UNILATERAL EXOPHTHALMOS AS THE DEBUT OF A NON-SECRETORY MULTIPLE MYELOMA

EXOFTALMOS UNILATERAL COMO DEBUT DE MIELOMA MÚLTIPLE NO SECRETOR

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ABSTRACT

Case report: A 56 year-old male presented blurred vision and diplopia for 2 months, left unilateral exophthalmos, restricted ocular motility and papilledema. The imaging proofs showed osteolytic lesions in the left sphenoid bone, fourth rib and fourth dorsal vertebral body with associated masses of soft tissues. Biopsy was performed and the diagnosis of plasma cell neoplasm was established. The diagnosis of non-secretory multiple myeloma was made by analytical criteria and bone marrow biopsy. Local radiotherapy and polychemotherapy was prescribed.

Conclusions: The ophthalmologist can play an important role in the diagnosis of systemic neoplasms that require the intervention of a multidisciplinary team (Arch Soc Esp Oftalmol 2009; 84: 631-634).

Key words: Plasma cell neoplasms, multiple myeloma, exophthalmos, orbit tumors.

RESUMEN

Caso clínico: Varón de 56 años con visión borrosa y diplopía de 2 meses de evolución, exoftalmos unilateral izquierdo con miopatía restrictiva y papiledema. Las pruebas de imagen mostraron lesiones osteolíticas en esfenoides izquierdo, 4.ª costilla y cuerpo de la cuarta vértebra dorsal, con masas de partes blandas asociadas. La punción aspiración con aguja fina de la lesión confirmó el diagnóstico de plasmocitoma. Se diagnosticó de mieloma múltiple no secretor por criterios analíticos y biopsia de médula ósea. Se instauró tratamiento con radioterapia local y poliquimioterapia.

Conclusiones: El oftalmólogo puede desempeñar un papel primordial en el diagnóstico de neoplasias sistémicas, que requieren la intervención de un equipo multidisciplinario.

Palabras claves: Neoplasias de células plasmáticas, mieloma múltiple, exoftalmos, tumores de órbita.
INTRODUCTION

Plasmatic cell neoplasias account for 1-2% of malign neoplasias. They are classified according to their origin as multicentric, such as multiple myeloma (MM) which courses with bone marrow infiltration, and localized, with origin in the bone such as solitary bone plasmocytoma, or in soft tissue such as extra-medullar solitary plasmocytoma. The involvement of the orbit is infrequent and in most cases it represents the first expression of a MM (1).

This short paper presents the case of orbitary involvement due to a plasmatic cell neoplasia which began with unilateral exophthalmos, emphasizing the importance of early diagnostic in these lesions.

CASE REPORT

A 56-year old male with visual acuity reduction (VA) in the left eye (LE) and diplopia with 2 months of evolution. Upon exploration, a left unilateral exophthalmos was revealed with restriction of ocular motility. The VA was 20/20 in right eye and 20/30 in LE. Anterior segment biomicroscopy was normal and the funduscopic assessment revealed a papillary edema in LE (fig. 1A). An orbital-cranial computerized axial tomography (CAT) showed the presence of osteolytic lesions in the major and minor wing of the left sphenoid and a soft mass extending to the temporal fossa, the infra-temporal fossa and orbit (fig. 2A). An extension chest-abdomen CAT revealed osteolytic lesions involving the posterior arch of the 4th rib and the vertebral body of D4 with associated soft mass. A puncture-aspiration with thin needle (PAAF) was made in the lesion with a result compatible with plasmatic cell neoplasia. The serum and urinary proteingram was normal and the bone marrow biopsy was positive, establishing the diagnostic of non-secretory MM. Treatment was established with local radiotherapy prior to polychemotherapy. After 3 months, the patient’s VA in LE was 20/20 and the funduscopic assessment did not produce significant findings (fig. 1B). The orbital-cranial and chest-abdomen CAT (fig. 2B) exhibited the presence of slight lytic lesions in the locations referred to above without associated soft mass.

DISCUSSION

MM is a plasmatic cell neoplasia which segregates monoclonal para-proteins. It accounts for 10% of hematological neoplasias, with a prevalence of 5.5 cases/100,000 inhabitants (1). In turn, non-secretory MM represents 1-4% of myeloma cases, with a mean diagnostic age of ten years under the diagnostic age of classic MM forms (2).

The ocular expressions of MM involve the conjunctiva, cornea, iris, cilliary body, choroids and sclerotic (3). The involvement of the orbit is rare, with about 50 cases published in the literature (1). Orbitary involvement is generally unilateral and

![Fig. 1: Ocular fundus images. A) showing papillary edema in left eye and B) without significant findings after treatment.](image-url)
more frequently affects the sphenoid (3). In the majority of cases, the initial symptoms are insidious with slowly progressing exophthalmos together with pain, diplopia and VA reduction (4). The intracranial expansion expresses via papilledema and paralysis of cranial pairs. Also it is frequent to find non-specific symptoms such as fever, discomfort and anorexia (3). A careful evaluation of symptoms, prescription of imaging tests as well as extension study were crucial to establish the early diagnostic of MM in the present case.

In the presence of a orbital neoplasia of plasmatic cells it is important to prove the presence of systemic expressions in order to establish an early MM diagnostic. The diagnosis of localized forms such as solitary bone plasmocytoma and extra-medullar solitary plasmocytoma is made in the presence of a plasmocytoma without systemic findings after discarding the involvement of lymphatic nodules and axial skeleton, carrying out a bone marrow biopsy and image tests (5). The difference with MM lies in the invasion of the bone marrow in addition to analytical changes and systemic expressions which are typical of this neoplasia. Approximately two thirds of solitary bone plasmocytoma and one third of patients with extra-medullar solitary plasmocytoma will progress to MM (5).

Non-secretory MM is a special form of myeloma. The clinical signs are common to the classic form, sharing the typical osteolytic lesions and the presence of plasmocytomas as main diagnostic criteria. The peculiarity of this type of myeloma is the absence of paraproteins in blood and urine, with the ensuing diagnostic difficulty (2). The absence of secretion of the M component makes kidney involvement rate and improves the prognosis (2).

In the case presented here, PAAF of the lesions demonstrated a plasmocytoma, the extension study revealed osteolytic lesions in other locations and the bone marrow biopsy showed an infiltration of plasmatic cells, whereas the serum and urinary proteins were normal. On the basis of these clinical findings, the diagnostic of non-secretory MM was established. As conclusion, we emphasize that the ophthalmologist can play an essential role in the early diagnostic of systemic neoplasias, which require the intervention of a multidisciplinary team.
REFERENCES


