Mediterranean BioMedical Journals
International Journal of Medicine and Surgery
Volume 3, Issue 2, 2016
DOI: 10.15342/ijms.v3i2.103

CLINICAL CASE

Multiple Myeloma Revealed by a Sphenoid Plasmocytoma A Case Report

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Received 15 September 2016; Revised 19 December 2016; Accepted 24 December 2016.

ABSTRACT
Plasma cell neoplasms can manifest as a solitary or multiple plasmocytomas and may be associated with or progressing to multiple myeloma (MM). Cranial and intracranial plasmocytomas revealing multiple myeloma are very rare and only few cases are reported in the literature.
We report the case of a sphenoid plasmocytoma that revealed a multiple myeloma in a 56 year-old woman with 3 months history of temporal headache and diplopia. Magnetic resonance imaging (MRI) and computed tomography (CT) showed a sphenoid mass. An endoscopic sphenoidal biopsy was performed and the histopathological exams showed a plasmocytoma with a positive staining for CD138. Further biological studies confirmed the diagnosis of multiple myeloma with a monoclonal gamma peak of immunoglobulin (Ig) A. The patient started systemic chemotherapy and received decompressive radiation therapy on the sphenoidal sinus. She remained in remission for 8 months and died from renal dysfunction.
Although the sphenoid plasmocytoma is a very rare presentation of multiple myeloma it should be considered for effective patient management and prognosis improvement.

KEY WORDS: Multiple Myeloma; Plasmocytoma; Sphenoid; Skull Base.

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INTRODUCTION
Multiple Myeloma (MM) is a malignant neoplastic disorder arising from bone marrow and was described for the first time by MacIntyre in 1850. [1] It is characterized by plasma cell tumors that are producing immunoglobulins and/or light chains.[2] The presentation of MM are unspecific and various clinical features can occur, such as bone pain, weakness, fatigue, fever and infection. It can be associated with plasmocytoma at the diagnosis or more frequently developed subsequently in 50 % of the cases. [1]
Plasmocytomas may be solitary or multiple, arising from osseous or non osseous sites. When involving the skull base, plasmocytoma could produce pain, epistaxis, rhinorhea, nasal obstruction and cranial nerve palsies. [3] Cranial localization of plasmocytoma is very rare especially when associated to MM at diagnosis. We present the case of a 56 year-old woman with a MM revealed by sphenoid plasmocytoma. Only few cases in the literature reported intracranial plasmocytoma revealing MM. Our case is another one showing clinicopathological characteristics and therapeutic outcomes of this specific association.

CASES REPORT
A 56 year-old woman complained for three months of temporal headache and diplopia.
She consulted an ophthalmologist and her physical examination showed a palsy of the left sixth cranial nerve responsible of diplopia; without any other clinical findings. A Facial CT and MRI (figure 1, 2) showed an expansive voluminous mass: 50 mm*38 mm in the sphenoid eroding bone anteriorly and extending to the left temporal region and the left cavernous sinus.

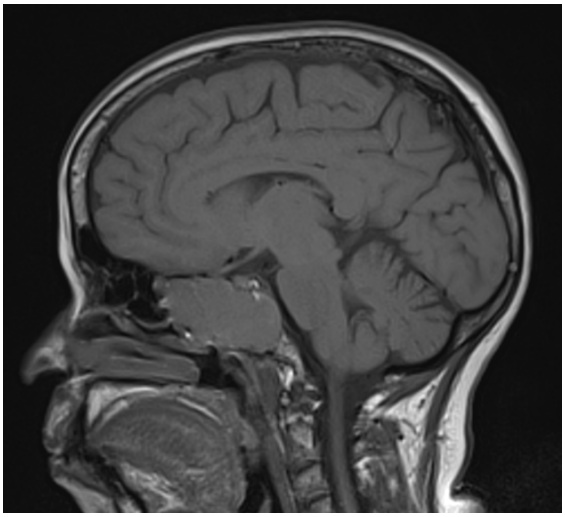

Figure 1: T1 weighted sagital view of a cranial MRI showing a sphenoid extending tumor

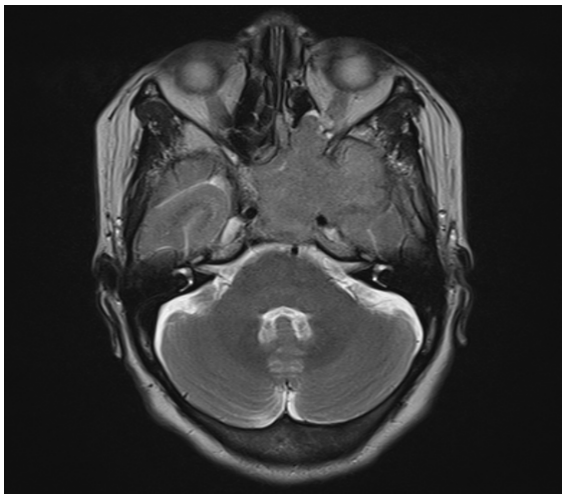

Figure 2: T2 weighted axial view of a brain MRI showing the sphenoid tumor extending on the left to the temporal area

The sphenoidal endoscopic biopsy under general anesthesia revealed microscopically, a monotonous proliferation of atypical plasma cells with hyperchromatic nuclei (figure 3) staining positively for CD138 confirming the diagnosis of plasmocytoma (figure 4).

Laboratory test revealed a mild normocytic anemia with a hemoglobin level of 11.5 g/dl. Blood chemistry analyses were normal. Serum protein electrophoresis detected a monoclonal gamma peak with Ig A kappa on serum immunofluorescence electrophoresis. Urine was negative for Bence–Jones proteinuria. Beta 2 microglobulin level was 4 mg/dl.

A bone narrow aspiration showed more than 15% of plasma cells sometimes in clusters with some multinucleated cells and plasmablasts . The diagnosis of multiple myeloma stage II of Salmon and Durie, with the involvement of the sphenoid was made.

Immediately a systemic chemotherapy based on dexamethazon and zoledronic acid was initiated and a decompressive radiotherapy was given, 40 Gy in 2Gy per fraction by 2 opposed lateral fields in 28 days. The patient was going to continue the systemic therapy but her condition deteriorated and she died from renal failure 8 months after her admission.

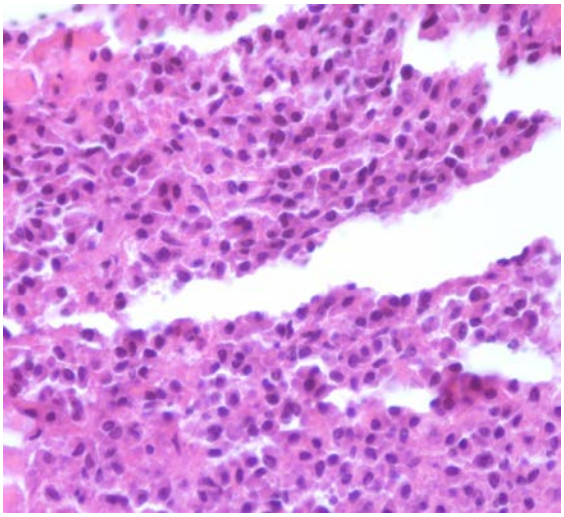

Figure3: microphotography showing round tumor cells with eccentrically located nuclei. The nuclei show hyperchromasia. (Hemalun eosin , objective ×40).

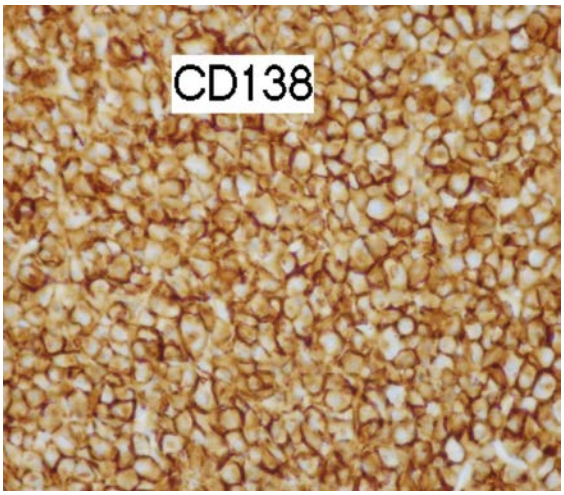

Figure 4: high power view of the tumor showing strong staining for CD138.

DISCUSSION

MM is a bone marrow serous malignancy occurring in the elderly, more frequent in men and usually affecting the axial skeleton [2, 4]. It is a malignant disease characterized by monoclonal protein in serum and/or urine due to plasma cells, lytic lesions in bone, renal failure, anemia, susceptibility to infections and hypercalcemia. It is rarely revealed by a plasmocytoma as in our case [5]. When involving the skull base as in our observation, plasma cells neoplasms could be asymptomatic. If it's a voluminous mass it may result in tumefaction, pain, headache, single or multiple cranial nerve palsies, or seizure if intracranial extension [6, 7]. In our case the patient had a diplopia caused by the left sixth nerve palsy by compression and temporal headache.

Well-defined destructive masses arising from osseous structures are easily shown by CT and MRI. Even if CT is better than MRI in delineating bone erosions, MRI has the best assessment because of its superiority in the study of both bone marrow and meninges. [1]

Plasma cell neoplasm is highly radiosensitive. When it is a manifestation of MM as in our case, the treatment of

choice is combination of irradiation and systemic chemotherapy followed by stem cell transplant.[8]
Skull base plasmocytomas are treated by subtotal resection followed by irradiation, or irradiation alone. [9] Radiosurgery could be used when the tumor volume is suitable. [10]
Before radiotherapy a neurosurgical opinion should be sought and during irradiation the patient should receive corticosteroid to avoid acute complications due to inflammation.
Disease stage at diagnosis, and laboratory prognostic factors have impact on survival of patients with MM. Despite aggressive systemic and local therapy, the survival of patients with intracranial MM ranges from 1 to 96 months after diagnosis. [1]
Unfortunately our case confirmed the poor outcomes and prognosis of the association of MM and an expansible plasmocytoma of the skull base and showed the importance of an early diagnosis for effective management of patients.

CONCLUSION

The MM associated to sphenoid plasmocytoma with intracranial involvement leads to a fatal outcome. Because cranial nerve involvement and headache are unusual in plasma cell neoplasms; diagnosis and treatment were delayed. In the context of anaemia and lytic bony lesion as in our case; MM should always be rapidly considered.

LIST OF ABBREVIATIONS

MM Multiple Myeloma
MRI Magnetic Resonance Imaging
CT Computed Tomography

ACKNOWLEDGEMENT

Declared none.

PATIENT CONSENT

Written informed consent was obtained from the patient for publication of this case report.

COMPETING INTERESTS

The authors declare no competing interests.

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