

Unilateral ptosis as the initial sign of multiple myeloma[☆]



Ptosis palpebral unilateral como debut de mieloma múltiple

Multiple myeloma is the third most frequent haematologic neoplasia, after non-Hodgkin lymphoma and chronic lymphocytic leukaemia. It is characterised by the uncontrolled proliferation of plasma cells in the bone marrow leading to the overproduction of monoclonal immunoglobulins.¹

It has an incidence of 5.5 cases/100 000 person-years² and is slightly more frequent in men (male/female ratio, 1.6:1). Mean age of onset is 68 years¹; presentation in patients younger than 40 is rare.³

Bone pain, the most frequent symptom of the disease, is present in at least 70% of cases. Patients with myeloma may also present anaemia due to tumour invasion of the bone marrow, hypercalcaemia secondary to bone destruction, renal failure caused by immunoglobulin deposition, and a predisposition to bacterial infections, especially pneumonia or pyelonephritis.^{1,4}

Orbital involvement is rare in multiple myeloma. We present a case of multiple myeloma with an unusual clinical onset of isolated ptosis.

Clinical case

Our patient was a 73-year-old woman with no relevant medical history, who was transferred to our neurology department from the ophthalmology department due to a 3-week history of left-sided ptosis. The ptosis had developed suddenly and did not fluctuate. A few days after our patient first noticed the ptosis, she began to experience occasional episodes of diplopia lasting for several minutes. She reported no bone pain or constitutional symptoms. Physical examination revealed 6-mm left-sided ptosis; no exophthalmos or impaired extraocular movements were seen. Fundus examination revealed no abnormalities.

The neurological examination and general physical examination also showed normal results.

A cranial MRI scan and a CT scan of the paranasal sinuses revealed a lytic lesion on the left orbital floor associated with a soft tissue mass extending to the maxillary sinus and affecting the bony wall of the left nasal cavity. Furthermore, another lytic lesion was found on the right mandibular ramus. A PET scan showed infiltration of the left maxillary sinus, left humerus, and some ribs. Results from the orbital mass biopsy were compatible with plasmacytoma (CD138+, MUM1+, intense positivity for CD56), although lymphoplasmacytic lymphoma could not be ruled out due to the location of the mass. Blood tests revealed mild pancytopenia, hypogammaglobulinaemia, and the presence of a monoclonal component (0.24 g/dL). Immunoelectrophoresis showed a gamma band compatible with IgG-kappa, while a

urine test found free kappa light chains. In view of these findings, we decided to perform a bone marrow biopsy, which confirmed the diagnosis of multiple myeloma with plasma cell infiltration (30%).

Discussion

Very few patients with multiple myeloma present neurological symptoms. However, given the characteristics of this entity, there is a wide range of neurological clinical syndromes which may be present during disease progression or even very occasionally representing some of the initial symptoms. Manifestations will depend on the location and size of the tumour.^{1,3,5} Compression fractures may cause spinal cord compression and/or radiculopathy. Diffuse infiltration of the subarachnoid space (myelomatous meningitis) may also occur. Hypercalcaemia can present with symptoms of encephalopathy. Tumour expansion from facial or cranial bone structures can lead to the involvement of cranial nerves or intracerebral space-occupying lesions. Lastly, amyloid deposition on peripheral nerves can cause mono or polyneuropathy.¹

Orbital involvement is rare in the context of multiple myeloma, with very few cases published in the literature. In 2009, Burkat et al.⁴ conducted a literature review and found a total of 71 cases. In only a third of these were orbital symptoms the initial manifestation of the disease; in the remaining cases, orbital involvement occurred after diagnosis of multiple myeloma.

Myelomas can affect any intraorbital structure. Globes are rarely affected; the most frequent manifestations include retinal haemorrhages, macular detachment, cotton-wool spots, and crystal or copper deposition in the cornea.⁴

Orbital multiple myeloma normally presents as a solitary soft-tissue tumour originating from bone deposits and is frequently associated with bone destruction.⁵ According to the literature, involvement is predominantly unilateral (88%); masses are more frequently located in the posterior part of the orbital cavity (69%), and up to 90% of lesions are located in the extraconal space.⁴

The most frequent initial symptom is exophthalmos (81%) followed by loss of vision (23%), diplopia (23%), palpebral oedema (21%), and ptosis to a lesser extent (13%).⁴ These data confirm the rareness of our case, which manifested with isolated unilateral ptosis. According to most authors, symptoms begin insidiously.^{3,6,7}

The presence of isolated unilateral ptosis allowed us to rule out oculomotor nerve nucleus or fascicle involvement as a possible cause. Consequently, ptosis could only be due to a dysfunction of either the levator palpebrae superioris muscle or the superior branch of cranial nerve III, which emerges from the posterior part of the orbit.

Neuroimaging results, which show a tumour on the orbital floor, do not reveal direct compression on either of these structures. We hypothesise that the tumour mass may have displaced the orbital contents rostrally, compressing the levator palpebrae superioris muscle against the roof of the orbit.

In our case, myeloma was an IgG secretor, as is the case in the majority of myelomas involving the orbit, to an even greater extent than myelomas in other areas.⁶

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Orbital involvement in myeloma tends to signify a more aggressive form of the disease, and survival rates are significantly poorer in these cases.⁴

Although fewer than 1% of all orbital tumours are myelomas,¹ this possibility should be considered in the differential diagnosis. Careful communication and coordinated teamwork among the different services implicated (in our case, the departments of neurology, ophthalmology, otorhinolaryngology, and haematology) are crucial to reach an accurate diagnosis when myeloma presents in this location.

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Subarachnoid haemorrhage from a ruptured intracranial mirror-like aneurysm. A case report and literature review[☆]



Hemorragia subaracnoidea por rotura de aneurisma especular intracraneal. A propósito de un caso y revisión de la literatura

Dear Editor:

Subarachnoid haemorrhages (SAH) are responsible for 6% to 8% of all acute cerebrovascular events, 5% of deaths, and 25% of the potential life years lost. Their importance lies in the fact that they tend to affect younger patients than ischaemic stroke and result in high morbidity and mortality rates. In addition, a SAH is both a pathological process in itself and an epiphenomenon of an underlying disease that must be treated.^{1–5} We present the case of a patient with aneurysmal SAH (aSAH) manifesting with intermittent symptoms who was found to have mirror aneurysms in both middle cerebral arteries (MCA).

Our patient was a 42-year-old man who presented at the emergency department with fluctuating non-traumatic clinical symptoms consisting of a holocranial headache of moderate intensity, which had started 4 days previously

while he was playing volleyball. The headache was self-limiting but later reappeared in association with presyncopal symptoms and meningismus. The patient described the headache as 'the worst headache of his life'. Suspicion of aSAH led to a request for an urgent brain CT scan and a general blood test. The blood test yielded normal results and the CT scan revealed an aSAH (Fig. 1A). The vascular study also included an intracranial CT angiography (Fig. 1B), which showed an aneurysm on each MCA (distal M2 segment). The haemorrhage was attributed to the aneurysm on the left MCA, which was confirmed by cerebral arteriography (Fig. 2). The patient was diagnosed with aSAH (Hunt & Hess grade 1, WFNS grade 1, Fisher grade 3). After starting on oral nimodipine 60 mg/4 h, our patient was admitted and underwent an emergency pterional craniotomy (clipping the aneurysm on the M2 segment of the left MCA); the same procedure was repeated for the unruptured aneurysm on the right MCA 30 days later. The clinical and radiological outcomes were favourable, and the patient recovered *ad integrum*.

Despite the many achievements in SAH diagnosis and treatment, SAH is still a serious entity: 10% of patients die before arriving at the hospital, 25% die within the first 24 hours after bleeding, 45% die within the following 30 days, 50% present sequelae, and only 33% achieve good outcomes after treatment.^{1–5} The incidence of ruptured intracranial aneurysms increases with age (≥ 60 years) and aneurysm size (≥ 5 mm), and is greater in women, patients with posterior circulation aneurysms, and symptomatic patients. In order of frequency, intracranial aneurysms are located in the anterior communicating and anterior cerebral arteries (35%), the middle cerebral artery (25%), the posterior communicating artery (22%), and arteries of the posterior circulation (18%). Between 15% and 33.5% of patients with SAHs have multiple aneurysms.^{1–4,6} Mirror or twin aneurysms, a rare subtype of

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