Pancreatitis, panniculitis, and polyarthritis syndrome complicated with systemic intraosseous fat necrosis

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Dear Editor,
The patient was a 54-year-old male with alcoholism and multiple episodes of pancreatitis. Abdominal pain reappeared in November 2022, with severe systemic infection and malnutrition, along with subcutaneous nodules and knee pain. Computed tomography (CT) of the knee in November 2022 (Fig. 1A) showed effusion in both knee capsules and no signs of bone destruction. The final diagnosis was “pancreatitis, panniculitis and polyarthritis syndrome” (PPP). After treatment, the symptoms of pancreatitis and panniculitis resolved, but gradually swelling and pain developed in the bilateral ankle, shoulder, elbow and some metacarpophalangeal joints. Joint pain persisted, which was temporarily relieved by glucocorticoids but not cured. Four months later, the patient was readmitted to the hospital, and while at home, he had two mild episodes of acute pancreatitis. At this point, the patient’s
joint swelling mitigated, however, the pain symptoms remain unchanged, with significant impairment of movement of the limbs and significant pain to the touch. The symptoms were most severe in the left knee. Magnetic resonance imaging (MRI) of the left knee (March 28, 2023) (Fig. 1B) indicated osteonecrosis and osteomyelitis. Considering similar symptoms in multiple joints of the patient’s body, whole-body 99mTc-MDP bone scan (April 5, 2023) (Fig. 1C) revealed abnormal tracer uptake in the mandible, limbs and certain joints.

Discussion

PPP syndrome is relatively rare and was first reported in 1973. Pathogeny includes acute and chronic pancreatitis, pancreatic tumors, pancreatic pseudocysts and pancreatic trauma. The pathogenic factor is related to pancreatic enzymes (1). The polyarthritis of PPP syndrome is characterized by multiple serositis and necrosis of intraosseous fat, and it typically presents as symmetric or asymmetric with involvement of multiple joints (2). In the majority of patients, polyarthritis symptoms resolve upon improvement of pancreatic disease. However, a minority may develop chronic arthritis. MRI can be used for early detection of intraosseous fat necrosis, bone marrow edema and periarticular soft tissue panniculitis (3). 99mTc-MDP bone scan also plays an important role in diagnosing systemic intraosseous fat necrosis (4). The treatment of PPP syndrome should focus on the treatment of pancreatic disease, and we believe that frequent episodes of pancreatitis were the main cause of the persistent polyarthritis symptoms in this patient, which continued to progress even after this patient’s pancreatic enzymes returned to normal. This implies that once extra-pancreatic complications are triggered in patients with PPP syndrome, the complications may continue to progress and do not resolve with remission of the pancreatic disease.

References


Fig. 1. A. Computed tomography (CT) scan of the knee. B. Left knee magnetic resonance imaging (MRI). C. 99mTc-MDP bone scan; increased tracer uptake was observed in the mandible, bilateral shoulder, bilateral humerus and bilateral femur, and decreased in the bilateral elbow, hip, knee and ankle.