

POEMS syndrome: a puzzling case

Lopes C¹, Costa T¹, Mateus M², Branco JC¹

ACTA REUMATOL PORT. 2019;44:84-85

A 63-year-old woman with a personal history of asthma, arterial hypertension and dyslipidemia was admitted to the Rheumatology Department. She had a record of 14 years of random polyarthralgias, fatigue, myalgias, distal paresthesias and muscle weakness of the upper and lower limbs with increasing walking difficulty. She also referred a progressive increase in abdominal volume and inguinal masses for the last 6 years. Physical examination revealed progressive hypesthesia, skin hyperpigmentation, hemangiomas (Figure 1), acrocyanosis, multiple peripheral lymph nodes and splenomegaly. Blood tests showed persistent thrombocytosis over the last 4 years, increased serum creatinine (1.50 mg/dL) and B2-microglobulin (4.56 mg/L). C-reactive protein and sedimentation rate were normal. Hormonal study showed a primary hypothyroidism and hyperprolactinemia. During the last 5 years, serum protein electrophoresis revealed monoclonal immunoglobulin G-lambda (IgG- λ) paraprotein. She had multiple radiographic blastic and lytic lesions (skull, sternum, dorsal and lumbar spine, sacrum, iliac wings, femurs and humerus) (Figure 2). She had been submitted to six myelograms and bone marrow biopsies, all revealing plasmacytosis of undetermined significance. A guided biopsy to a subtrochanteric lesion done 5 years before revealed plasma cells aggregates. Computerized tomography (CT) scans showed splenomegaly and lymphadenopathies (including the Waldayer ring, axillary and inguinal lymph nodes) for the last 6 years. Needle electromyography confirmed a polyneuropathy and measurement of vascular endothelial growth factor (VEGF) level was normal. A presumptive diagnosis of POEMS syn-



FIGURE 1. Glomeruloid hemangioma on the right thigh

drome was established based on the clinical, histopathological, imagiological and laboratorial findings. The patient was oriented to a reference center for appropriate follow-up and an autologous peripheral blood stem cell transplantation was performed with clinical improvement.

Polyneuropathy, organomegaly, endocrinopathy, M protein, and skin changes (POEMS) syndrome, also known as Crow-Fukase syndrome, osteosclerotic myeloma and Takatsuki syndrome, is a rare paraneoplastic syndrome^{1,2,3,4}. The diagnosis is based on having both the polyradiculoneuropathy and the monoclonal plasma cell disorder (almost always λ),

1. Rheumatology, Hospital de Egas Moniz, CHLO, Lisbon, Portugal; CEDOC, NOVA Medical School - Faculdade de Ciências Médicas - NOVA University of Lisbon, Lisbon, Portugal

2. Rheumatology, Hospital de Egas Moniz, CHLO, Lisbon, Portugal

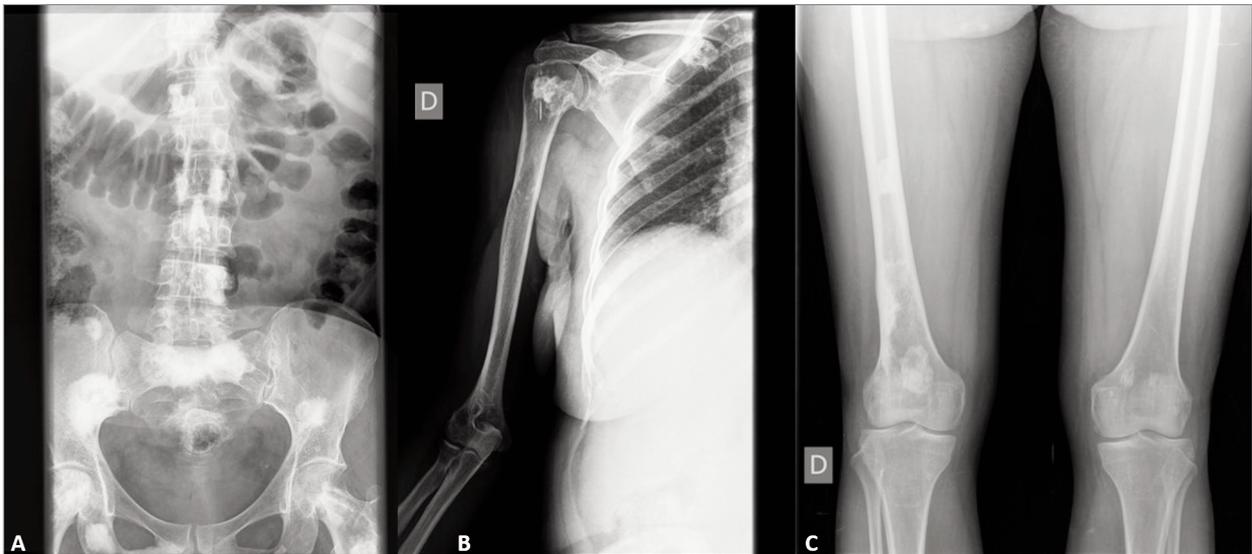


FIGURE 2. A) multiple foci of osteosclerotic lesions on plain radiograph in some vertebrae of dorsal and lumbar spine, sacrum and iliac wings; B and C) mixed lytic and osteosclerotic bone lesions on plain radiograph of femurs and humerus

at least one of the other three major criteria (Castleman disease, sclerotic bone lesions or elevated VEGF) and at least one minor criteria (organomegaly, extravascular volume overload, endocrinopathy, skin changes, papilledema or thrombocytosis/polycythemia)¹⁻⁴.

Among cutaneous manifestations, granulomas might appear in course of the disease as firm papular lesions, with erythematous or violet coloration, on the trunk and proximal limbs. They may present variable histological characteristics, the most common being cherry hemangioma, lobular capillary hemangioma and, less frequently, glomerular hemangioma.⁵

Bone lesions occur in approximately 95% of patients, and can be mistaken for benign bone islands, aneurysmal bone cysts, non-ossifying fibromas, and fibrous dysplasia^{1,4}. The characteristic findings are single or various, pure or mixed, osteosclerotic focal lesions: some densely sclerotic, others lytic with a sclerotic rim, and still others presenting a mixed soap bubble appearance^{1,2,4}. Bone windows of CT body images are often very enlightening. The lesions are found predominantly in the axial and proximal appendicular skeleton and may appear years before the diagnosis.²

The diagnosis of POEMS syndrome is challenging

due to its low prevalence, multiorgan involvement and diverse clinical manifestations². Treatment of POEMS syndrome depends on a multidisciplinary team preferentially in reference centers.

CORRESPONDENCE TO

Carina Lopes
Hospital Egas Moniz
R. da Junqueira 126, 1349-019 Lisboa
E-mail: carina.alves.lopes@gmail.com

REFERENCES

1. Dispenzieri A. POEMS syndrome: Update on diagnosis, risk-stratification, and management. *Am. J. Hematol* 2015; 90: 952-962.
2. Arana C, León J, Gómez-Moreno G, Pérez-Cano R, Hernández T. POEMS Syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal Gammopathy and Skin Changes) Treated with Autologous Hematopoietic Stem Cell Transplantation: A Case Report and Literature Review. *Am J Case Rep* 2015; 16: 124-129.
3. Dispenzieri A. POEMS syndrome. *Blood Review* 2007; 21: 285-299.
4. Li J, Zhou D. New advances in the diagnosis and treatment of POEMS syndrome. *British Journal of Haematology* 2013; 161: 303-315.
5. Dispenzieri A. How I treat POEMS syndrome. *Blood* 2012; 119:5650-5658.