Lupus-associated pancreatitis complicated by pancreatic pseudocyst and central nervous system vasculitis

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Systemic lupus erythematosus (SLE) is an autoimmune disease that can affect various organ systems, including the skin and mucous membranes, joints, kidney, brain, serous membranes, lung, heart and occasionally the gastrointestinal tract. Single organ systems may be involved or they can be involved in any combination. SLE may also involve the pancreas and in rare cases it may cause acute pancreatitis with a broad spectrum of complications. We report on a case of a female patient with SLE who developed acute pancreatitis with pseudocyst formation, subsequently complicated by central nervous system (CNS) vasculitis. Abdominal computed tomography (CT) showed an edematous swelling of the pancreas and a pseudocyst measuring 4×3 cm. Brain magnetic resonance imaging (MRI) showed multiple high signal intensity lesions in both cerebral hemispheres. The pseudocyst did not completely resolve with high-dose steroid therapy and it was later complicated by infection and rupture. After a surgical drainage for the complicated pseudocyst, her clinical symptoms and signs were markedly improved. This case shows that prolonged observation of pancreatic pseudocysts with the expectation of spontaneous resolution may expose the SLE patient to unwarranted risks (e. g., infection or rupture). Early drainage for pancreatic pseudocyst might be considered and especially in a patient with lupus-associated pancreatitis.

Autoimmune lymphoproliferative syndrome-like syndrome presented as lupus-like syndrome with mycobacterial joint infection evolved into the lymphoma

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The autoimmune lymphoproliferative syndrome (ALPS) and ALPS-like syndrome are variable clinical conditions characterized by lymphoproliferative disease, autoimmune cytopenias and susceptibility to malignancy. Case: A 59-year old woman was admitted to the hospital for intractable generalized pain and stiffness with multiple swollen joints for 2 weeks. A low-grade fever, intermittent hypotension and confusion were associated with the pain. The evaluation revealed multiple joint bony erosions with effusion and a ruptured Baker’s cyst and positive AFB testing on the joint biopsy of the right wrist. In addition, there were a macular skin rash with telangiectasia and perivascular lymphocyte infiltration, a cytopenia without abnormal cells, a hepatosplenomegaly, a pericardial thickness with effusion and pleural effusion. The patient was treated with anti-mycobacterial drugs, NSAIDs and glucocorticoids for 10 months. But with the symptoms worsening the patient developed cervical lymph node enlargements and was diagnosed as a diffuse large B cell lymphoma with hemophagocytosis on biopsy. The patient had acceleration of the pancytopenia, jaundice and confusion despite a normal brain MRI and neurology examination. The patient underwent emergency R-CHOP chemotherapy for HLH associated with malignant lymphoma. This case illustrates the clinical course resembling an autoimmune lymphoproliferative syndrome presenting with a lupus-like syndrome and mycobacterial joint infection that evolved to lymphoma with hemophagocytic lymphohistiocytosis.