

A rare case of splenic infarct presenting with acute Abdominal pain due to polyarteritis nodosa: case report and review of the literature

Poliarteritis nodozaya baęlı akut karın aęrısı ile bařvuran nadir grlen bir splenik infarkt olgusu: olgu sunumu ve literatrn gzden geirilmesi

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Kırk drt yařında erkek hasta acil servise birkaç saat devam eden řiddetli sol st kadran aęrısı ile bařvurdu. Bilgisayarlı tomografi (BT) ve dijital substraksiyon anjiyografide (DSA) dalakta infarkts tespit edildi. Piyesin patolojik incelemesinde diffz akut vasklit, tromboz, panvasklit ile karakterize poliarteritis nodoza tanısı konuldu. Bu tanıyı koymak oęunlukla klinik bulguların deęiřkenlięi nedeniyle zordur. Arteriyel tutulumun yerine ve yaygınlıęına baęlı deęiřiklik gstermektedir.

Anahtar szckler : Dalak infarkts, poliarteritis nodoza, vasklit, acil cerrahi

A 44 year-old man presented to the Emergency Department of American Hospital with severe pain persisting in the left upper quadrant for several hours. A computed tomographic scanning (CT scan) and celiac digital subtraction angiography (DSA) of the abdomen demonstrated a splenic infarct. Histopathological examinations showed diffuse acute vasculitis, thrombosis, panvasculitis which led us to diagnose the case as "polyarteritis nodosa". The diagnosis is usually difficult to establish and it is usually delayed due to variable clinical manifestations dependent on the site and the extent of arterial involvement.

Key words: Splenic infarct, polyarteritis nodosa, vasculitis, emergency surgery

Splenic infarction is a rare event encountered in the emergency departments. In younger patients, splenic infarctions are most commonly associated with sickle cell disease. Other potential causes are valvular heart disease, polycythemia vera, valve replacement, and endocarditis.^[1] A rare multi-system disease with an incidence of about 5/million

per year, polyarteritis nodosa (PAN) occasionally may cause splenic vasculitis, usually at arteriolar bifurcations, leading to multiple systemic infarcts with extensive confluent systemic necrosis.^[2]

We herein present a patient with splenic infarct related to PAN, which may indeed present in various forms depending on the organ involved.

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CASE REPORT

A forty four year- old man applied to the emergency service with severe abdominal pain and nausea of about 24 hours duration. Initial physical examination revealed increased tenderness on the epigastric region and on the left upper quadrant without rebound or defence. On the auscultation of the abdomen, mildly increased intestinal sounds could be heard. White cell count was $11000/m^3$ and laboratory test results were normal. Gastroscopy was performed because of long standing complaints of chronic gastritis which showed findings of "active gastritis". Despite medical treatment with proton pump inhibitors (omeprazole 30 mg IV 1x1), complaints of the patient aggravated. Total abdominal CT with contrast was planned which showed a hypodense area 5 x 7 cm in diameter in spleen. This hypodense area suggested infarction (Figure 1). No other intraabdominal pathology was defined. These signs indicated celiac DSA which manifested a non-perfused avascular triangular zone in the middle part of spleen suggestive of an infarcted area (Figure 2). Treatment with enoxaparine sodium 0.4 mL (1x1), levofloxacin 500 mg (1x1tb) was initiated. Subsequently, because of increased tenderness, rebound and defence in the epigastric area and left upper quadrant and leucocytosis (11000 to $17000/mm^3$), the patient underwent splenectomy with the diagnosis of acute abdomen (infarcted spleen). Histopathological examination of the spleen showed



Figure 1: In CT with contrast, hypodense area manifesting a splenic infarct



Figure 2: DSA showing avascular triangular non-perfused zone suggestive of an infarct in the middle part of spleen.

diffuse acute vasculitis, thrombosis, massive fresh bleeding and necrobiosis, hyaline arteriosclerosis, panvasculitis with fibrinoid necrosis at the minimal level in the trabecular arteries which led us to the diagnosis of "polyarteritis nodosa". Consultation from the Department of Rheumatology directed the treatment with pulsatile prednisolone 30 mg IV which was later replaced with oral prednisolone (30 mg/day). At the fifth postoperative day, levels of liver enzymes increased; the blood biochemistry was as follows: AST:45 IU/L, ALT:94 IU/L, GGT:98 IU/L, Albumin:3.1g/dL, Glucose:85 mg/dL, Ca:8.5 mg/dL, Na:140 mmol/L, K:4.3 mmol/L, Cl:106 mmol/L, lupus anti-coagulant (+), HbsAg (-), Anti-HBs total (-), CRP:91.4 U/L, ESR: 67 mm (1hour). Celiac DSA was repeated in order to rule out a hepatic infarction. However the branches of hepatic and renal arteries were defined as normal according to the celiac DSA. Parenteral cyclophosphamide 150 mg IV was initiated as a result of Rheumatology and Hepatology consultations. The general condition of the patient improved and he was discharged from the hospital with oral prednisolone (30 mg daily). Thirty-one days after the operation, the patient applied to our department again because of distension and tenderness in the abdomen. Abdominal CT with contrast media showed diffuse hemorrhage at the splenectomy site. Celiac DSA was repeated which showed contrast extravasation that was in harmony with ongoing bleeding at the distal end of pancreatic ar-

tery (Figure 3). Bleeding could be stopped by angiographic embolisation. However, tenderness on the right upper quadrant of abdomen and WBC ($17000/\text{mm}^3$), AST (221 IU/L), ALT (1956 IU/L) increased subsequently. Repeated abdominal CT with contrast media showed a non-perfused, hypodense area which was in harmony with infarction in the left lobe of liver. Due to his well general health, a conservative approach was decided. The patient responded well to treatment with oral 150 mg cyclophosphamide and incremental oral steroid doses up to 80 mg daily. It was decided to follow the patient on an out-patient basis when the complaints regressed and no other complication of infarction was observed.

DISCUSSION

Splenic infarction can occur in a variety of clinical situations. Patients under 40 years of age typically have an associated hematological disorder such as sickle cell disease. Patients over 40 years of age typically have experienced an embolic event.^[3] While it is well known that sickle cell disease can precipitate splenic infarction, patients with sickle cell trait, as well as hemoglobin SC disease^[4] may develop splenic infarction^[5]. Other unique causes of splenic infarction include polycythemia vera,^[6] Gaucher's disease,^[7] pancreatitis,^[8] endocarditis,^[9] malignancy,^[10] as well as

collagen-vascular diseases.

Polyarteritis, also termed "periarteritis nodosa" and "polyarteritis nodosa" is a multi-system disease characterised by acute inflammation and fibrinoid necrosis of medium sized and small arteries.^[11,12] Histologically, all layers of the vessel wall are involved in varying stages of inflammation, followed by chronic mononuclear cell infiltration and fibrosis.^[11,12,13] Multiple saccular or fusiform aneurysms up to one cm may occur in the renal, hepatic, and visceral vasculature in 60-80% of cases.^[14] Infarction due to arterial occlusion and hemorrhage because of aneurysmal rupture are the two most important features of this disease. Early mortality is high because of the acute vascular compromise of such critical organs as the kidney, heart, and central nervous system. PAN is slightly more common in males and usually occurs in the third to sixth decade, though it may affect patients from infancy to advanced age.^[2]

Clinical symptomatology of splenic infarct consists primarily of left upper quadrant pain. Pain associated with splenic infarct may have a pleuritic component, and may radiate to the left shoulder. In half of patients, anemia and a leucocytosis may develop.^[3]

There are several diagnostic modalities that may be utilised for the establishment of definitive diagnosis of splenic infarction. A KUB (kidney, ureter, and bladder) is never diagnostic, but certain findings aids in diagnosis. Left upper quadrant gas or air-fluid levels are found only with massive infarction.^[15,16]

Ultrasound has been used to image splenic infarction^[17,18]; however an interval of 24 hours or more after the infarction is required for the identification of a demarcation line between normal and infarcted tissue.^[18]

In approximately one third of the cases, CT scan will show classic wedge-shaped low-attenuation area of infarction with rind enhancement. Depending on the amount of ischemia or hemorrhage in this area, infarcts may be seen as areas of low or mixed attenuation.^[19]

Arteriography may assist in diagnosis, and may provide information on prognosis by showing the extent of the disease. Angiographic findings include arterial saccular or fusiform aneurysms, and narrowing or occlusion of the arteries.^[20] They oc-



Figure 3: In DSA contrast extravasation demonstrating continuous bleeding from the distal end of pancreatic artery.

cur commonly in the kidney, in the liver, and in mesenteric, cerebral, splenic, pulmonary, lumbar, intercostal, inferior phrenic, hypogastric, and gastro-duodenal arteries.^[10] The aneurysms may disappear with improvement of the vasculitis.^[21] However, the angiographic findings mentioned above may also be associated with other diseases, such as Wegener's granulomatosis, CSS, systemic lupus erythematosus, thrombotic thrombocytopenic purpura, infective endocarditis, atrial myxoma, angitis associated with drug abuse, and metastatic malignancy involving peritoneum.^[22,23]

The most important diagnostic test is biopsy of the involved organ to demonstrate the characteristic histologic changes of PAN. The most accessible affected areas are skin, sural nerve, muscle, kidney, and testicles.^[10]

High-dose corticosteroids (eg, prednisone 60 mg/day in divided doses) may prevent progression and appear to induce partial or near-complete remission in about 30% of the patients. Immunosuppressive drugs, either alone or initially with corticosteroids are used with some success when corticosteroids alone are inadequate. Cyclophosphamide (2 to 3 mg/kg/day PO) may be given to patients who do not respond to corticosteroids during the first few weeks of therapy or for whom prohibitively high doses of corticosteroids appear necessary to control disease (by these criteria, most patients qualify for steroid therapy).^[24]

Diagnosis of PAN is difficult to make and it is usually delayed due to diversity of symptoms. Clinically, PAN is mostly confused with infectious diseases, conditions causing acute abdomen, bleeding peptic ulcer, glomerulonephritis, coronary artery disease, myositis, and polyneuritis.

Splenectomy is indicated for persistent symptoms or for complications of the infarct such as hemorrhage, abscess, or splenic pseudocyst. Nonoperative management of a complication has been associated with a significant mortality rate. Specific treatment aimed at correcting the identified underlying cause should be undertaken. Older patients with suspected thromboembolic etiologies should be considered for anticoagulation or vena cava filter placement.

The diagnosis of splenic infarction can best be made by use of CT, liver-spleen scans, or angiography. Most patients recover without the need for

splenectomy, however underlying causes must be treated in order to prevent recurrences of PAN.

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