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PLASMACYTOMA OF THE SKULL MIMICKING A MENINGIOMATWO CASE REPORTS AND REVIEW OF THE LITERATURE

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ABSTRACT

Cranial plasmacytoma is a rare tumor that can be isolated or part of a multiple myeloma context. The differential diagnosis between plasmacytoma and meningioma is often challenging, as both can present radiological similarities. In this article, we present two cases of cranial plasmacytoma in women with a cranial mass initially diagnosed as meningioma based on imaging. In the first case, the plasmacytoma was isolated, without progression to multiple myeloma, while the second case revealed multiple myeloma. Both patients underwent surgical excision. This work emphasizes the importance of performing a preoperative assessment, including a biopsy, especially in cases of suspected meningioma, to avoid missing a cranial plasmacytoma or even multiple myeloma.

KEYWORDS: skull plasmacytoma, meningioma, multiple myeloma, biopsy, preoperative assessment.

INTRODUCTION

Plasmacytoma is a tumor consisting of a monoclonal proliferation of malignant plasma cells, whether or not it is part of multiple myeloma.[1]

The location of plasmacytoma in the skull is rare.[1]

The therapeutic management of solitary plasmacytoma is primarily based on radiotherapy. However, when it is part of multiple myeloma, treatment is based on polychemotherapy.[2]

We report two cases of plasmacytoma of the skull in two women presenting with a painful cranial mass, where the initial diagnosis of meningioma was made based on imaging.

OBSERVATIONS

Case 1

A 57-year-old diabetic woman presented with painful cranial swelling in the right parietal region, the volume of which had been gradually increasing over the past seven months. She showed neither signs of intracranial hypertension nor sensory-motor deficit. The progression of her illness led the patient to consult a neurologist in December 2019.

A brain MRI revealed an extra-axial expansive process located in the right frontoparietal region, manifested by an isosignal concerning the white matter on the different sequences.

This lesion took on strong contrast, with regular contours and clear boundaries, measuring 40 mm in the anteroposterior direction and 36 mm transversely.

The lesion suppressed the underlying brain parenchyma without invasion but invaded the skull bone, suggesting a meningioma.

The patient underwent surgery with complete excision of the cranial vault mass and its intracranial extension, followed by cranioplasty.

Pathological examination revealed the presence of a proliferation of round cells, often large, showing plasmacytoid differentiation.

These cells showed large, sometimes nucleolated nuclei and more or less abundant eosinophilic cytoplasm. These cells were arranged in diffuse areas with infiltration of bone tissue.

This aspect was in favor of a plasmacytoma. These cells were immunoreactive to CD138, indicating the presence of a plasmacytoma.

An assessment to look for multiple myeloma was carried out, without signs of anemia, disturbance of serum calcium, or renal dysfunction. Serum protein

electrophoresis, combined with immunofixation, showed a monoclonal IgG Kappa peak at 1.5 g/dL. Bence-Jones protein was negative, while the light chain ratio was within the normal range. The myelogram revealed a proportion of plasma cells less than 10%. Whole-body MRI and PET scan showed no myeloma infiltration.

The patient has been in remission for more than four years following surgical excision, with regular follow-up.

Case 2

A 66-year-old woman consulted in 2020 for a painful right temporal swelling which had been gradually increasing in size for several months, without signs of intracranial hypertension or sensory-motor deficit, and with a preserved general condition.

An X-ray of the skull revealed a bone gap image next to the right temporal skin swelling, justifying additional CT scanning.

The brain scan showed a spontaneously dense tissue formation, extra-axial right parietal, massively enhanced after contrast injection, with regular contours, measuring 65 mm by 45 mm, accompanied by lysis of the bony vault with extra-axial extension cranial, suggestive of a meningioma of the right parietal vault Fig 1.

The patient underwent a large lumpectomy with cranioplasty.

Histopathological analysis showed tumor proliferation in sheets of monomorphic cells with a plasmacytoid appearance, with eosinophilic cytoplasms and peripheral nuclei Fig 2. Immunohistochemistry confirmed a plasmacytoma with CD138 immunoreactive cells Fig 3.

A work-up to look for multiple myeloma showed an absence of anemia, normal serum calcium, normal renal function, a monoclonal peak of IgG Kappa gamma globulins quantified at 44.2 g/L, Kappa light chains at 13.72 µg/dL, and Lambda at 4.14 µg/dL with a Kappa/Lambda ratio of 3.13 and a positive Bence-Jones protein.

The myelogram revealed 20% clonal plasma cells. A PET scan revealed lytic lesions in the right rib K8 and left femoral head and hilar lymph node hypermetabolism of non-specific appearance.

The diagnosis of IgG Kappa multiple myeloma was confirmed. Initially, the patient received VCD (bortezomib, cyclophosphamide, dexamethasone), achieving a very good partial response after six cycles, and was subsequently placed under surveillance.

Two years later, she experienced both clinical and biochemical relapse. Despite being treated with DKD (daratumumab, carfilzomib, dexamethasone), her disease progressed, and she ultimately passed away.

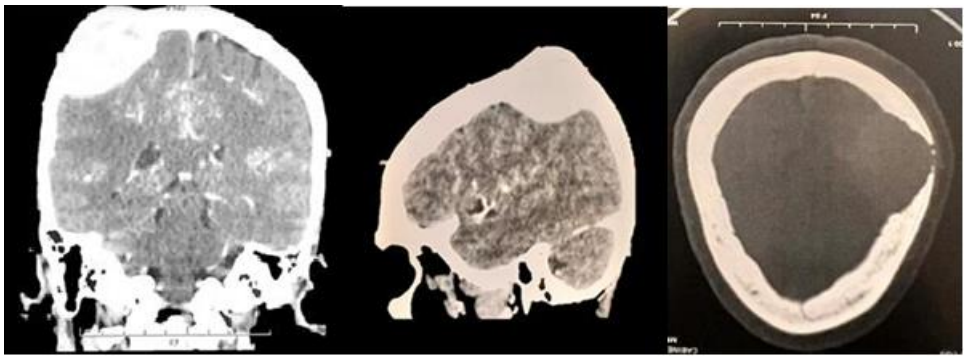

Fig 1: CT images of plasmacytoma of the right parietal vault mimicking a meningioma.

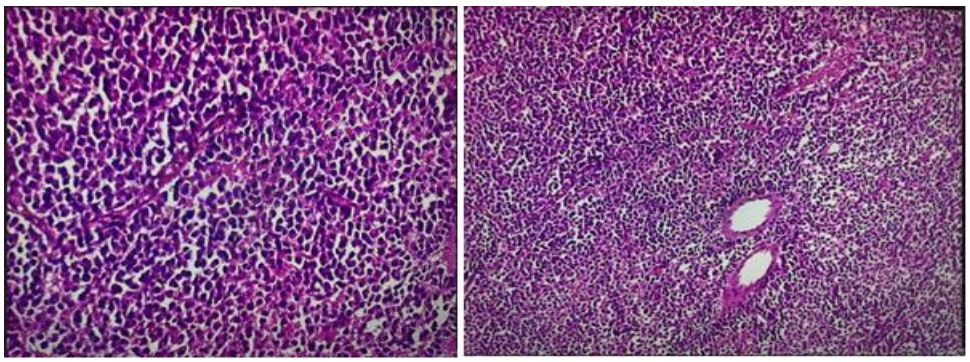

Fig 2: Pathological examination of the surgical specimen showing a sheet of plasma cells.

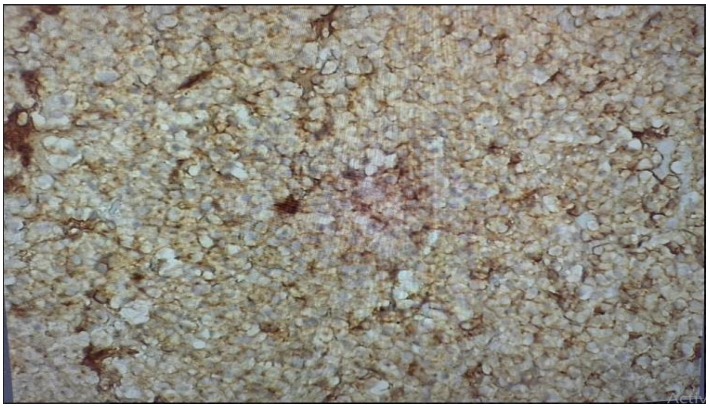

Fig 3: Immunohistochemical staining of CD38.

DISCUSSION

Plasmacytoma is a malignant tumor characterized by the proliferation of monoclonal plasma cells, which can be solitary or indicative of multiple myeloma.[3] The diagnosis of solitary plasmacytoma is based on several criteria: histological confirmation of the lesion's uniqueness, absence of radiological damage, and absence of signs of marrow invasion.[4,5]

The average age at diagnosis is between 40 and 50 years, approximately ten years less than for multiple myeloma, with a male predominance.[4] In our series, the average age of our patients is 61.5 years (range 57-66 years), slightly higher due to the late discovery of multiple myeloma.

Solitary cranial plasmacytoma is a rare tumor that develops in the dura mater, skull bones, or,more rarely, in the brain. Representing only 0.7% of all solitary plasmacytomas, it most oftenoccurs as a manifestation of multiple myeloma6. Involvement of the cranial vault is very rare, mainly affecting the base of the skull.[6,7]

Until 2001, to our knowledge, 44 cases of solitary intracranial plasmacytomas were reportedin the Anglo-Saxon literature.[6] These plasmacytomas frequently affect the parietal bone, while the involvement of the frontal bone is less common or even very rare.[6,7] Solitary plasmacytoma located at the vault can be indicative of multiple myeloma, but it does not transform into multiple myeloma, whereas intracranial plasmacytomas of the vault and base of the skull are much more likely to progress to multiple myeloma.[3,6,8] Hence the systematic monitoring of solitary intracranial plasmacytoma in search of multiple myeloma, especially if it is a plasmacytoma of the base of the skull, as demonstrated by the histopathological study carriedout by Schwartz et al.[9]

The radiological appearance of plasmacytoma shows a well-defined bone gap, without peripheral sclerosis on standard radiography. CT scan reveals a lytic lesion of the vault, which can be lobulated, without peripheral sclerosis, and it is a homogeneous lesion that takes contrast markedly. MRI is the test of choice for exploring intracranial plasmacytomas. Plasmacytoma appears on T1 isosignal MRI with clear enhancement after gadolinium injection, allowing the specification of extraosseous tumor extensions, particularly intracranially, as well as the relationship with the underlying brain parenchyma.[10,11] Cerebral angiography, if performed, shows a vascular blush.[12]

Both cases exhibited typical radiological characteristics of plasmacytomas, including well-defined osteolytic lesions without associated bone sclerosis. CT and MRI scans revealed lesions with significant contrast enhancement, demonstrating sharp margins and extensions into both bone and surrounding soft tissues. These radiological findings are consistent with the classical descriptions of plasmacytomas in the literature, highlighting their tendency for clear contrast uptake and expansive growth patterns involving osseous and extraosseous structures.

There is no pathognomonic radiological sign for the diagnosis of plasmacytoma. It can be confused with other lytic lesions of the vault, particularly meningioma, which presents similar radiological characteristics: an extra-axial mass with a broad base on the dura mater, hyperdense on CT, isosignal on T1-weighted MRI, and variable signal intensity on T2 with strong enhancement after contrast injection.[12,13,14] The diagnosis is made by histological examination during a biopsy.

Meningioma is a generally benign tumor and the most common tumor of the central nervous system and its coverings, representing over a third of all such tumors. Intracranial meningiomas can secondarily invade the cranial vault. Primary intraosseous meningioma is rare, representing only 1% of meningiomas, called osteomeningioma. However, the term osteomeningioma is often used to describe hyperostoses reactive to an underlying meningioma.[15]

A predominantly dural-based cranial plasmacytoma

mimicking a meningioma has been well described in the literature, usually involving the leptomeninges and skull.[16,17]

Due to their rarity, the literature on the management of cranial plasmacytomas is limited. Neurosurgeons tend to plan surgical resection without further evaluation of the lesion. Plasmacytomas are one of the few tumors that are radiosensitive and often do not require surgical reduction unless the lesion is compressing vital structures. If correctly diagnosed, craniotomy with resection of the intracranial mass can be avoided.[18]

In cases of plasmacytoma associated with multiple myeloma, most authors recommend a combination of chemotherapy and radiotherapy as the initial treatment for these lesions, even in the presence of a mass effect exerted by the tumor. This approach is favored due to the high sensitivity of these lesions to these treatments.[19,20]

However, surgery is often reserved for patients who are refractory to these treatments or who cannot undergo radiotherapy due to the proximity of sensitive structures, such as the optic chiasm.[9]

Our two cases represent the two previously described scenarios: a solitary plasmacytoma in the first case and a plasmacytoma that turned out to be a manifestation of multiple myeloma in the second case.

Given that solitary plasmacytoma of the skull can successfully mimic meningioma on imaging and that intraosseous meningiomas are rare, benign ectopic tumors with progressive evolution, and their definitive diagnosis is histological[21], we recommend conducting a preoperative assessment for multiple myeloma or even performing a biopsy before making a therapeutic decision. This approach can avoid potentially unnecessary major surgery when plasmacytoma is in the differential diagnosis.

CONCLUSION

Our work highlights the benefit of performing a preoperative assessment, which may include a biopsy in the case of a meningioma, to avoid missing a skull plasmacytoma or even multiple myeloma.

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