INTRODUCTION

The POEMS syndrome is a rare systemic disease related to multiple myeloma and defined by the presence of polyneuropathy (mixed sensitive-motor type and predominantly demyelinating), organomegaly, endocrinopathy, monoclonal component (monoclonal gammopathy IgA-1 or IgG) and skin lesions (diffuse hyperpigmentation, acropachy).

According to the criterion established by Dispenzieri, 2 major criteria are required (polyneuropathy and monoclonal component) plus at least one minor criterion (sclerotic bone lesions, Castleman’s disease, organomegaly, ede-
ma, endocrinopathy, skin changes or papilledema) for diagnosing the POEMS syndrome (2).

**CASE REPORT**

A 54-year-old patient exhibiting paresthesia in lower limbs and bilateral papilledema, bigger in the left eye (LE) than the right eye (RE) and with hemorrhage in the nervous fiber layer of the LE (fig. 1). The patient referred temporary visual loss although upon diagnosis she was non-symptomatic at the ophthalmological level with a visual acuity (VA) of 1.0 in both eyes and a normal SITA Standard 24:2 computerized campimetry. A nuclear magnetic resonance (NMR) was performed with normal results, and an electromiogram (EMG) with mixed polyneuropathy. Analytics revealed a monoclonal IgA-1 gammapathy and the skin exploration exhibited hyper-pigmentation and Hippocratic fingers (fig. 2). The patient was diagnosed with the POEMS disease and treatment was established with Melfalan 250 µg/kg/day for five days every six weeks and prednisone 30mg every 48 hours, maintained for one year.

In 2000 the patient exhibited a reduction in visual acuity. Exploration showed a maximum corrected visual acuity of 0.5 in each eye, associated to a relative afferent pupillary defect in the RE. This is not frequent in the papilledema associated to POEMS but there was no other associated optic neuropathy. The exploration also revealed color vision reduction in the same eye (4/15 in Ishihara test) and progression of the papillary edema. The visual field evidenced an increase in the bilateral blind spot and an arch-shaped scotoma in the LE (fig. 3).

The pressure of the intracranial fluid was of 28 cmH 2 O (normal values are below 25 cmH 2 O). Upon confirmation of high intracranial fluid pressure, treatment was established with acetazolamide 250 mg every eight hours for one week, after which the pressure returned to normal levels.

After one month, the systemic condition worsened. The hematology service proposed a bone marrow transplant which was declined by the patient. It was decided to substitute the Melfalan treatment with Rituximab, an anti-Ag CD20 monoclonal antibody.

The last exploration, which was carried out three months after the Rituximab treatment, revealed a corrected VA of 1 in both eyes, a relative afferent pupillary defect in the RE, partial involvement of green and red vision, while the perimetric assessment revealed an increase of a blind spot in both eyes with a nasal step in the LE (fig. 4). Eye fundus showed a bilateral chronic papillary edema. The visual condition was controlled without the need for treatment.
DISCUSSION

On rare occasions, papilledema is the initial expression of systemic diseases such as the Guillain-Barré or POEMS syndromes [1].

At the ophthalmological level, patients may exhibit papilledema (37-73 %) (2), peripapillary drusen and/or peri-papillary choroidal neovascularization. The papillary edema is usually chronic and bilateral with blind spot increases, with or without involvement of the VA (according to the stage of the disease) without evolution to optical atrophy.

As regards the pathogeny and physiopathology of this disease, there are a number of theories: vasculitis in the papilla (2,5), infiltration of the optic nerve (2,5), hyper-proteinrachy and intra-cranial hypertension (2,5) and finally increased vascular permeability regulated by cytokines (3).

To date, the most usual treatments were radiotherapy (over localized osteo-sclerotic lesions and plasmocytomes), chelating agents (melfalan and cyclophosphamide) and corticoids. Of late, these treatments are being displaced by more specific therapies such as monoclonal antibodies like rituximab (anti CD20) in doses of four rituximab 375 mg/m 2 infusions at weekly intervals (4) or the anti-VEGF such as bevacizumab.

Treatment for papilledema will only be necessary in the case of visual clinical symptoms and will vary according to the etiology. In the case of a patient with the POEMS syndrome and papilledema, a lumbar puncture must be made to determine intra-cranial pressure. If it exceeds 25 cm H 2 O we would consider high intracranial pressure as the cause of the papilledema. In that case, we would treat with oral acetazolamide to achieve clinical stabilization. If there is no improvement we would consider the therapeutic possibility of opening the sheath of the optic nerve, although to our knowledge there are no cases requiring fenestration in this type of patients. If the intracranial pressure is above 25 cm H 2 O we would consider the papilledema secondary to an infiltration process and would assess several therapeutic options, including high doses of systemic corticoids or brachytherapy on the optic nerve, or the more recent use of monoclonal antibodies (5).

REFERENCES