A patient with Castleman’s disease mimicking acute pancreatitis

Key words: Castleman’s disease. Giant lymph node hyperplasia. Acute pancreatitis.

DOI: 10.17235/reed.2017.5120/2017

Dear Editor,

Castleman’s disease is a rare condition that gastroenterologists should be aware of. We present a case of Castleman’s disease mimicking acute pancreatitis.

Case report

A 34-year-old man presented with epigastric pain, fever and abdominal distention for five days. There was no history of alcohol abuse or drug abuse. Physical examination showed multiple cervical, axillary and inguinal lymphadenopathy. Laboratory tests showed increased levels of inflammatory markers (white cell count of 17.19 × 10^9/l, 83.1% neutrophils and C-reactive protein of 92 mg/l) and serum amylase was normal. Rheumatologic, infectious and viral tests were all negative. Computed tomography (CT) revealed acute edematous pancreatitis, massive ascites, pelvic, perihepatic and pericardial effusion (Fig. 1A and B). There was no evidence of gallstones and no other underlying etiological factors of acute pancreatitis were detected. The patient was treated with antibiotics. The fever and abdominal pain subsided but the abdominal distention did not improve. An excisional biopsy from the inguinal lymph node revealed mixed forms of Castleman’s disease (Fig. 1C). Immunohistochemical analysis showed expression of CD3, CD20, CD21 and CD38. The patient underwent six courses of CHOP chemotherapy. The symptoms disappeared and a repeat CT showed absorption of the above effusion. The patient remains well and there was no recurrence after one year of follow-up.

Discussion

Castleman’s disease (CD) is a rare lymphoproliferative disorder and its pathogenesis remains poorly understood (1). On a clinical basis, the disease is separated into uni-centric CD and multi-centric CD, and it is pathologically classified as hyaline
vascular variant (HVV), plasma cell variant (PCV) and mixed forms as both HVV and PCV (1,2). CD involving the pancreas is extremely rare and, to date, around 20 cases have been reported. However, almost all these cases presented as a pancreatic mass or tumor, and over half of them were HVV (3,4). To our knowledge, this is the first reported CD case presenting as acute pancreatitis. The acute pancreatitis and multiple effusion in the present case may be attributed to the acute systemic inflammatory reaction caused by excessive proinflammatory hypercytokinemia in the setting of multi-centric CD (5). This case is reminiscent of acute pancreatitis with multiple lymphadenopathy that should raise a suspicion of CD.

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References