Internal Medicine Quiz – Case 5

A 60-year-old man was evaluated for arthritic complaints of recent onset. He was a smoker with 100 pack-years. His past medical history was notable for Paget's disease of bones (PDB), diagnosed 10 years before, due to low back pain, an elevated alkaline phosphatase and a positive bone scan. He had received biphosphonates and the disease was currently considered as inactive.

Currently, he complained of weight loss, malaise, shortness of breath and redness, oedema and morning stiffness of elbows, knees, and metacarpophalangeal joints of both hands.

On presentation he had a low grade fever (37.3 °C) and arthritis of the elbows, knees, shoulders and of the metacarpophalangeal and interphalangeal joints of both hands, with markedly reduced range of motion of the affected joints. Skeletal deformities were not present, while generalized muscular atrophy was evident. No hepatomegaly, splenomegaly or lymphadenopathy were noted.

An initial laboratory evaluation revealed anemia (Ht 30%, with hypochromic [MCH 23 pg] and microcytic [MCV 76 fL] indices), diffuse hyperglobulinemia, stool positive for occult blood, low serum iron (20 μg/dL) and serum ferritin (50 ng/mL). Inflammatory indices were elevated (erythrocyte sedimentation rate of 80 mm/hour and CRP at 25 mg/dL), alkaline phosphatase was 227 IU/L and tumor markers were negative. The rheumatoid factor was elevated at 75 IU/mL, while testing for anti-cyclic citrullinated peptide antibodies and antinuclear antibodies was negative.

A tuberculin skin reaction was negative. A lip biopsy was negative for lymphocytic infiltration. Gastroscopy revealed hiatus hernia with esophagitis, antral and gastric body gastritis. The histologic study of the gastric biopsies showed infiltration by a gastric MALT lymphoma (Wotherspoon score index 5). On colonoscopy hemorrhoids were noted. Bone marrow smear did not reveal infiltration from lymphoproliferative disease. Findings consistent with PDB were noted on bone marrow biopsy.

CT scans of the abdomen and thorax revealed no pathologic findings except diffuse lytic and regenerative changes of the bones of the pelvis and the spine (fig. 1), compatible with PDB.

X-rays of the joints, pelvis, and skull were obtained and showed images typical of PDB as well as an erosive arthritis (figures 2, 3).

Comment

Inflammatory arthritis is not a feature of PDB, while arthritic pain is usually not prominent and may be caused directly by a pagetic lesion involving para-articular areas or, more often, indirectly from complications stemming from the abnormal bone, such as degenerative arthritis, nerve impingement. Pain may also herald the development of neoplasia, such as osteosarcoma or benign giant cell tumors, which occur in 0.7–1.0% of cases and can have an extraskeletal location. The coexistence of gastric MALT lymphoma and asymptomatic Paget's disease of bones has been also reported previously.

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