

Maxillary Solitary Recurrent Plasmacytoma: A Case Report

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Rezumat

Plasmocitomul maxilar solitar multiplu: prezentare de caz

Plasmocitomul solitar reprezintă o formă rară de neoplazie. Face parte din grupul gamapatiilor monoclonale. Este o proliferare de celule plasmatiche sub formă de masă solitară medulară sau extramedulară. Simptomatologia de debut este discretă. Fiind o afecțiune rară datele din literatură sunt destul de puține. Foarte important este diagnosticul precoce, deoarece prezintă risc ridicat de transformare în mielom multiplu. Prezentăm cazul unui pacient de 52 ani cu plasmocitom solitar multiplu. Formele tumorale au fost diagnosticate de-a lungul unei perioade de 14 ani. Medulogramele de control nu au arătat o invazie medulară. Inițial a răspuns la chimioterapie, dar tumora a progresat determinând hematologul să recomande rezecția chirurgicală. Localizarea maxilară a tumorii a determinat excizia totală a tumorii cu refacerea planșoului orbitei și păstrarea funcționalității globului ocular. Această evoluție a pacientului vine în sprijinul tratamentului combinat chirurgical și chimio-radioterapic în tratamentul acestei patologii.

Cuvinte cheie: plasmocitom solitar, tumoră maxilar, reabilitare funcțională

Abstract

Solitary plasmacytoma is a very rare form of neoplasia, part of the monoclonal gammopathies. It represents a tumoral proliferation of plasma cells in the form of a solitary mass which can be located in the bone marrow or extramedullary. Initial symptoms are vague and nonspecific. Being such a rare affliction, there is little information in the literature. Early diagnosis is difficult but very important due to therapy outcome. A high risk of progression towards a multiple myeloma has been reported. We present a rare case of a 52-year-old patient diagnosed with multiple solitary plasmacytomas. The tumours were separated from one another in time, over a 14 years period. The various medullograms did not show any sign of medullary plasma cell infiltrate. Initially, the affliction responded to chemotherapy, but later the haematologist recommended surgical resections followed by reconstruction. The maxillary localization required excision of the tumour with the preservation of the eye bulb despite the destruction of the orbital floor and with the regain of ocular functionality as well as aesthetic rehabilitation. This evolution highlights the benefits of surgical treatment in conjunction with chemotherapy in the treatment of this entity.

Key words: solitary plasmacytoma, maxillary tumor, functional rehabilitation

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Introduction

Plasma cell neoplasms are clonal proliferations of plasma cells characterized by secretion of a single homogeneous immunoglobulin known as monoclonal component (1).

Among the monoclonal gammopathies there are: multiple myeloma (MM) and the solitary plasmacytoma classified as: solitary plasmacytoma of the bone (SPB) and extramedullary plasmacytoma (EMP). The International Myeloma Working Group describes a third form – the solitary multiple plasmacytoma (2).

Multiple myeloma is a generalized tumoral proliferation of plasma cells and it represents the most frequent primary malignant bone tumour. Solitary plasmacytoma is a very rare neoplasia and it represents a monoclonal proliferation of plasma cells under the form of a solitary mass which can be situated in the bone marrow or extramedullary.

Solitary plasmacytoma of the bone represent less than 5% of plasma cell neoplasms. It mainly involves the axial skeleton, especially the vertebrae and in the majority of cases progresses to multiple myeloma. The symptoms are dependent on tumour localization. The most common symptom is pain at the site of the skeletal lesion or a pathological fracture. For the vertebral localizations there can also be compressions of nerves leading to severe pain and even neurological damage. Often, the discovery of plasmacytoma is made accidentally after a fracture through a routine X-ray examination (2).

Extramedullary plasmacytoma appears outside the bone, in the soft tissues, and it has a better prognostic than the solitary plasmacytoma of the bone. Less than 30% of extramedullary plasmacytomas progress towards multiple myeloma. They can involve any anatomical region but in most of the cases are localized in the head and neck region. 80% originate in the submucous tissue of the superior aerodigestive tract. (3,4,5) They can also be found in the parotid gland, thyroid, breast or in the gastrointestinal tract (6).

Multiple solitary plasmacytomas affects the soft tissue or the bone, and may be recurrent but with no evidence of bone marrow involvement or other skeletal lesions. (2)

All these clinical-pathological entities differ in terms of location, tumour progression, and overall survival rate.

We present a rare case of a male patient diagnosed with well differentiated multiple solitary plasmacytomas.

Case report

A 52-year-old man was admitted to the OMF Surgery Department of the Clinical Emergency Hospital Sibiu in November 2010 at the recommendation of the haematologist for evaluation and possible surgical cure of a tumoral proliferation localized in the region of the right maxilla. The evolution of the tumour was known since 2008, while the patient was under treatment at the Haematology Department of the Academic Emergency Hospital Bucharest.

In his medical history he presented a spontaneous fracture in 1998 in the left humeral diaphysis which led to the discovery of an Ig G secretory plasmacytoma, for which he underwent 5 cures of VMCP (Vincristine, Melphalan, Cyclophosphamide, and Prednisone). In 2003 the tumour relapsed and the patient was directed to the Orthopaedic Department of the Fundeni Hospital Bucharest, where he underwent excision of the humeral tumour and reconstruc-

tion with non-vascularised fibula bone graft.

In 2006 the tumour reappeared in the right ulna. Treatment of choice was surgical resection and chemotherapy.

In July 2008 he was admitted to the Orthopaedics Department of the Academic Emergency Hospital Bucharest for a tumoral mass in the left calf which was surgically removed with resection in oncological limits. The repeated medullograms did not show any medullary plasma cell infiltrate, and by then skeletal radiographies showed no other concomitant lesions.

The patient was submitted to a bone scintigraphy in September 2008 which showed areas of high capture of the RT in the left tibia, the left tarsal bones and on the anterior border of the coxal bone on the right side, indicating the progression towards multiple myeloma.

He was immediately directed to the Haematological Department of the Emergency Academic Hospital Bucharest given the progression to multiple myeloma and the appearance of a maxillary proliferation. The patient underwent a total of 6 cures of VAD (vincristine, Adriamycin, Dexamethasone), the last one being in March 2009. Under this treatment the maxillary tumour regressed. Thalidomide was administrated for a year without observing significant adverse reactions.

In May 2010 the maxillary tumour relapsed and extended to the right orbit. The therapy with VAD was reinitiated and the patient underwent 3 sessions. The result was unsatisfactory: the tumour did not recede.

The lack of a response to chemotherapy in the maxillary tumour determined the haematologists to direct the patient to our OMF Department in November 2010.

When the patient presented in our Department we noticed a voluminous tumour in the right zygomatic-maxillary region extended in the right orbit and in the soft tissues with a mass effect over the eye bulb, but without visual acuity disturbances. Palpebral oedema and unilateral nasal drainage could be also observed (Fig. 1A).

Anterior rhinoscopy showed the obstruction of the nasal cavity due to the tumoral extension from the maxillary sinus to the nasal fossa deforming even the nasal septum.

The CT scan showed an expansive, iodophile, mass process, which projected in the malar and orbital region on the right side, occupying the maxillary sinus completely on this side: the dimensions were 6/5 cm axially and 5 cm cranial-caudal. Destruction of the orbital floor and the anterior and medial walls of the right maxillary sinus, without the palatal platform (Fig. 1B) were noticed.

Given the destructive form of the lesion and the lack of response to chemotherapy the haematologist recommended surgical excision of the tumour followed by self-transplant with stem cells after the surgical intervention.

The tumour was resected completely, the resection piece had the dimensions of 90/75/60mm and expanded through the zygomatic-maxillary bone complex, embedding the floor and the lateral wall of the orbit; the alveolar palatal portion was not involved (Fig. 2A). The skin of the genial zygomatic region, free of tumour infiltration, was spared. The right external carotid was ligatured due to the haemorrhage risk of the tumour. The

Figure 1. (A) Massive tumour in the right zygomatic-maxillary region extended in the right orbit and in the soft tissues with the narrowing of the palpebral opening and with mass effect on the eye bulb. (B) CT scan highlights an expansive tissue mass occupying the right maxillary sinus completely, having projection in the nasal, malar and orbital region on the same side

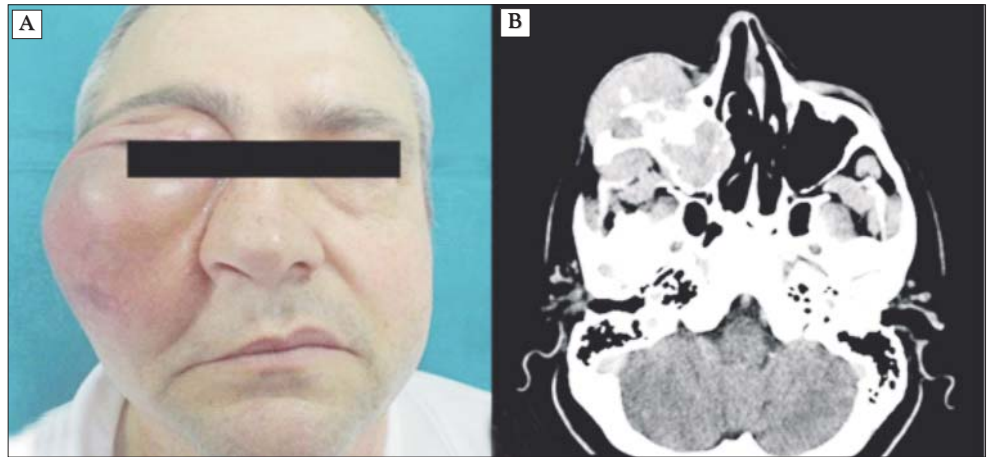


Figure 2. (A) Intra-operative photo of the tumour; (B) Reconstruction of the orbital floor

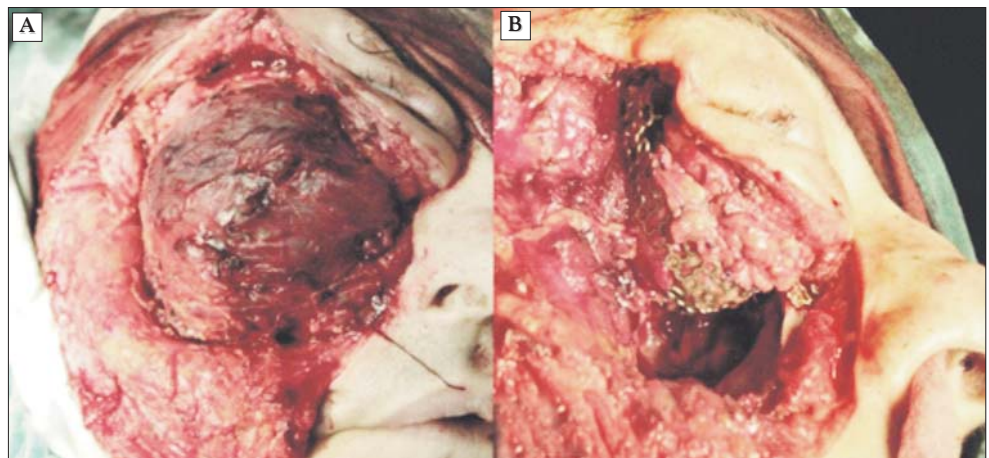
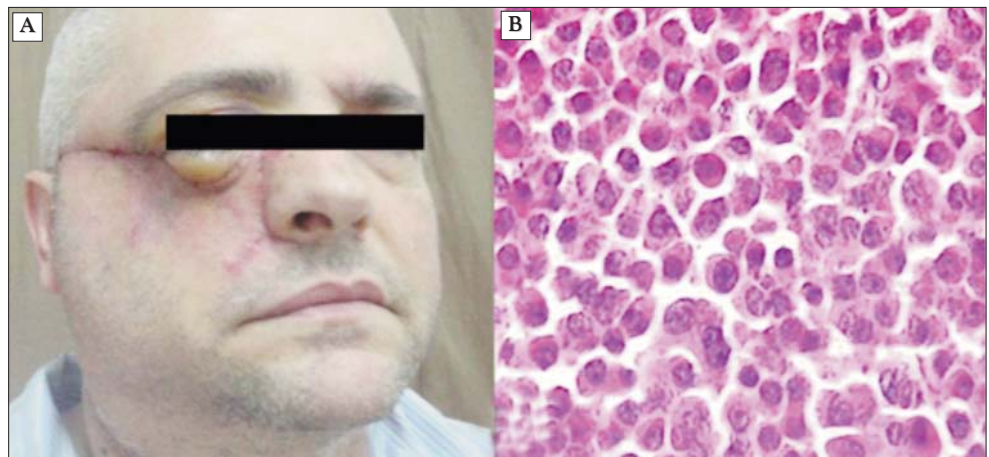


Figure 3. (A) Post-operative clinical exam after 7 days. (B) Histological examination shows plasma cell proliferation with different degrees of atypia and maturity (H.E. 40 X)



reconstruction of the orbital platform with titanium mesh allowed the preservation of the eye bulb's functionality (Fig. 2B).

At 20 days follow up normal eye function was observed (Fig. 3A).

The histologic examination of the excised piece showed fibrous tissue infiltrated with plasma cells with different degrees of atypia pleading for a multiple myeloma (Fig. 3B).

Two years later, the onset of kidney insufficiency through the evolution of the multiple myeloma, caused the death of the patient, without any sign of local relapse.

Discussions

Solitary plasmacytomas are rare afflictions with vague and nonspecific initial symptoms, and are therefore hard to detect

The early diagnosis of a specific clinical-pathological entity is very important in the choice of the therapy and survival rate. Differential diagnosis must be undertaken carefully as early signs are not very conclusive (7). The evidence of plasma cell infiltrate in the HP examination orients

the diagnosis of solitary plasmacytoma, but it is insufficient since the diagnosis of multiple myeloma has to be excluded through medullar puncture, blood and urine tests, as well as through imagistic explorations. For the diagnosis of solitary plasmacytoma, the imagistic investigations of the whole skeleton have to prove the presence of a unique zone of bone destruction due to the clonal plasma cells without other osteolytic lesions or tissue involvement. Medullograms have to highlight the infiltration of the bone marrow with plasma cells below 5% of all the nucleate cells. Also, the serological and urine examinations have to show low concentration of the monoclonal protein in the serum or urine as well as the absence of anaemia, hypocalcaemia or kidney involvement attributed to multiple myeloma (2).

The case presented was monitored by the haematologist for over 14 years. The patient was diagnosed with multirecurrent solitary plasmacytoma having multiple localizations - humerus, ulna, calf, upper maxillary - separated from one another in time. Initially it responded to chemotherapy, then surgical resections were necessary followed by reconstruction.

The maxillary localization had an evolution of 2 years, obtaining transitional remissions after chemotherapy. Later, the tumoral growth could not be controlled anymore, the destruction of the inferior wall of the orbit and the orbital invasion imposing surgical resection.

Conclusions

The group of afflictions embedding the neoplasia with plasmacytes represents different manifestations of a sum of pathologies.

The election therapy is radiotherapy with doses between 40-50 Gy, these tumours being intensely radiosensitive (2,8).

Some studies recommend combined chemotherapy along with other therapies in patients with high risk of evolution towards multiple myeloma (9).

For some cases which do not respond well to radiant treatment and in which the lesions have destructive features, there are also treatments which combine surgical excision with radiotherapy described in the literature (8).

In the presented case, despite the histological examination, the bone marrow puncture and the imagistic explorations established the progression towards multiple myeloma. Complete surgical resection was the treatment of choice for this maxillary chemotherapy non-respondent tumour. The patient survived for a period of two years after the maxillary tumour excision, with no local recurrence, with ocular functionality as well as aesthetic rehabilitation.

This evolution underlines the benefits of surgery combined with radio-chemotherapy in the treatment of this entity.

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Competing interests

The authors declare that they have no Competing interests.

Ethical approval

Not required.

Author's contribution

The authors of the present paper "Maxillary Solitar Recurrent Plasmacytoma: a case report" were effectively and actively involved in this paper conception, wording as in this case prosthetic and surgical treatment. The primar development of the manuscript as well as the critical analysis were performed both individually and by the team. The setting of the final version were sustained by the entire collective, being supervised by Prof. Dr. Viorel Ibric Cioranu.

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