POEMS Syndrome: an uncommon cause of pleural effusion

Clarissa Canella[1,2], Bruno Schau[3], Edson Marchiori[4]

A 38-year-old man presented with dyspnea, progressively worsening back pain, and numbness and weakness in his feet and hands. Physical examination revealed hepatosplenomegaly and diffuse skin thickening with generalized hyperpigmentation. Laboratory analysis showed hypothyroidism and hyperprolactinemia. Computed tomography revealed left pleural effusion, hepatosplenomegaly, and multiple sclerotic lesions involving the axial skeleton (Figure 1). The final diagnosis was POEMS syndrome.

POEMS syndrome is a multisystemic disease occurring secondary to plasma cell dyscrasia. The acronym stands for a range of distinct features: polyneuropathy (P), organomegaly (O), endocrinopathy (E), monoclonal plasma-cell proliferative disorder (M), and skin changes (S). Many other clinical signs, such as sclerotic bone lesions, papilledema, ascites, pleural effusion (related to extravascular volume overload), pericardial effusion, pulmonary hypertension, Castleman disease, thrombocytosis and erythrocytosis, and an increased serum vascular endothelial growth factor level, may be present. POEMS syndrome is diagnosed based on the presence of polyradiculoneuropathy, clonal plasma cell disorder, and at least one additional major criterion and one minor criterion. The disease is potentially fatal, and the patients' quality of life deteriorates due to progressive neuropathy, massive peripheral edema, pleural effusion, and ascites.[1,2] Clinicians who encounter pleural effusion and some of these other signs should be aware of the possibility of POEMS syndrome.

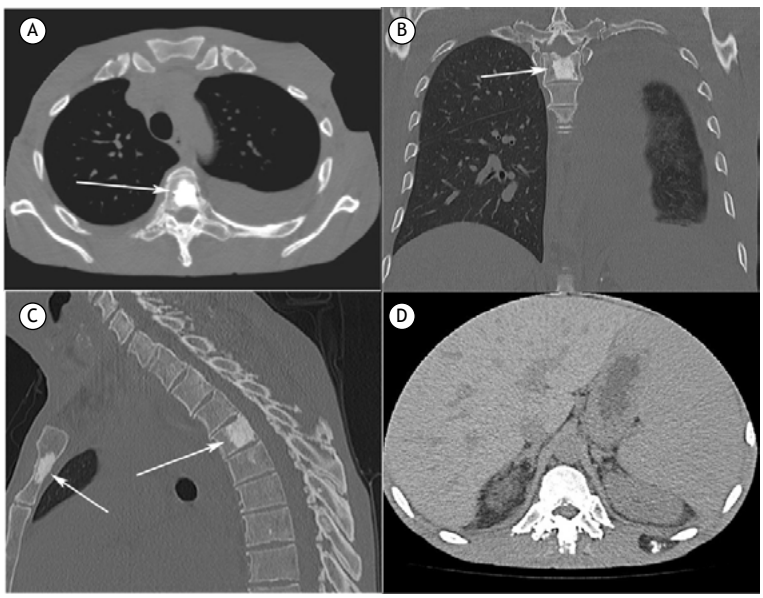

Figure 1. Computed tomography performed with mediastinal window settings and axial (A), coronal (B), and sagittal (C) reconstruction showing left pleural effusion. Note also the presence of multiple sclerotic lesions in the vertebral bodies and sternum (arrows). (D) Axial view of the upper abdomen demonstrating hepatosplenomegaly.

REFERENCES

1. Dispenzieri A. POEMS Syndrome: 2019 Update on diagnosis, risk-stratification, and management. Am J Hematol. 2019;94(7):812-827. https://doi.org/10.1002/ajh.25495.
2. Nozza A. POEMS Syndrome: An Update. Mediterr J Hematol Infect Dis. 2017;9(1):e2017051. https://doi.org/10.4084/MJHID.2017.051.

1. Clínica de Diagnóstico por Imagem, Rio de Janeiro (RJ), Brasil.
2. Universidade Federal Fluminense, Rio de Janeiro (RJ), Brasil.
3. Hospital Federal dos Servidores do Estado, Rio de Janeiro (RJ), Brasil.
4. Universidade Federal do Rio de Janeiro, Rio de Janeiro (RJ), Brasil.