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Pleural effusion revealing a rare haematological disease

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ABSTRACT: *Introduction:* POEMS syndrome consisting of polyneuropathy (P), organomegaly (O), endocrinopathy (E), monoclonal gammopathy (M) and skin changes (S), is a rare paraneoplastic syndrome resulting from an underlying plasma cell disorder. The multi-organ involvement associated with this syndrome extends beyond those listed in the acronym, such as extravascular volume overload (pleural effusion, ascites, oedema), sclerotic bone lesions, papilledema and thrombocytosis, and not all of the features listed in the acronym are required for diagnosis. *Observation:* we report a rare case of POEMS syndrome in an 83-year-old man with bilateral pleural effusions, initially attributed to pleural tuberculosis given the endemic context. *Conclusion:* the pleural involvement in patients with POEMS in our endemic tuberculosis context makes diagnosis difficult, delaying treatment. However, a good history and physical examination followed by appropriate investigations can differentiate this syndrome from other diseases.

KEYWORDS: POEMS Syndrome, pleural effusion, tuberculosis, Morocco.

1 INTRODUCTION

POEMS syndrome, consisting of polyneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell disease and skin changes, is a rare multisystem auto-inflammatory disorder described by Bardwick in 1980 [1].

Other features of this disease not included in the acronym, such as extravascular volume overload, papilledema, sclerotic bone lesions, thrombocytosis/erythrocytosis, elevated VEGF levels, abnormal pulmonary function tests and multicentric Castleman disease [2, 3], may be early signs of the disease.

This was the case in our patient, whose diagnosis of POEMS was made on the basis of bilateral pleural effusions in the context of endemic tuberculosis, which made the diagnosis difficult.

We would like to report this rare case and invite clinicians to consider the whole clinical picture, but also to work in multidisciplinary groups.

2 OBSERVATION

An 83-year-old man with a history of treated hypertension presented with a bilateral pleural effusion associated with asthenia, which had developed 3 months prior to admission, and who had previously been diagnosed with tuberculosis based on the epidemiological context, a positive quantiferon and an exudative pleural fluid with a lymphocytic predominance.

The initial pleural biopsy was inconclusive. The patient was started on anti-tuberculosis treatment with worsening clinical symptoms.

After discussion in the multidisciplinary consultation, a POEMS syndrome was retained due to:

- Sensory-motor deficit in both lower limbs with EMG confirmation of the demyelinating nature of the neuropathy,
- Monoclonal gammopathy (IgG lambda),
- Hyperpigmentation predominantly on the extremities with white, ridged nails,
- Extravascular volume overload (bilateral pleural effusions with dyspnea, peripheral oedema and minimal pericardial effusion),
- Bone marrow had 12% lambda monoclonal plasma cells,
- Plasma VEGF 1179 ng/ml.

A thoracoscopy was performed due to suspected tuberculosis and showed a thickened pleura with septal effusion without suspicious lesions. Histology was consistent with subacute inflammatory remodelling. Complement PCR testing on a pleural fragment was negative, ruling out evolving pleural tuberculosis.

Due to his advanced age and poor performance status, the patient was deemed ineligible for high-dose chemotherapy followed by peripheral blood stem cell transplantation.

He was started on Lenalidomide 25 mg daily for 21 days of a 28-day cycle with once-weekly Dexaméthasone 20 mg.

By his third cycle of therapy, his pleural effusion and oedema had completely resolved, his performance status had significantly improved, and a control EMG showed a 25% improvement in his neuropathy. After six months of treatment, VEGF levels decreased from 1170 pg/ml to 314 pg/ml.

3 DISCUSSION

POEMS syndrome is a rare disorder and its global incidence is unknown. It is considered to be a disease of Asian descent, mainly in China and Japan, with an approximate incidence of 0.3 per 100,000 in Japan [3]. However, many series have been published in Europe and the USA in recent years [4, 11].

The multisystem involvement of POEMS and its clinical polymorphism (the constellation of non-specific signs and symptoms) affecting different species usually delay diagnosis, with a median of 13-18 months [5, 8] between onset of symptoms and diagnosis of POEMS [3].

As in our patient, the presenting sign was a bilateral exudative pleural effusion, which delayed the diagnosis in the context of tuberculosis endemia.

The incidence of pleural effusion in POEMS syndrome is variable, ranging from 34.7% in the YONG WANG study to 50% in the TOMOKI study [7, 9]. The pathogenesis of pleural effusion in POEMS syndrome remains unclear, although a possible role for cytokines such as vascular endothelial growth factor (VEGF) in endothelial permeability has been suggested [1, 3].

In our Moroccan context of endemic tuberculosis, any exudative pleural effusion makes tuberculosis the first diagnosis. Indeed, the incidence of tuberculosis in Morocco is 94 cases per 100,000 inhabitants in 2021 [10], with a trend towards extrapulmonary forms, where pleural involvement is second only to lymph node involvement. As pleural TB is paucibacillary, it is more difficult to diagnose with certainty and pleural biopsy, the gold standard for diagnosis, is only conclusive in 50-60% of cases [6].

As in the case of our patient who presented with a worsening of his pleural effusion under antibacillary treatment, we performed a thoracoscopy to confirm the diagnosis of pleural tuberculosis and the biopsies came back in favour of an inflammatory remodelling and the absence of tuberculosis.

The diagnosis of POEMS syndrome was made on the basis of the peripheral neuropathy and the presence of the monoclonal protein on serum protein electrophoresis.

In a retrospective Chinese study of 96 patients with POEMS syndrome, pleural effusion was the most common chest involvement with an incidence of 42.7% (41 of 96 patients) and all effusions were exudative [8]. An incidence that is still quite significant and why not revise the diagnostic criteria for POEMS syndrome?

This interesting result [7, 8, 9] makes us to think about other diagnoses of exudative pleural effusion, especially the rarer ones, which have a prognostic impact on patients despite the context of endemic tuberculosis, and not to hesitate to revise the initial diagnosis in case of a non-response to antibacillary treatment or in the absence of solid arguments to maintain the diagnosis of pleural tuberculosis.

4 CONCLUSION

POEMS syndrome is potentially fatal and patients' quality of life is compromised by progressive neuropathy, massive peripheral oedema, pleural effusion and ascites. We have reported this case to remind clinicians of this rare condition and to prevent misdiagnosis.

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