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Clinical picture

QJM

Osteosclerotic myeloma with POEMS syndrome

Clinical presentation

A previously healthy 46-year-old man presented with erectile dysfunction for several months and a feeling of coldness in his legs and pain in his lower back for 2 weeks. On physical examination, the spleen was palpable 7 cm below the costal margin. Both skin hyperpigmentation and hypertrichosis were present on his extremities. The complete blood cell count with differential was normal. Serum total calcium and albumin were 2.2 mmol/l (normal: 2.02–2.6 mmol/l) and 4.1 g/dl (normal: 3.5–5.0 g/dl), respectively. Abdominal plain film radiography revealed multiple scattered osteosclerotic nodules, including in the lumbosacral spine, ribs, pelvic bones and upper femurs (Figure 1). Electromyographic studies showed prolongation of distal latencies and severe attenuation of compound muscle action potential in four limbs. Abdominal computed tomography revealed hepatosplenomegaly and normal prostate size without lymphadenopathy. IgG lambda monoclonal protein was detected by serum immunofixation test, and bone marrow study showed clonal plasma cells. Osteosclerotic myeloma with POEMS (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal protein, and Skin changes) syndrome was diagnosed. His symptoms improved after chemotherapy.

Originally described by Scheinker in 1938, and the acronym of POEMS was coined in 1980 by Bardwick *et al.*; POEMS syndrome (as known as Crow–Fukase syndrome, PEP syndrome, or Takatsuki syndrome) is a rare medical disorder, characterized by the presence of: (P) polyneuropathy, (O) organomegaly, (E) endocrinopathy, (M) M-protein and (S) skin abnomalities.[1,2] Several other associated diseases such as osteosclerotic myeloma or Castleman's disease are usually seen. Although it has been reported mostly in Asia areas, there are affected individuals from other ethnic backgrounds.[3]

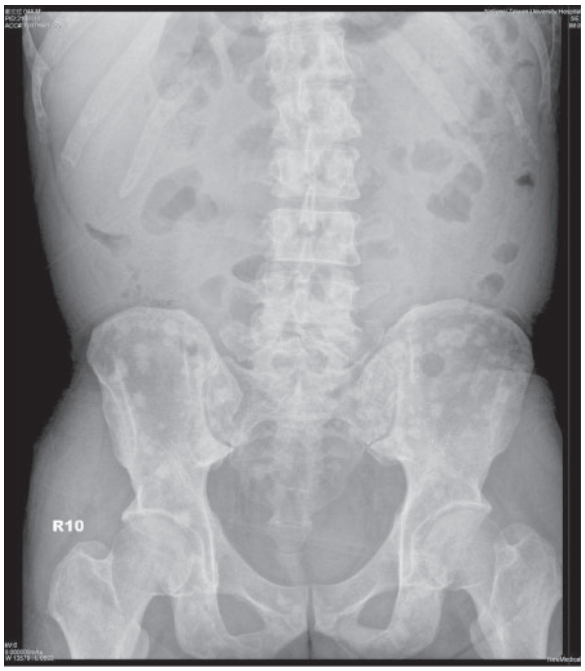

Figure 1. Abdominal plain film revealed multiple scattered osteosclerotic nodules, including in the lumbosacral spine, ribs, pelvic bones and upper femurs.

The exact pathophysiologic mechanism is still unknown and there is no single test that establishes the diagnosis of POEMS syndrome. The striking osteosclerotic changes revealed by plain film in our patient played an important role in the diagnosis. Including the lambda light chain, lots of cytokines have been observed increased in patients with POEMS syndromes, which may be associated with the pathogenesis.[1] The choice of treatment depends on the clinical conditions, including corticosteroid, radiation therapy and chemotherapy.

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References

1. Dispenzieri A, Kyle RA, Lacy MQ, Rajkumar SV, Therneau TM, Larson DR, *et al.* POEMS syndrome: definitions and long-term outcome. *Blood* 2003; **101**:2496–506.
2. 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 (Baltimore)* 1980; **59**:311–22.
3. Soubrier MJ, Dubost JJ, Sauvezie BJ. POEMS syndrome: a study of 25 cases and a review of the literature. French Study Group on POEMS syndrome. *Am J Med* 1994; **97**:543–53.