International Journal of Academic Health and Medical Research (IJAHMR)
ISSN: 2643-9824
Vol. 8 Issue 5 May - 2024, Pages: 41-44

Malar Bone Plasmocytoma : Case Report

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The authors declare no conflict of interest

Abstract : *The plasmacytoma tumoral process described as bone infiltration by a malignant plasmocyte line cells in the form of a solitary mass that may be located in the bone marrow or extramedullary. Initial symptoms are vague and nonspecific. In view of the rarity of the condition and the lack of data in the literature, we have chosen to focus on this theme. Our case is a 57-year-old female patient who presented left jugal mass with firm consistency evolving for two years. The patient's medical history has no specific features. A CT scan of the facial mass, which revealed a lesional process in the right malar bone. A biopsy was performed; immunohistochemistry was in favor of a plasmacytoma with Monotype kappa. The patient started chemotherapy.*

INTRODUCTION:

The plasmacytoma is a neoplastic and monoclonal proliferation of plasma cells that usually arises within bones. These are relatively rare malignant tumors of the head and neck region. The incidence of these tumors is around 2.6-3.3/100,000 populations. (1). they are characterized by a monoclonal neoplastic proliferation of plasma cells and histologically, they are indistinguishable. They can appear as multiple myeloma (MM), solitary plasmacytoma of bone (SPB), or extramedullary plasmacytoma (EMP). (2)

The majority of the lesions occur within the center of a single bone, most often in the spine, vertebrae, femur and pelvis (3) (4).Although SBP represents under 10% of plasma cell dyscrasias, it is a known precursor of multiple myeloma (MM), since more than 50% of SBP patients have undergone systemic dissemination to MM. The presence in the serum of an abnormal immunoglobulin free light chain, or the reappearance of a treated solitary lesion, are signs of tumor progression to MM. (5) (6) (7)

The male/female ration of solitary plasmacytoma is around 2/1, the average age is estimated at 55 years. (8)

Radiotherapy, radical extensive surgery or an association of these modalities represents the primary treatment. (9)

Very little has been reported on plasmacytomas of the oral and maxillofacial region, particularly in the malar bone.

CASE REPORT:

A 57-year-old female patient was referred to our department of oral and maxillofacial surgery of Ibn Sina hospital in Rabat. She presented right jugal mass with firm consistency evolving for two years. The patient's medical history has no specific features.

The physical exam found a right jugal mass without inflammatory signs opposite, painless, firm consistency, mobile in relation to the superficial plane and fixed in relation to the deep plane, measuring approximately 3 cm long, without vestibular expression . Cervical exam reveals no palpable adenopathy. Note that the sensitivity and motor skills of the face are preserved. (figure1)

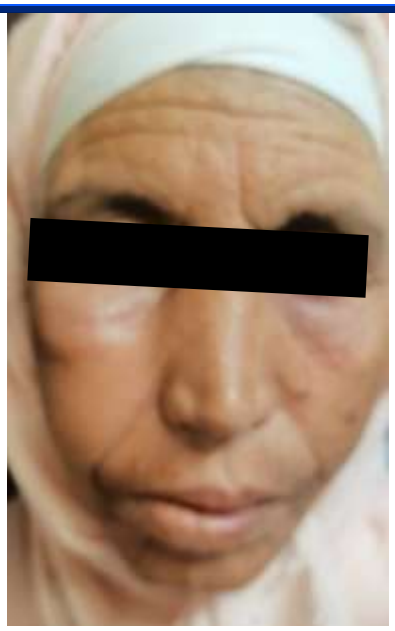
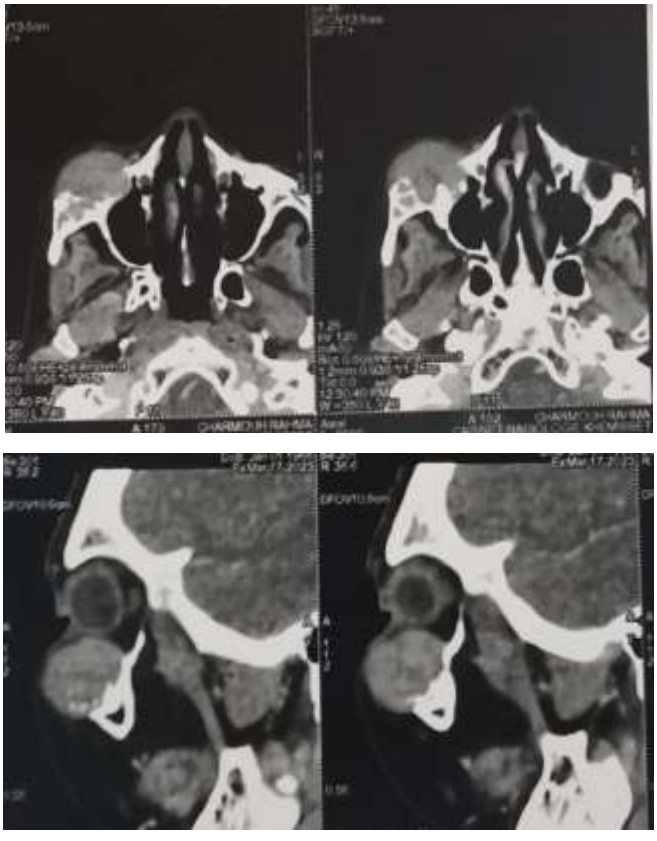


Figure 1: Image showing right jugal mass

The patient underwent a CT scan of the facial mass, which revealed a lesional process in the right malar bone with heterogeneous tissue density after injection of the contrast medium, reaching the floor of the orbit and the anterior wall of the maxillary sinus. (Figure 2)



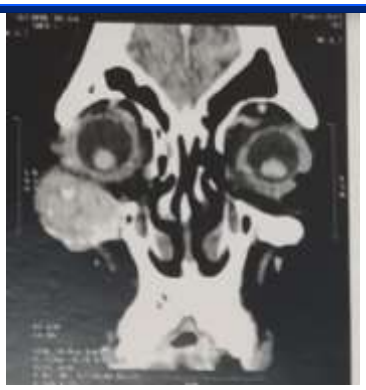


Figure 2: CT scan showing lesional process in the right malar bone.

A biopsy was performed; immunohistochemistry was in favor of a plasmacytoma with Monotype kappa. Given the presence of Bence Jones proteinuria, the patient was referred to the internal medicine department for an extension evaluation, which included an osteomedullary biopsy and a myelogram with a CTAP scan as well as a standard X-ray of the skall in search of geodes. Bone marrow puncture confirmed plasma cell proliferation.1 month later the patient started chemotherapy.

DISCUSSION:

Solitary plasmacytomas are rare affections whose initial symptoms are vague and non-specific, making them difficult to detect. Its etiology and pathogenesis have yet to be elucidated. It rarely occurs in the maxillofacial bones.

The relationship between SBP and MM is still controversial. Most experts consider SBP to be a different clinical disease from MM, as there are major differences in clinical manifestations, course and prognosis and that SBP should be considered an early stage of MM. The average age of onset of SBP is about 10 years earlier than MM. Although a few patients have stable lesions for a long time, many patients (about 10–60%) will eventually progress to MM within 3 years. Progression to MM is the main factor affecting the survival of patients with SBP. (10)

Many specialists recommend local radiotherapy as the optimal treatment for SBP. If bone destruction is severe and involves the stability of the bone structure, surgery and radiotherapy may be considered. (11) (12)

An early diagnosis of a particular clinico-pathological entity is very important for the choice of therapy and survival rate. Differential diagnosis must be carried out with care, as early signs are not very conclusive. (13)

Skeletal radiography is a conventional, routine imaging method for the diagnosis of SBP. Orthopantomography is used for maxillofacial lesions. CT has high density resolution and can show the destruction of fine bone structure earlier than X-rays. CT in combination with various reconstruction techniques can clearly show the mode of destruction, characteristics, degree and extent of bone destruction in the maxillofacial SBP. It can also show the residual bone ridge in the area of bone destruction, proliferation and sclerosis of the surrounding cortex, as well as multiple tumor ruptures through the cortex. Although the above findings can also be visualized by MRI, CT can show bone changes more clearly. CT is therefore an important diagnostic imaging method for maxillofacial SBP. In more recent years, PET/CT has been progressively used to assess the efficacy of SBP therapy and differentiate it from MM. (14)

The detection of a plasma cell infiltrate on histopathology is a guide to the diagnosis of solitary plasmacytoma, but it is not sufficient, as the diagnosis of multiple myeloma must be ruled out by bone marrow aspiration, blood and urine tests, as well as imagistic investigations. To diagnose solitary plasmacytoma, radiological investigations of the entire skeleton must reveal the presence of a single area of bone destruction due to clonal plasma cells, with no other osteolytic lesions or tissue damage. Bone marrow infiltration by plasma cells of less than 5% of all nucleated cells should be revealed by medullograms. Serological and urinary examinations should also show low concentrations of monoclonal protein in serum or urine, and the absence of anemia, hypocalcemia or renal impairment attributed to multiple myeloma. (15)

In a review, V. Ibric Cioranu et al. their case was referred to a hematologist and monitored for over 14 years. The patient was diagnosed with multirecurrent solitary plasmacytoma having multiple localizations. In our case, the patient was referred to the

internal medicine department. After several investigations, joined the Hughes M et al (15). study the diagnosis of multipe myeloma was accepted, and chemotherapy was initiated.

Unlike V. Ibric Cioranu1 et al's case, our patient did not require surgery.

CONCLUSION:

The group of affections that integrate plasma cell neoplasia represent various manifestations. The investigations such as histopathological examination and immunohistochemistry are important. Treatments differ according to the diagnosis of multiple myeloma or SBP.

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