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CASE REPORT

ACUTE PANCREATITIS CAUSED BY PRIMARY PANCREATIC LYMPHOMA IN A GERIATRIC PATIENT WITH SARCOIDOSIS: REPORT OF A COMPLEX CASE

ABSTRACT

The purpose of reporting this case is to raise the awareness of the rapid worsening clinical presentation of acute pancreatitis in an older patient, unexpected encounter with pancreatic lymphoma, and sarcoidosis-lymphoma syndrome. An 80-year-old woman was diagnosed with sarcoidosis 16 years ago. Following recent hospitalization because of acute pancreatitis, non-obstructive and non-invasive pancreatic mass was found on radiological images. Histopathology confirmed the mass to be diffuse large B-cell lymphoma. We report this case because of its complicated the etiology of acute pancreatitis in an older patient, presentation of pancreatic lymphoma, and immune pathogenesis of sarcoidosis-lymphoma syndrome.

Key Words: Pancreatitis; Lymphoma, Non-Hodgkin; Sarcoidosis

OLGU SUNUMU

SARKOİDOZLU GERİATRİK HASTADA PRİMER PANKREATİK LENFOMANIN SEBEP OLDUĞU AKUT PANKREATİT: KOMPLEKS BİR OLGU SUNUMU

Öz

Bu olgu sunumunun amacı, yaşlı bir hastada hızla kötüleşen akut pankreatit kliniğine, pankreatik lenfoma ile beklenmedik karşılaşmaya, sarkoidoz-lenfoma sendromuna dikkat çekmektir. 16 yıl önce Sarkoidoz tanısı alan 80 yaşındaki kadın hastada, akut pankreatit tanısı ile hastaneye yatışını takiben, radyolojik incelemede tıkayıcı veya invaziv olmayan pankreatik kitle saptandı. Histopatolojik inceleme kitlenin B hücreli lenfoma olduğunu gösterdi. Yaşlı bir hastadaki akut pankreatitin kompleks etiyolojisi, pankreatik lenfomanın ortaya çıkışı ve sarkoidoz-lenfoma sendromunun immün patogenezi sebebiyle bu olguyu sunulmuştur.

Anahtar Sözcükler: Pankreatit; Lenfoma, Non-Hodgkin; Sarkoidoz

INTRODUCTION

The World Health Report has emphasized the acceleration of the aging of the population worldwide, with a marked increase in age related diseases in many countries. Here we report a case of an 80-year-old woman who developed B cell non-Hodgkin lymphoma subsequent to 16 years after the first diagnosis of sarcoidosis. She then presented to the hospital with acute severe pancreatitis. This provides evidence of a rare and complicated circumstance relating to a complex combination of these clinical entities.

CASE REPORT

An 80-year-old woman was admitted to the emergency department of a hospital with progressively worsening abdominal pain and nausea. Physical examination revealed significant abdominal tenderness and distention on palpation with no rebound. She was diagnosed with sarcoidosis 16 years previously, she had presented with bilateral hilar lymphadenopathy and mediastinal and lung parenchyma symptoms. She was treated with a low dose steroid for an unknown time period. She underwent regular clinical and radiological follow-up examinations that revealed no relapse or progression of the disease. Following hospitalisation, initial laboratory tests showed elevated levels of amylase 386 U/L (normal reference range 20-160U/L), lipase (1424 U/L (8-78U/L) and C-reactive protein (150 mg/dL (0-0.5mg/dL) with normal liver and kidney function test results. The preliminary diagnosis was acute pancreatitis, with low severity scores and good prognosis according to Ranson's criteria, Acute Physiology and Chronic Health Evaluation (Apache) II score and Bedside Index of Severity in Acute Pancreatitis. Subsequently, urgent treatment was provided. Abdominal computed tomography (CT) revealed bulky, homogeneous, diffuse infiltrative mass extending from the pancreatic head and corpus to the gastro-colic ligament, paraaortic and paracaval areas, and left kidney and its vein

(Figure 1). The size of the mass was approximately 120x78x96 mm. There was no pancreatico-biliary duct dilatation, although the mass was surrounding the adjacent celiac trunk, splenic artery, hepatic artery and the proximal side of the portal vein without invasion (Figure 2). Furthermore, there was no peripheral lymphadenopathy except for stable paratracheal and subcarinal lymphadenopathies seen on thoracic CT compared with previous scans. There were also high levels of lactate dehydrogenase [LDH, 1674 U/L (125-245U/L)] and beta-2 microglobuline [6.29 mg/L (0.9-2.6 mg/L)] along side a large non-obstructive and non-invasive pancreatic mass. Furthermore, immunoglobulin G4 (IgG4) was within normal limits and cancer antigen 19-9 was mildly elevated at 73 IU/mL (0-37 IU/mL). CT-guided tru-cut biopsy of the pancreatic mass was performed to elucidate the underlying cause. Histopathologic analysis revealed a diffuse large B-cell lymphoma and immunohistochemistry of the specimen demonstrated cells positive for CD19 and CD20 but negative for CD3 (Figure 3). Before histopathological diagnosis, steroid therapy was administered based on radiological results that suggested pancreatic lymphoma. However, there was no reduction in the severity of pancreatitis following treatment. Subsequently, the patient died of rapid and irreversible pulmonary and renal system failure 10 days after the diagnosis of pancreatitis.

Ethical approval for this case was received from the local ethics committee of our institute. A written informed consent was obtained from her parents before this report was prepared.

DISCUSSION

This is an interesting case, although it is complicated in many aspects. First, the gallstone is the most common etiology of acute pancreatitis in older patients, whereas, this patient presented with primary pancreatic lymphoma (PPL), an uncommon cause of acute pancreatitis. PPL accounts for only 1% of extranodal lymphomas and 0.5% of all pancreatic masses (1). The rare and often subclenic



characteristics of PPL make it a diagnostic challenge (2). Primary pancreatic lymphoma concurrent with acute pancreatitis, as in the present study patient, is an infrequent presentation. Several factors were more suggestive of PPL rather than pancreatic adenocarcinoma or lipomatous pseudohypertrophy. These included the presence of normal bilirubin and enzymes of cholestasis, along with the absence of pancreatic and biliary duct dilatation despite the large size of mass. Furthermore, there was no heavy fat composition and no invasion or occlusion of the abdominal vascularity (3,4). The absence of pancreatic calcification and necrosis provided additional evidence for the diagnosis of PPL.

Several existing reports suggest that patients with sarcoidosis have a higher risk of developing lymphoma in later life. The patient's history of sarcoidosis would, therefore, support diagnosis of pancreatic lymphoma regarding sarcoidosis-lymphoma syndrome (5,6). While it remains controversial, it is noteworthy that there was a long interval between the two diagnoses in our patient, which might provide a possible basis for a proposed immunopathogenic model. Our patient was

evaluated both in terms of autoimmune pancreatitis and sarcoidosis with pancreatic involvement. Several analyses showed normal serum IgG4 level, no plasma cell infiltration, and storiform fibrosis or obliterative phlebitis. This alongside no other organ involvement made a diagnosis of IgG4 related pancreatitis unlikely. The incidence of gastrointestinal involvement in sarcoidosis is less than 1.0%, and the patients with pancreatic involvement have also rarely been reported (5). In this patient with pancreatic involvement was excluded because no peripancreatic lymphadenopathy or granulomatous inflammation was observed on histopathological findings.

It is important to evaluate the severity of pancreatitis as early as possible in order to decrease mortality rates. High serum LDH and older age are independent risk factors of severity and poor outcome of acute pancreatitis according to several clinical scoring systems. However, the presence of malignancy with a high cell turnover may be another negative predictive sign for pancreatitis in older patients.

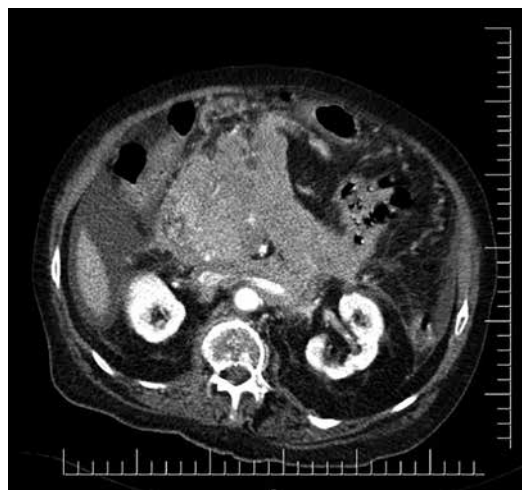


Figure 1. Abdominal computerised tomography imaging revealed a mass extending from the pancreatic head and corpus to the gastro-colic ligament, paraaortic and paracaval areas, and left kidney and its vein.

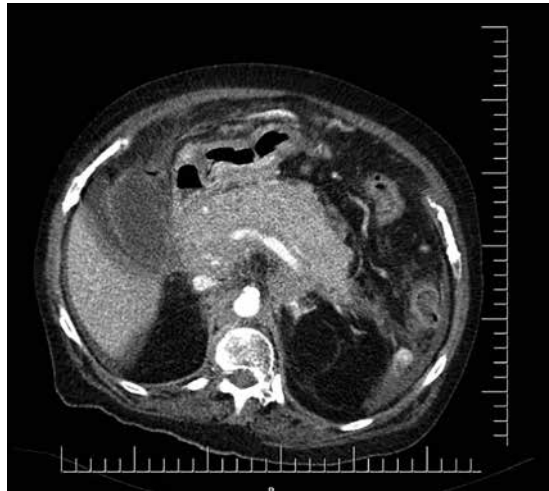


Figure 2. No pancreatic duct dilatation or vessel invasion was observed, despite the presence of a large pancreatic mass seen on abdominal computerised tomography imaging.

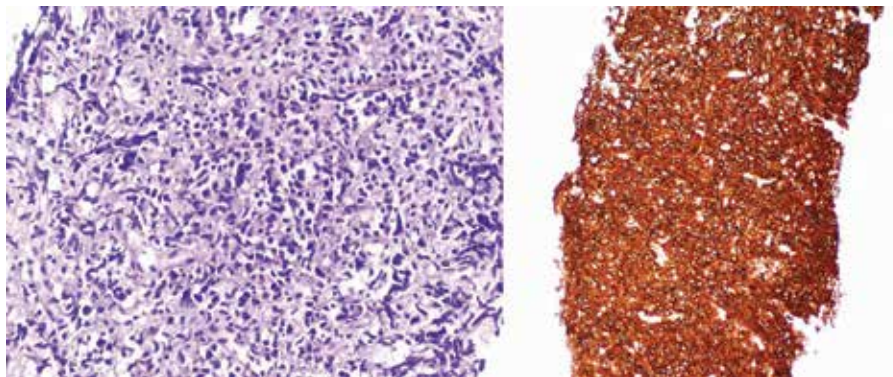


Figure 3. Left: the tumor was composed of large lymphoid cells with hyperchromatic nuclei and membrane-bound nucleoli (hemotoxylin-eosin, x200). **Right:** neoplastic cells showed diffuse membrane staining for the pan B-cell marker, CD 20 (CD 20, x100).

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