Osteolytic Lesion with Thick Sclerotic Walls and Demyelinating Polyneuropathy

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Systemic symptoms such as organomegaly and endocrine manifestations, in the presence of a peripheral neuropathy, may alert the clinician to POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes).

A 30-year-old man was admitted for the recent onset of a demyelinating polyneuropathy. Clinical and laboratory investigations disclosed lower limb edema, hepatosplenomegaly and lymphadenopathy, gynecomastia associated with hyperprolactinemia, hypothyroidism, adrenal and gonadotropic insufficiencies, and bilateral papilledema. Monoclonal gammopathy IgA λ was present. POEMS syndrome was suspected. Serum vascular endothelial growth

factor (VEGF) level was 953 pg/ml (normal < 115). Pelvis radiography (Figure 1) showed an expansile osteolytic lesion mainly in the left iliopectic ramus, contrasting with preserved or thickened cortical margins and thick sclerotic intralésional walls. Three other small lytic lesions with a sclerotic rim were observed. Biopsy of the largest bone lesion showed a massive infiltration by atypical plasma cells (Figure 2), which were clonal lambda on immunostaining (Figure 3). POEMS was confirmed and, because there were > 2 bone lesions, chemotherapy was started, as well as cementoplasty of the large osteolytic lesion^{1,2}.

In the presence of a peripheral neuropathy, systemic symptoms such as organomegaly and endocrine manifesta-

Figure 1. Osteolytic lesion in the left iliopectic ramus with marked thick sclerotic walls.

Figure 2. Biopsy showed a massive infiltration by atypical plasma cells (original magnification $\times 400$).

Figure 3. Plasma cells showed clonal lambda on immunostaining (original magnification $\times 400$).

tions should suggest POEMS syndrome³ and lead to bone radiograph, VEGF serum level measurement, and a search for monoclonal gammopathy.

REFERENCES

1. Dispenzieri A. POEMS syndrome: 2011 update on diagnosis, risk-stratification, and management. *Am J Hematol* 2011;86:591-601.
2. Dispenzieri A. How I treat POEMS syndrome. *Blood* 2012;119:5650-8.
3. Bardwick PA, Zvaifler NJ, Gill GN, Newman D, Greenway GD, Resnick DL. Plasma cell dyscrasia with polyneuropathy, organomegaly, endocrinopathy, M protein, and skin changes: the POEMS syndrome. Report on two cases and a review of the literature. *Medicine* 1980;59:311-22.