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Ascites with Elevated Protein Content as the Presenting Sign of POEMS Syndrome: Case Report

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Abstract **Case Report**

POEMS syndrome is a rare multisystemic disease that occurs in the setting of a plasma cell dyscrasia. Peripheral neuropathy is the dominant clinical feature of this disorder and is often the inaugural sign. We report the case of a 46-year-old patient, followed-up for chronic polyradiculoneuropathy, admitted in our department for etiological assessment of an ascites with elevated protein content and important weight loss. The diagnosis of POEMS syndrome was established in the presence of two major criteria associated to several minor criteria (organomegaly, ascites, papillary edema and skin changes, hypothyroidism). Dexamethasone was the initial treatment used for our patient then it was combined to thalidomide. The clinical outcome was good.

Keywords: Ascites, POEMS, polyradiculoneuropathy.

INTRODUCTION

Rich in protein ascites is often due to malignancies or infections like peritoneal tuberculosis; however the POEMS syndrome is also an etiology that remains rare. This syndrome is a multisystem disorder that associates in the complete forms a polyneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell disorder and skin abnormalities. We report the case of a patient admitted for etiological assessment of a high protein ascites whose investigations were in favor of a POEMS syndrome.

OBSERVATION

A 46-year-old patient, suffering since 2015 from chronic polyradiculoneuropathy, is admitted in our department for progressive abdominal distension, associated with cramp-like abdominal pain evolving for several days before admission. These abdominal signs were associated with paresthesia in the lower and upper limbs accompanied by asthenia and weight loss.

Clinical examination found a patient with WHO Performance status grade 2, BMI = 22 kg / m2, edema of the lower limbs, ascites of average abundance, splenomegaly, soft-edged and regular Hepatomegaly (liver size=14cm), multiple homogeneous bilateral axillary and inguinal adenonapthies, peripheral neurogenic syndrome, with darkened areolae and white nails.

The biological assessment showed a hypoalbuminemia, the ascites fluid was rich in protein, with IgA lambda monoclonal gammopathy. Abdominopelvic CT scan showed hepatomegaly, splenomegaly, abdominal lymphadenopathies and lytic vertebral and rib lesions.

EMG was in favor of severe peripheral neurogenic damage. *CSF* analysis revealed *albuminocytologic dissociation*. Papilledema and hypothyroidism were also discovered in our patient.

The diagnosis of POEMS syndrome was established in the presence of two mandatory major criteria (monoclonal plasma cell disorder, peripheral neuropathy) associated to several minor criteria (organomegaly, ascites, Papilledema, skin changes and hypothyroidism). Dexamethasone was the initial treatment used for our patient then it was combined to thalidomide. The clinical outcome was good with resolution of the ascites and improvement of the neuropathy.

DISCUSSION

The POEMS syndrome is more common in Japan than in Western countries according to Dispenzieri *et al*., [1] and Nakanishi *et al*., [2]. The male predominance is reported, the average age of onset is between 40 and 50 years. Typically, the POEMS syndrome combines the five main signs contained in the acronym POEMS. Some authors require the presence of two major criteria (monoclonal plasma cell disorder, peripheral neuropathy) with a minor criterion among the following criteria: Sclerotic bone lesions, Castleman's disease, organomegaly, extravascular volume overload (edema, pleural effusion, or ascites), endocrinopathy (adrenal, thyroid,b pituitary, gonadal, parathyroid, and pancreaticb), skin changes (hyperpigmentation, hypertrichosis, angiomas, white nails) Papilledema. These criteria were proposed by the Dispenzierri *et al*., [1] team. Polyneuropathy is the constant element, found in 100% of cases in the series of Nakanishi *et al*., [2]. It is a chronic polyradiculoneuropathy manifested by paresthesia of the feet (71%), pain (12%), gait disturbances (17%), areflexia (68%), and proprioception disorders (78%). In the Soubrier [4] review, polyneuropathy is often the revealing sign. In our case, the neurological signs were inaugural. Apart from the five main manifestations of the POEMS syndrome, the clinical presentation is very polymorphic, as it has been demonstrated in most studies [1-3] which may include: febrile syndrome, a change in general condition, renal failure, stroke, diarrhea, pleural effusions, *Primary pulmonary hypertension* (PPH) and heart failure. Soubrier, M [3] insisted that patients with unexplained peripheral neuropathy should benefit from electrophoresis and immunoelectrophoresis in search of a monoclonal component pointing towards this diagnosis to reduce the delay in therapeutic management. Treatment with thalidomide or lenalidomide, prescribed by Tomás *et al*., [5], has been successful with decreases in VEGF and neuropathy. Our patient was treated with dexamethasone followed by thalidomide with a good clinical response. The combination melphalan dexamethasone has given good results in some cases as it was presented in the Méndez-Herrera *et al*., review [6]. The bone marrow transplant would be indicated in forms refractory to corticosteroid therapy, chemotherapy and radiotherapy. Bevacizumab, an anti-VEGF monoclonal antibody, dramatically improved 2 patients but worsened another [4].

CONCLUSION

POEMS syndrome is a rare but serious disease. Therefore, it is necessary to think about electrophoresis and immunoelectrophoresis, especially in patients with unexplained peripheral neuropathy wich can help establish the diagnosis and reduce the diagnostic and therapeutic delay.

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