

Polyarteritis nodosa presenting with necrotising appendicitis and hepatic aneurysm rupture

Nekrotizan apandisit ve hepatic anevrizma rüptürü ile birlikte seyreden bir poliarteritis nodosa olgusu

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Polyarteritis nodosa is a necrotizing vasculitis which affects small and medium-sized arteries. The clinical features of the disease vary according to the site and extent of involvement. Aneurysmal dilatation and thrombosis in the arteries of the liver and gastrointestinal tract have been reported in polyarteritis nodosa. However, rupture of the hepatic aneurysm and necrotizing appendicitis due to polyarteritis nodosa are uncommon; indeed a case in which these have occurred together has not been reported to date. In this case report, a patient with necrotizing appendicitis and hepatic aneurysm rupture due to underlying polyarteritis nodosa is presented. Extensive intraabdominal hematoma and appendicitis were treated surgically and complete recovery was achieved.

Key words: Hepatic aneurysm rupture, necrotizing appendicitis, polyarteritis nodosa.

Poliarteritis nodosa küçük ve orta büyüklükteki arterleri tutan sistemik bir vaskülitir. Klinik seyri tutulan organa göre değişiklik gösterir. PAN'a bağlı olarak gastrointestinal traktus ve karaciğerdeki arterlerde anevrizmal dilatasyonlar ve trombus oluşumu daha önce bildirilmiştir. Ancak hepatic anevrizma rüptürü ve akut apandisit ise çok nadir görülür. Ayrıca her ikisinin birlikte görüldüğü bir olgu da henüz yayınlanmamıştır. Bu yazıda akut hepatic anevrizma rüptürü ile eş zamanlı akut gangrenöz apandisit ile ortaya çıkan bir PAN olgusu sunulmaktadır. Hasta cerrahi ve tıbbi tedavi yöntemleriyle sağlığına kavuşmuştur.

Anahtar kelimeler: Hepatic anevrizma rüptürü, nekrotizan apandisit, poliarteritis nodosa.

INTRODUCTION

Polyarteritis nodosa (PAN) is a systemic vasculitis that is characterized by necrotizing inflammation in small and-medium sized arteries. It was first described by Kussmaul and Maier (1). The incidence is unknown, but this uncommon disease affects people in their fourth and fifth decades of life, with the male to female ratio being 2/3:1(2). Multiple organs, especially the kidney, skin, peripheric nerves, striated muscle and intestine can be affected (1). Gastrointestinal organs such as the liver, appendix and gallbladder are rarely involved (3,4).

Although PAN may affect any organ with vascular and thromboembolic manifestations and consequent tissue ischemia, clinical progress and prognosis depends upon the involved organ and the severity of the disease (5). Common clinical findings in these cases are fever, weakness, fatigue, abdominal pain, peripheral neuropathy, asym-

metric polyarthritis and various cutaneous lesions (1). Patients with renal involvement may have perirenal and retroperitoneal hematoma and secondary changes include aneurysm formation, hemorrhage and thrombosis (3,6,7). The aneurysm may undergo necrosis and heal with fibrosis, but occasionally it will rupture and cause hemorrhage. We report an exceptional case who presented with hepatic aneurysm rupture and necrotizing appendicitis as the first manifestation of underlying PAN. The patient also developed ischemic lesions in the left kidney and head of pancreas.

CASE REPORT

A 58 year-old male was admitted to our hospital with fever, fatigue, myalgia, abdominal pain and diarrhea. He was initially diagnosed with gastroenteritis and treated symptomatically.

However, within a week he had lost four kg in weight and began to experience increasing abdominal pain radiating to the epigastrium and right hypocondrium, with his general condition deteriorating. Because of the severe abdominal pain, he was diagnosed as acute abdomen and transferred to our Surgical Department.

When he was first examined, the patient had a fever (38.5°C, axilla), with a blood pressure of 160/100 mmHg and pulse of 90/min. Physical examination revealed mild hepatomegaly, while the abdomen was markedly and diffusely painful on palpation. Laboratory analyses showed an erythrocyte sedimentation rate (ESR) of 68 mm/h, leukocytosis (white-cell count, 23200/mm³), hypoalbuminemia (albumin, 2.7g/dl) and increased serum levels of transaminases (ALT, 412 IU/L, AST, 552 IU/L). Urinalysis and creatinine levels were normal. While Anti-HIV, anti-HAV, anti-HBV, and anti-HCV antibodies were negative.

Abdominal ultrasonography demonstrated increased liver echogenity in segment three, six, and seven. Non-enhanced abdominal computed tomography showed a hyperdense area, two cm in diameter, in the cortex of the left kidney and a hypodense area 1.5 cm in diameter in the head of the pancreas. This appearance remained after IV administration of radio-contrast media. Enhanced computed tomography also revealed multiple heterogenous lesions in segment three, six, and seven of the liver. The largest of these lesions was 90mmX92 mm in dimension and located in segment seven (Figure 1). These heterogenous appearances in the liver seemed to be result of subcapsular hematoma. In selective celiac truncus angiography, there were multiple aneurysmal dilatations of the hepatic, splenic and renal arteries. Defibrinated blood was aspirated by paracentesis.

With these findings, a diagnosis of hepatic subcapsular hematoma was made and urgent laparotomy performed. There was a diffuse hematoma in the abdomen and the appendix was gangrenously inflamed at laparotomy. The liver appeared ischemic and edematous, with ruptured subcapsular hematoma detected in segment five, six and seven. The hematoma was drained, appendectomy performed and biopsies were taken from the liver.

On histopathological examination of the liver biopsies, there was coagulation necrosis in large

areas, mixed inflammatory cell infiltration in necrotic tissues and portal areas and bleeding focuses in the parenchyma close to portal areas. The epithelial cells had spilled into the lumen of the bile ducts and in one area there was fibrinoid necrosis with mixed cell infiltration in a middle-sized arterial wall, which is usually seen in polyarteritis nodosa.

The appendectomy specimen showed transmural necrosis on histopathological evaluation. In perivascular areas and the submucosal small arterial and venous walls, mixed inflammatory cell infiltration was seen. In the mesentery of the appendix, the middle-sized arterial wall had fibrinoid necrosis and mixed inflammatory cell infiltration related to polyarteritis nodosa was evident.

In the postoperative period the patient's general condition improved significantly and liver enzymes decreased to within normal ranges within a week. The patient was transferred to a rheumatology department for treatment and follow up of polyarteritis nodosa on the 10th postoperative day.

DISCUSSION

Polyarteritis nodosa is a rarely seen systemic vasculitis involving small to medium-sized arteries, characterized by focal, panmural, necrotizing and inflammatory lesions with a tendency to vessel bifurcation (1). The clinical course may be limited or serious, depending on the affected organ. Although PAN may occur with visceral infarction, as in our case, extraordinary aspects such as

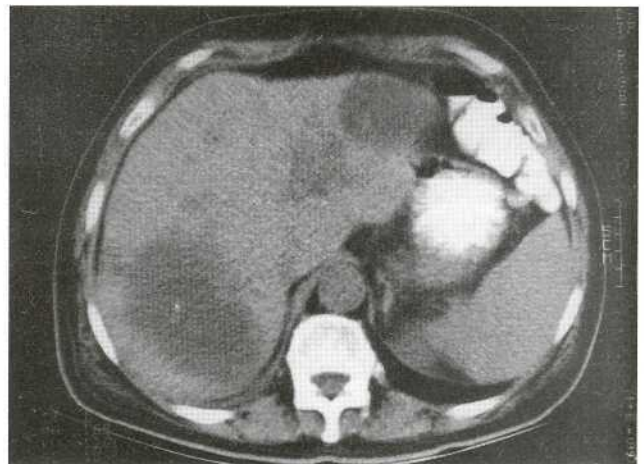


Figure 1. Enhanced abdominal CT showed multiple heterogenous lesions in segment 3, 6 and 7 of the liver.

dramatic presentation of disease, multiple visceral involvement and unusual localization of infarctions have been reported (1,2). Involvement of hepatic and gastrointestinal tract arteries have also been reported although hepatic involvement and hepatic aneurysm rupture are rarely seen (8-10). Necrotizing vasculitis of appendix vermiformis and acute appendicitis as a clinical manifestation of PAN are also uncommon; the largest series in the literature consists of 12 cases reported by Moyana who demonstrated the close relationship between necrotizing arteritis of appendix vermiformis and polyarteritis nodosa (11). In the acute phase, fibrinoid necrosis and mixed type inflammatory cell infiltration involving the whole wall are observed.

Since our patient had no previous history of polyarteritis nodosa, his abdominal pain was initially thought to be due to a gastroenteritis. In spite of the medical treatment, however, his general condition and liver function tests deteriorated and further radiological investigations, especially CT scan and selective celiac angiography, led to the diagnosis of subcapsular hematoma and suspicion of underlying polyarteritis nodosa, preoperatively. Multiple heterogeneous lesions in segment three, six and seven of the liver in the CT scan indicated subcapsular hematoma, and multiple aneurysmal dilatations of the hepatic, splenic and renal arteries in selective celiac angiography led us to consider the possibility of underlying polyarteritis nodosa, although we did not expect to find necrotizing appendicitis preoperatively. Histopathologic examination of the liver biopsies supported the di-

agnosis of polyarteritis nodosa as did histopathologic examinations of appendix vermiformis, which was not expected.

Since PAN is an uncommon vasculitis and the symptoms are non-specific, the diagnosis is always difficult. The lesions may occur in visceral organs and diagnosis often depends upon microscopic tissue examination (12). In our patient, a provisional diagnosis was initially made by angiography and subsequently confirmed by tissue biopsy. The American College of Rheumatology have established ten criteria for diagnosis of polyarteritis nodosa and stated that a patient with vasculitis may only be diagnosed with polyarteritis nodosa if at least three of these 10 criteria are present (13). In our patient, the presence of five of these criteria (diastolic pressure >90mm Hg, myalgias, muscular weakness, weight loss >4 kg, angiographic abnormalities and PMN in a medium sized artery supported the diagnosis of polyarteritis nodosa.

Necrotizing vasculitis of appendix vermiformis and acute appendicitis as a clinical manifestation of PAN is uncommon. Hepatic aneurysm rupture is also rarely seen (14). Indeed, more than one complication in a patient is fairly rare. Intrahepatic bleeding and hemobilia with complicated peritonitis have been reported in only one case with perirenal and hepatic hematoma in only one other (10,14). In our case hepatic aneurysmal rupture and necrotizing appendicitis were seen simultaneously. Although PAN may occur with visceral infarction, the involvement of liver and appendix vermiformis in the same case is very unusual and thus its rarity is worthy of report.

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