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A RARE CASE: SPONTANEOUS SPLENIC RUPTURE BECAUSE OF SPLENIC ANGIOSARCOMA

Nadir Görülen Bir Olgu: Spontan Dalak Rüptürü

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ABSTRACT

Primary splenic angiosarcoma and associated spontaneous splenic rupture are very rare condition. In this article, we discussed a patient who had the complaint of sudden onset abdominal pain, had shock findings in the first examination and who was diagnosed with splenic rupture in the later evaluation. In our case, the patient was diagnosed with angiosarcoma by post-operative pathology material results. Since mortality rate is very high in angiosarcoma patients who develop splenic rupture, splenectomy should be performed in the early period, if patients are considering the diagnosis of splenic angiosarcoma.

Keywords: *Emergency department, spontaneous splenic rupture, angiosarcoma*

ÖZ

Primer dalak anjiosarkomu ve buna bağlı olarak görülen spontan dalak rüptürü çok nadir görülen bir durumdur. Bu yazıda ani başlangıçlı karın ağrısı ile başvuran ve geliş muayenesinde şok bulguları olan, sonrasında yapılan değerlendirmede dalak rüptürü tespit edilen olgu tartışıldı. Olgumuzda anjiosarkom tanısı postop patoloji materyal sonucu ile konuldu. Dalak rüptürü gelişen anjiosarkom olgularında mortalite çok yüksek seyrettiğinden, hastalarda dalak anjiosarkom tanısı düşünüldüğü takdirde erken dönemde splenektomi uygulanmalıdır.

Anahtar kelimeler: Acil servis, spontan dalak rüptürü, anjiosarkom

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INTRODUCTION

Splenic rupture is usually a life-threatening condition observed following blunt abdominal trauma but non-traumatic splenic rupture is a condition that is observed rarely. This usually occurs in a pathological spleen and may rarely be observed in a healthy spleen (1). Splenic angiosarcoma is one of these reasons. Spleen angiosarcomas constitutes less than 1% of all angiosarcomas. Most cases present with complaints such as abdominal pain, fatigue, weight loss and back pain, and a small number of patients present with intra-abdominal bleeding symptoms related to spontaneous rupture (2). Patients aged 65 years and over constitute approximately 9-10% of patients admitted to the emergency department due to complaint of abdominal pain and more than half of these patients are hospitalized. More than one third of hospitalized patients need surgery. In patients who are above 65 years and who present with the complaint of abdominal pain, mortality rate was found to be 11-14% (3). In this article, we aimed to describe a case of spontaneous splenic rupture presented in our emergency department due to abdominal pain. In this case splenic angiosarcoma induced intra-abdominal bleeding and hypovolemic shock.

CASE

Eighty six years old female patient presented to our emergency department with her relatives due to complaint of abdominal pain that started after a meal about an hour ago. The patient described the pain in the epigastric region as a constant blunt pain that does not radiate. Her medical history does not include any known characteristic other than hypertension and dementia. Her family history is not fully known.

In the physical examination, general condition is moderate, consciousness is prone to sleeping, partial orientation and cooperation. Respiratory sounds were normal, no rales, no ronchi. Heart sounds S1 and S2 were rhythmic. No additional sounds or murmurs. In the abdominal examination, tenderness was noted in the epigastric region, abdominal defence was present and no rebound was detected. Blood pressure was found as 100/60 mmHg, respiratory rate as 16/minute, oxygen saturation as 96% and pulse as 110/minutes. We asked the patient for complete blood count (CBC), biochemistry, cardiac enzymes and blood gas analyse. In the complete blood count of the patient: Hemoglobin (Hgb) was found as 11.6 g/dl, hematocrit (Hct) as 35.5%, platelets (Plt) as 248000, white blood cell (Wbc) as 14700, and cardiac enzyme values and biochemistry values were found to be in normal range. In electrocardiography (ECG), sinus rhythm was detected and no ST-T change was noted.

Contrast-enhanced abdominal computed tomography (CT) was requested because the patient's physical examination findings were compatible with acute abdomen. In the contrast-enhanced abdominal CT as shown in figure 1, common contrasting defects were observed in the splenic parenchyma and free fluid appearance was observed starting around the spleen and spreading in the abdomen with a depth of about 6 cm in the pelvis. In addition, a large number of cystic lesions were present in the liver parenchyma.

As a result of the contrast-enhanced abdominal CT, after determining compatibility with the diagnosis of splenic rupture, control CBC was sent. In the control CBC, Hgb was found as 10.2 g/dl, Hct as 30.8%, Plt as 241000 and Wbc as 17520.

The patient was referred to the general surgery clinic due to diagnosis of spontaneous splenic rupture with her current findings. She was admitted to the department of general surgery for operation. The material sent to the pathology after splenectomy was detected as angiosarcoma according to the result of the histopathological examination. The patient died on the 30th post-operative day due to multiple organ failure.

DISCUSSION

Spontaneous splenic rupture is a condition that is observed rarely. This usually occurs in a pathological spleen and may rarely be observed in a healthy spleen (1). Spontaneous rupture is observed in 13-39% of angiosarcoma cases, as in our case (4). Although the rate of spontaneous rupture is high this is very rare condition because primary angiosarcoma of the spleen is observed in 0.15-0.26 cases per million (5). In addi-

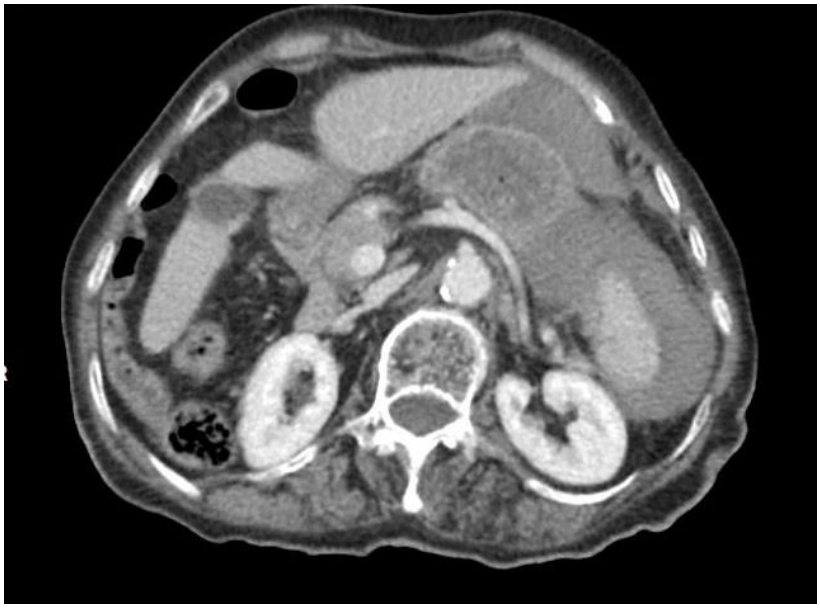

Figure 1: Contrast-enhanced abdominal CT; image of splenic rupture, diffuse free fluid around spleen

tion it is seen in various ages, it is more common in the 5th-6th decades and occurs equally in males and females (4).

Trauma may have a role in the etiology of spontaneous splenic rupture in the angiosarcoma such as turning in bed, coughing or defecation but this cannot be clearly determined (6). We were unable to clearly determine whether there was a minor trauma as mentioned above, although there was no history explaining a major injury in our case.

Splenic angiosarcoma patients may usually present with symptoms, such as abdominal pain, weakness, fatigue, loss of appetite and weight loss due to splenomegaly (7). In our case, patient presented to the emergency department with the complaint of sudden onset of abdominal pain after a meal, and it is not compatible with the cases in the literature. Pathological rupture of the spleen with malignant involvement is an event observed very rarely. Pathological rupture male/female ratio is roughly 3:1. This rate varies greatly in some diseases. For example, this rate can reach 3:1 in acute myeloblastic leukemia (AML), 8:1 in acute lymphoblastic leukemia (ALL), 7:1 in chronic lymphocytic leukemia (CLL) and 19 in chronic myelocytic leukemia. On the other hand, the male/female rate splenic rupture in non-Hodgkin's lymphoma (NHL) and hairy cell leukemia (HCL) was determined as 1:1 (8). However in a clinical study, no correlation was observed between rupture and age, gender, spleen size and degree of anemia (8).

Mortality rate is high in the case of rupture. Splenic angiosarcoma is a very aggressive tumor with poor prognosis and a six-month survival rate of 20% (10). At the time of diagnosis most cases are metastatic. Metastases occur mainly in liver and in lung, lymph nodes and bone (11). At the time of presentation to the emergency department, she was not diagnosed with angiosarcoma and she was diagnosed with post-operative pathology material. In the contrast-enhanced abdominal CT examination of the patient in the emergency department, no evidence supporting metastasis was detected in the lungs, bone tissues and other tissues.

The treatment angiosarcoma consists of splenectomy followed by chemotherapy combined with radiotherapy. In patients who develop spontaneous splenic rupture, prognosis is worse; therefore, performing splenectomy before developing of rupture can increase the rate of survival (10). Our patient was not diagnosed with angiosarcoma before presenting to the emergency department. As in our case, this is not always possible and patients can, although rarely, present to the hospital with splenic rupture.

CONCLUSION

In conclusion, although splenic angiosarcoma is a very rare disease, idiopathic anemia should be considered in the presence of abdominal pain and splenomegaly. Moreover, sponta-

neous splenic rupture should also be considered in the differential diagnosis of patients with hypovolemic shock findings accompanying acute abdomen findings such as hypotension tachycardia.

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