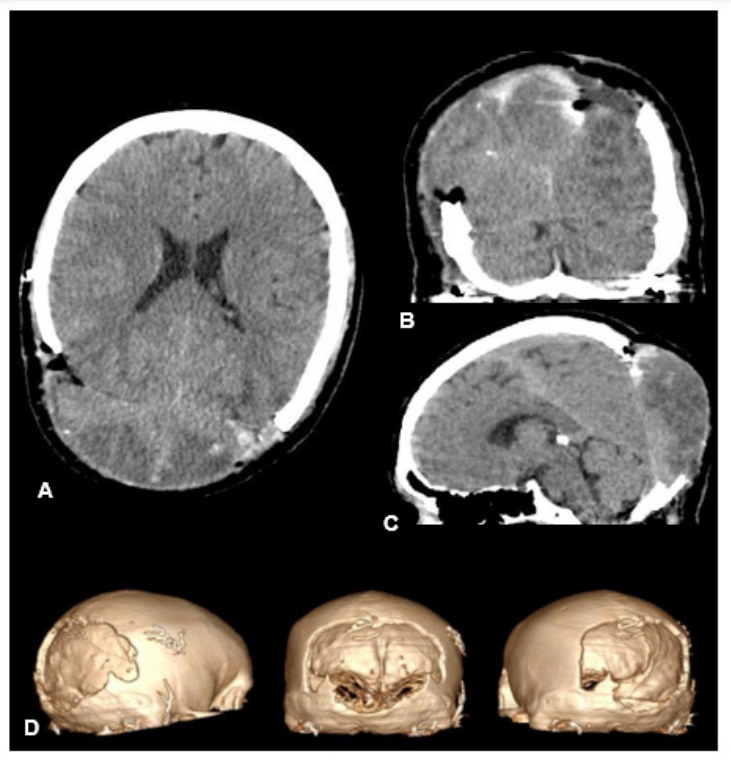

Figure 1: Simple CT scan of skull A, B and C Axial, Coronal and Sagittal sections, respectively, where there is evidence of a solution in the continuity of the bone table in the parieto-occipital region, lesion of extra-axial heterogeneous characteristics, with a dural interface and flattening of underlying structures. D. 3D reconstruction of the bone defect.

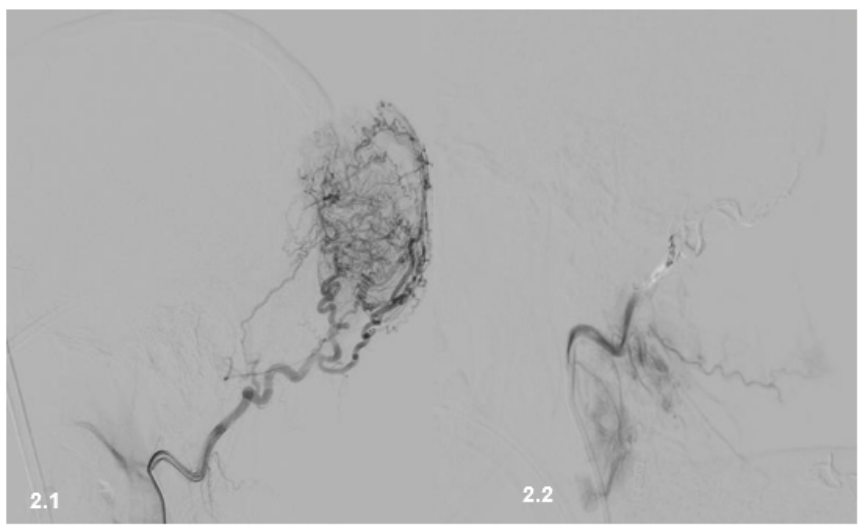

Figure 2: Selective arteriography: 2.1. Right external carotid artery (RECA) in its occipital branch nourishes most of the fistulous tract. 2.2 RECA post-embolization and absence of flow.

External right carotid artery in its occipital branch nourishes most of the fistulous tract (See Figure 2 2.1), so it was embolized with 1 Coil Axium 3D 10x20, 1 Coil Axium Helix 12x12, whereby permeability was observed thus administering 1 Onyx Vial and

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completely occluding such involvement. (See Figure 2 2.2) Also observed was the involvement of distal branches of the Middle Meningeal Artery and branches of the posterior Meningeal Artery, which were also embolized with Onyx. After completing the embolization procedure, the patient was surgically operated 48 hours later: With the patient in a prone position and with the cranial fixation system in three points, after asepsis and antisepsis protocol of the region and placement of sterile fields, a horseshoe-shaped incision with occipital base was made, cut by planes, retracting the skin flap in the previous direction and fixing to the fields. Perilesional subperiosteal dissection was performed until the edges of the cranial defect and tumor in the central region were identified (See Figure 3 3.1). Microsurgical dissection and resection with bipolar electrocoagulation were performed until the lesion was completely removed (See Figure 3 3.2); which had a firm consistency, regular edges, highly vascularized grayish color, in addition to thrombosed vessels in the periphery (previous embolization). There was no meningeal compromise observed. An epidural drainage system was placed and closed in a conventional manner. The sample was sent to the Pathology Department which reported; Lesion made up of several types of Cells of different sizes, but the great majority of them (approximately 80% of plasma cells) were well moderated and a little differentiated, which showed extensive zones of necrosis, and sporadic calcifications between the tumor cells. Mitosis was scarce but the pleomorphism was very prominent, which gave it an unpredictable prognosis. The characteristics of this tumor correspond to a poorly differentiated plasma cell neoplasm (See Figure 4).

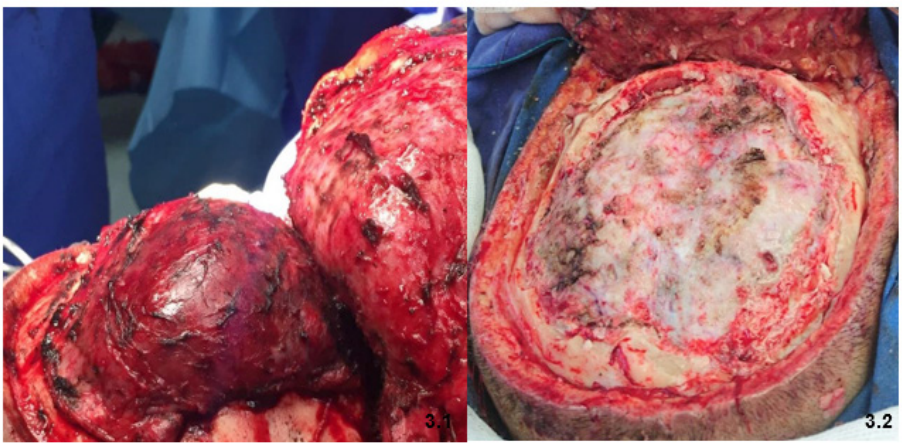

Figure 3: Intraoperative image: 3.1. Occipital peritorcular lesion with a greater diameter of 15 cm, firm consistency, regular edges, and heartbeat transmission. 3.2. Complete resection of the lesion with irregular osseous defect secondary to lytic reaction, without dura mater involvement.

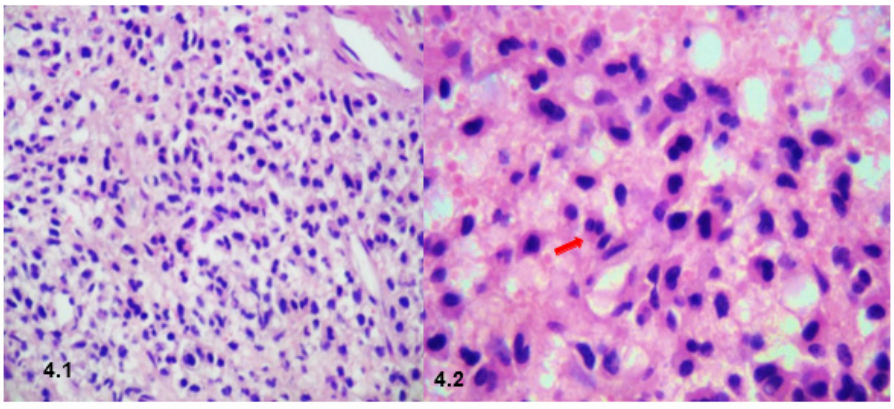

Figure 4: Microscopic photographs of the tumor 4.1 (H&E, x 40). Cells of different sizes (80% plasma cells) well moderated and poorly differentiated. Mitoses are scarce and pleomorphism is abundant. 4.2 (H&E, x 200). Binucleated plasma cells with eccentric nuclei, prominent nucleoli, a perinuclear halo and abundant cytoplasm (red arrow).

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Discussion

Multiple myeloma is a non-Hodgkin's low-grade B-cell lymphoma, characterized by a monoclonal proliferation of malignant plasma cells. It is a disease that usually originates in the bone marrow and eventually spreads to soft tissues or peripheral blood (plasma cell leukemia). The main causes and effects of the pain and disability are the replacement of the hematopoietic bone marrow (leading to anemia, leukopenia and thrombocytopenia and its sequelae), osteoporosis and bone destruction (leading to fractures and pain), kidney damage by paraproteins and systemic amyloidosis [7].

A solitary plasmacytoma is a single lesion without clinical, histological or radiological evidence of multiple myeloma [8]. Plasmacytomas are malignant bone tumors, which result from the proliferation of monoclonal plasma cells and are characterized by the secretion of immunoglobulin G and A (known as M protein) [9]. Plasmacytomas may be divided into multiple, solitary bone and solitary extraosseous or extramedullary plasmacytomas and are rare compared to multiple myeloma8. Solitary intracranial plasmacytomas are extremely rare [9]. The incidence of this disease has been reported in approximately 3/10 000 000 cases per year8. In comparison with a solitary plasmocytoma of the skull base, the prognosis for a solitary plasmacytoma of the cranial vault seems to be good, when there is no presence of multiple myelomatosis [6]. The diagnosis is based on the radiological findings of a solitary bone lesion, plasma cells in the biopsy, less than 5% of plasma cells in the bone marrow, less than 2.0 g/dL of monoclonal protein (Protein M) in the serum when present, a negative urinary test for Bence Jones protein, without hyperglobulinemia and hyperkalemia data and absence of anemia [8] (See below). In the case of our patient, all the criteria were corroborated to verify that we were in fact dealing with a solitary bone plasmacytoma. However, evidently it was not initially suspected in this diagnosis, therefore the studies were performed in the immediate postoperative period. The diagnosis of myeloma in the bone also needs to be differentiated from the eosinophilic granuloma, osteosarcoma and metastatic carcinoma [4]. Eosinophilic granuloma is mainly observed in children and young people, and they are mostly small simple lesions with well-defined borders. Osteosarcoma is a malignant primary bone tumor that is found mainly in long bones and it is very rarely found in the skull. Osteolytic metastatic carcinoma is the most common type of bone metastasis and is usually multiple. The limits of the metastases are not well defined, and a soft tissue mass may be adjacent to it. Meningiomas may cause a high-density peripheral osteoblastic reaction with homogeneous enhancement and have a dural tail [10-16].

Metastases due to solid tumors are much more common than multiple myeloma and may be difficult to discriminate. The characteristics in favor of multiple myeloma are:

a. Osteolytic lesions in the convexity of the skull and the diaphysis of long bones.

b. Non-reactive lytic lesions clearly outlined.

c. Scalloped (crescent-shaped erosions of cortical bone from its surface).

d. Marked osteoporosis.

e. Negative bone scan or cold lesions.

f. There is no primary tumor as a possible cause of bone metastases [7].

CT shows an osteolytic lesion without sclerotic borders, a hyperdense and homogeneous tumor that improves with the administration of the contrast medium6. However, neuroradiological findings generally lack specificity, since they are generally not different from meningioma, plasma cell granuloma, metastasis, lymphoma, dural sarcoma, infectious meningitis, and leptomeningeal carcinomatosis [10].

The international working group on Myeloma proposed new criteria for the diagnosis and classification of myeloma according to the available routine exams. According to the criteria, symptomatic myeloma requires the presence of M protein in the serum and urine, plasmacytosis in the bone marrow and damage related to the final organ. The criteria for asymptomatic or latent myeloma are M protein levels ≥30 g/l and/or ≥10% of bone marrow clonal cells, but there is no related impairment of organs or tissues. Cases with related deterioration of organs or tissues usually show elevated calcium levels, renal failure, anemia or bone lesions, which are attributed to the proliferation of plasma cells [4]. Histological staining with hematoxylin and eosin reveals a proliferation of atypical plasma cells, which usually show scattered chromatin, prominent nucleoli and a high cytoplasm nucleus relationship. In addition, malignant plasma cells may contain condensed or crystallized immunoglobulin resulting in multiple pale bluish white, grape-like accumulations, cherry-red refractive round bodies, vermillion staining glycogen-rich, overstuffed fibrils and crystalline rods, which identify specific cellular patterns [8]. Approximately 50% of plasmacytomas turn into multiple myeloma in 10 years of follow-up, and 10% of them return to a plasmacytoma. Recent studies using in-situ in-fluorescence hybridization indicate that all multiple myeloma cells harbor chromosomal abnormalities; a monosomy of chromosome 13, which occurs in 85% of all multiple myelomas, is associated with an adverse prognosis [2]. Due to its rarity, the literature on the treatment of cranial plasmacytoma is limited [11]. The consensus at this time suggests that for solitary plasmacytoma the treatment is gross total resection with adjuvant radiotherapy [11-16].

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Conclusion

Solitary bone plasmacytoma of the skull represents an extremely rare pathology, whose result is histopathological, and it is based on strict laboratory criteria. It is very important to differentiate it from multiple myeloma, given that its treatment and prognosis are usually very different; however, the follow up must be performed in the long run due to the potential risk of it turning into a systemic myelomatosis. The treatment of solitary plasmacytoma is full surgical removal of the tumor followed by postoperative radiotherapy.

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