

CASE REPORT

Extramedullary plasmacytoma of paranasal sinuses. A combined therapeutic strategy

Plasmacitoma extramidollare dei seni paranasali. Approccio chirurgico combinato

G. ATTANASIO, M. VICCARO, M. BARBARO, E. DE SETA, R. FILIPO
Department of Neurology and Otolaryngology, University "La Sapienza", Rome, Italy

Key words

Paranasal sinuses • Plasmacytoma • Surgical treatment

Parole chiave

Seni paranasali • Plasmocitoma • Trattamento chirurgico

Summary

Extramedullary plasmacytoma of the head and neck region is a rare malignant tumour comprising approximately 3% of all plasma cell tumours. Approximately 80-90% of extramedullary plasmacytomas involve the Mucosa-Associated Lymphoid Tissue of the upper airways, 75% of these involve the nasal and paranasal regions. Radiotherapy is considered the treatment of choice, surgery being limited to biopsy and to excision of residual disease. A case of extramedullary plasmacytoma of the nasal cavity and ethmoid sinus is reported, in which surgical excision is followed by complementary radiotherapy on the site of the tumour.

Riassunto

Il plasmacitoma extramidollare (PE) della testa e del collo è un tumore maligno raro che rappresenta circa il 3% di tutti i tumori delle plasmacellule. In circa l'80-90% dei casi il plasmacitoma extramidollare coinvolge il Tessuto Linfatico "Mucosa Associated" delle vie aereo-digestive superiori, e nel 75% dei casi si localizza a livello del naso e dei seni paranasali. La radioterapia è considerata il trattamento di scelta, essendo il ruolo della chirurgia limitato alla biopsia escissionale ed all'asportazione di eventuali residui di malattia. Riportiamo il caso di una donna di 67 anni ricoverata presso il nostro Dipartimento a seguito della comparsa, 5 mesi prima, di una tumefazione di colorito rosso a carico del canto interno e della radice del naso. L'esame biotico rilevò la presenza di un plasmacitoma CD79+ con catene leggere lambda. Il tumore venne rimosso chirurgicamente, con successiva irradiazione della sede del tumore.

Introduction

Extramedullary plasmacytoma is an uncommon plasma cell neoplasm which occurs in soft tissue. It shows a preference for the upper respiratory tract, especially the nasal cavity and paranasal sinus. It accounts for less than 1% of head and neck tumours.

Extramedullary plasmacytoma (EMP) of the head and neck region accounts for approximately 3% of all plasma cell tumours.

It typically arises in submucosal soft tissues of the upper respiratory tract and it is destructive with a tendency to local recurrence^{1,2}. The disease is more common in the male sex, during the fifth and sixth decades of life.

To exclude multiple myeloma or plasmacytoma of the bone, a systemic work-up and follow-up of the patient are mandatory, including serum protein electrophoresis, urinalysis for the Bence-Jones protein, skeletal survey and bone marrow biopsy^{3,4}.

The optimal management of EMP is controversial. Surgery can achieve high rates of local control in certain situations. However, radical excision is often impossible due to the size or the location of the tumour⁵. A case of EMP of the left nasal cavity and ethmoid sinus is reported, in which surgical excision followed by complementary radiotherapy on the site of the tumour has been performed.

Case report

A 67-year-old female was admitted to the ENT clinic on account of a reddish lump, which had been present for 5 months, involving the left medial canthus and the base of the nasal pyramid. Her past medical history revealed smoking habit, hypertension, uterine fibroma, and breast cancer treated with quadrantectomy two years earlier. Unilateral nasal obstruction, discharge, bleeding, but not diplopia, were reported. No

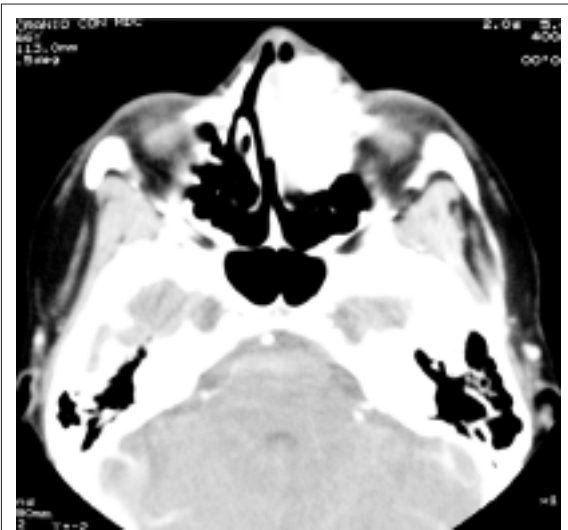


Fig. 1. Pre-operative CT scan showing soft tissue involving left nasal cavity and ethmoid sinus.

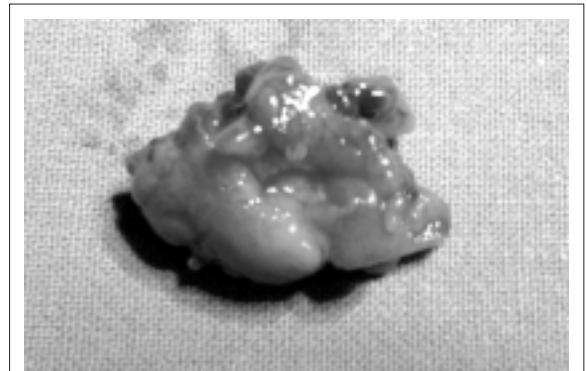


Fig. 2. Intra-operative specimen after *en bloc* excision.

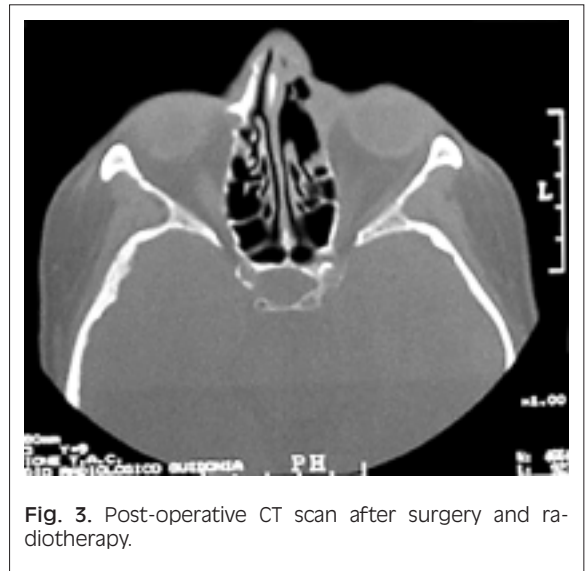


Fig. 3. Post-operative CT scan after surgery and radiotherapy.

palpable lateral neck nodes were present. Clinical examination showed a soft dark red mass in the left nasal cavity, at the level of the middle meatus. Computed tomography (CT) scan revealed a soft tissue involving the entire ethmoid sinus. The histological finding, following nasal biopsy, was plasmacytoma CD79a+ with light Lambda chain. A systemic work-up to exclude multiple myeloma was performed.

Renal and liver function were normal, blood profile revealed low plasmatic protein concentration (5.6 g/dl) and serum electrophoresis detected lambda monoclonal component. Bence-Jones protein was absent in urine. Bone marrow needle biopsy, skeletal survey and bone gamma scan were negative. Thus a diagnosis of EMP was made.

The tumour was removed surgically with the paranasal approach (Fig. 2). One month later, the patient received radiotherapy (RT) with a radiation dose of 40 Gray in 20 fractions. One-year follow-up revealed no recurrence (Fig. 3).

Discussion

Plasma cell tumours include the disseminated form, multiple myeloma, and the medullary (solitary plasmacytoma of the bone) and the extramedullary localized form.

Approximately 80-90% of EMP involve the Mucosa-Associated Lymphoid Tissue (MALT) of the upper airways, 75% of these involve the nasal and paranasal regions⁶.

Due to its presentation in the submucosa of the aere-

odigestive tracts, some Authors have suggested that the aetiology of EMP may be related to chronic stimulation caused by inhaled irritants or viral infection⁷. The most common clinical findings are: blocked nose, soft tissue mass (fleshy, yellowish grey to dark red sessile, polypoid, or pedunculated), epistaxis, nasal discharge, pain, more rarely cranial nerve palsy and neck lymphadenopathy.

In the present case, a systemic work-up including blood profile, renal and liver function, serum and urinary protein electrophoresis, serum immunoglobulin level, skeletal survey and bone marrow examination has been performed to exclude a systemic disease such as multiple myeloma.

RT is the treatment of choice in EMP localized in the head and neck, not extending through the floor of the anterior and middle cranial fossae and into the orbit, whereas the role of surgery is usually limited to biopsy and to excision of residual disease⁸. The progn-

sis for patients with extramedullary plasmacytoma is: 5-year survival rates between 30 and 82% and 10-year rates 50 to 90%.

In our patient, the size of the tumour and its location, close to the medial wall of the orbit, led us to perform first-step surgery followed by complementary RT, 30 days thereafter. This choice allowed us to minimize

the damage of RT on the orbit. No recurrence was detected at clinical examination or at CT scan performed one year after surgery.

Although the follow-up is not sufficiently long to consider the complete remission of disease as definitive, the therapeutic strategy adopted to date, in this case, might be considered valid.

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■ Received: June 30, 2005
Accepted: January 19, 2006

■ Address for correspondence: Prof. R. Filipo, Dipartimento di Otorinolaringoiatria e Neurologia, Università di Roma "La Sapienza", viale dell'Università 31, 00161 Roma, Italy. E-mail: roberto.filipo@uniroma1.it

The expert's comment

A. AGAZZI, F. PECCATORI
European Institute of Oncology, Milan, Italy

As the Authors state in this paper, the EMPs represent an uncommon feature in the scenario of Plasma Cell disorders. However, a number of EMPs present as nasal polyps which are common: upon removal, nasal polyps are often not subjected to histologic examination. Therefore, it is possible that the actual incidence of EMPs is higher than expected. While EMPs are most often observed in the head and neck, regional lymph nodes draining the site should be investigated because they can be involved in up to 25% of cases. EMPs can be associated with systemic presentation: Wiltshaw and Woodroof proposed, in 1979, a staging system for EMPs:

Stage I: tumour confined to primary site;
Stage II: involvement of drainage lymph nodes;
Stage III: evidence of metastatic spread (Multiple Myeloma).

According to the current literature, it is well recognized that EMPs are sensitive to radiation therapy and 4000-5000 cGys assure local control, in most patients. If biopsy only has been performed to obtain a diagnosis, complete excision is required. If complete local control is not achieved with radiation, surgical excision of the remaining lesion may be discussed because of the potential widespread of the disease. If more than one tumour has been detected, all involved sites should be irradiated too. Whether regional nodes must be irradiated prophylactically is not known even if it is routinely practiced in some Institutions. Again, multiple tumours may require a systemic approach (chemotherapy) including, in some instances, high-dose chemotherapy and stem cell transplantation.