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Locally Invasive Primary Splenic Angiosarcoma

Lokal İnvaziv Primer Splenik Anjiosarkom

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Abstract

Primary angiosarcoma of the spleen is a very rare vascular neoplasm, but it represents the most common non-hematolymphoid malignant tumor of the spleen. In this report, we present the case of a 48-year-old man with primary splenic angiosarcoma with local invasion to the left diaphragm and the radiological imaging findings for this cancer.

Key Words: Computed tomography, Neoplasm, Splenic angiosarcoma

Özet

Primer dalak anjiosarkomu oldukça nadir görülen bir malinite olup dalağın en sık görülen hematolenfojen olmayan tümörüdür.Bu olgu sunumunda sol diyafragmaya lokal invazyon yapmış primer dalak anjiosarkomu olan 48 yaşındaki erkek hastanın görüntüleme bulguları tartışılacaktır.

Anahtar Kelimeler: Bilgisayarlı tomografi, Dalak anjiosarkomu, Tümör

Introduction

Primary angiosarcoma of the spleen is a rare malignant neoplasm, but it is the most common primary neoplasm of the spleen arising from the vascular endothelium. The condition was first described by Langhans in 1879 [1]. As a very aggressive neoplasm, it has a very poor prognosis and has a high metastatic potential, and thus can spontaneously rupture. Therefore, patients with splenic angiosarcoma usually present with widespread metastasis disease or splenic rupture [2, 3]. Herein, we present the case of a 48-year-old man with primary splenic angiosarcoma with local invasion to the left diaphragm.

Case Report

A 48-year-old man was admitted with left upper quadrant pain and early satiation. There was no history of other systemic illness, fever or trauma. The laboratory tests revealed no anemia or thrombocytopenia, but an increased level of CA125 was found.

On ultrasound examination, splenomegaly with lobulated margins and heterogeneous echogenicity was seen in the parenchyma resulting from a huge mass. There were no associated enlarged lymph nodes in the abdomen. The liver and other abdominal organs were normal. On CT scan, the findings were similar to those of the ultrasound. In pre-contrast CT images, splenomegaly with lobulated margins and heterogeneous parenchyma were seen. There was no calcified focus. Post-contrast CT images showed multiple ill-defined confluent hypodense masses, which almost entirely replaced the spleen (Figure 1). On MR images, a huge mass consisting of multiple ill-defined variable-sized lesions was seen. The mass was heterogeneous; it was hypointense and hyperintense on T1- and T2-weighted images, respectively. The mass generally showed moderate contrast enhancement, less than normal splenic parenchyma. The upper portion of the mass revealed central mild hyperintensity on T1-weighted pre-contrast images and hyperintensity on T2-weighted images. Post-contrast T1-weighted images showed hypointensity in this region, suggesting hemorrhagic or non-hemorrhagic central necrosis (Figure 2). Because of the high risk of rupture and hemorrhage into the peritoneal cavity, fine needle aspiration or Tru-cut biopsy was not performed.

A laparotomic splenectomy was planned. During the surgery, the upper portion of the splenic mass was found to have invaded the left diaphragm. After total resection of the mass, an 8-cm defect in the left diaphragm appeared, and primary repair was performed after the consultation with thoracic surgeons. Pathologic examination revealed primary angiosarcoma of the spleen. Three months after the surgery, multiple pleural metastatic lesions were found on a PET-CT scan (Figure 3).

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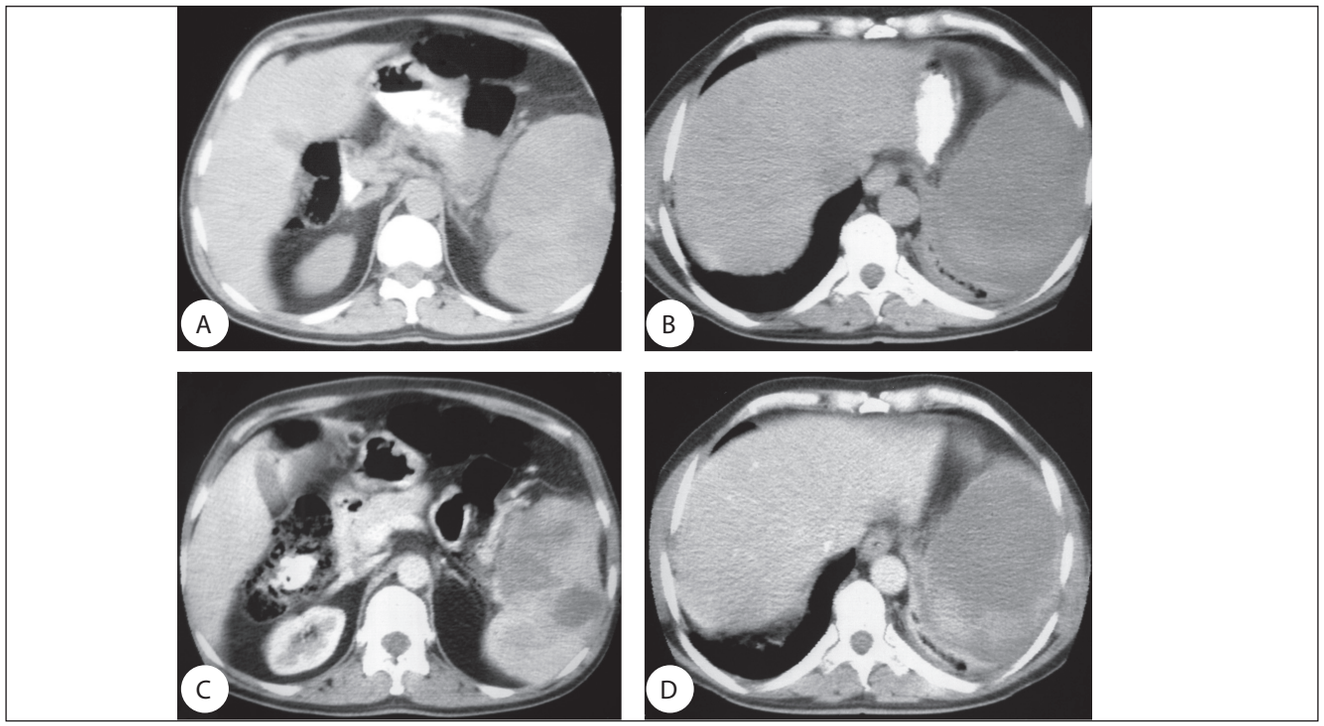

Figure 1. *Transverse CT scans.* ***(A, B)*** *Splenomegaly with lobulated margins and heterogeneous paranchyma is seen on pre-contrast images.* ***(C, D)*** *Multiple, variable-sized, ill-defined hypodense lesions that are less contrast enhanced than splenic paranchyma on post-contrast CT images.*

Discussion

Primary angiosarcoma of the spleen is a very rare vascular neoplasm, but it represents the most common non-hematolymphoid malignant tumor of the spleen. No more than 200 cases have been reported worldwide. The mean age of presentation is 59 years, with eight cases reported in the pediatric population [4, 5].

Unlike angiosarcomas of the liver; splenic angiosarcomas have no documented association with exposure to carcinogens such as thorium dioxide, vinyl chloride or arsenic. However, there have been case reports of splenic angiosarcoma associated with previous chemotherapy for testicular seminoma. The most common clinical presentation of splenic angiosarcomas is left upper quadrant pain; the others are fatigue, weight loss and fever [6]. On physical examination, splenomegaly is the most common finding. Splenic angiosarcomas frequently rupture spontaneously, which is the major complication and presents with signs of hemoperitoneum [7].

The laboratory findings associated with primary angiosarcoma of the spleen are anemia, thrombocytopenia, cytopenia, leukocytosis, thrombocytosis and elevated erythrocyte sedimentation. However, in our case, an elevated serum level of CA125 was the only abnormal laboratory finding. In the literature, there is only one case of cardiac angiosarcoma with an elevated CA125 level; the levels of CA125 are often elevated in patients with ovarian neoplasms and non-lung tumor metastases [8].

Primary angiosarcoma of the spleen is very aggressive and has a high potential of early metastasis that often involves the liver (89%), lungs (78%), lymph nodes (56%), bone (44%), brain, adrenal gland, omentum, bone marrow, gastrointestinal tract, and peritoneum. After therapy, metastasis can occur. In our case, there were pleural and costal metastases during the early post-operative period. This finding is unusual and stands in contrast to the more frequently reported metastatic sites in the literature. Moreover, the diaphragmatic metastasis seen in the current case has not been reported previously.

The main radiological finding of splenic angiosarcomas is splenomegaly. On ultrasound, heterogeneous echotexture of the mass with cystic areas as a result of necrosis and hemorrhage is frequently seen. Solid portions of the mass can demonstrate increased Doppler flow owing to hypervascularity of the tumor. On pre-contrast CT images, an ill-defined splenic mass is the most common appearance, and hyperdense areas can be seen as a result of hemorrhage within the tumor or of spontaneous rupture into the peritoneal cavity. On pre-contrast images, punctate calcifications and massive calcifications in a radial pattern have also been reported in patients with splenic angiosarcomas [9]. On post-contrast images,

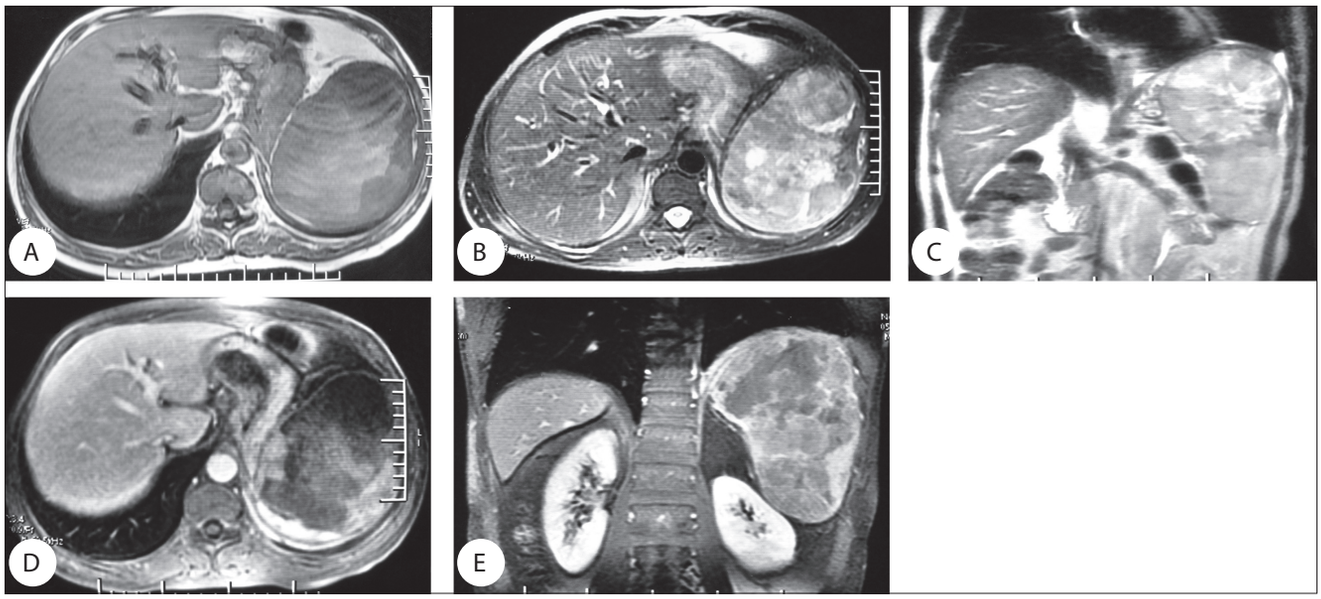

Figure 2. *MR Images. In comparison with the liver paranchyma, the splenic paranchyma is seen as heterogeneous and hypointense on T1-weighted images* ***(A)*** *and as hyperintense on T2-weighted images* ***(B, C)*** *After contrast administration, variable-sized, ill-defined, heterogeneously enhanced lesions are seen* ***(D, E)****. On T1- or T2-weighted images, the intensity of the solid masses changes from low to high signal because of the presence of hemorrhage or necrosis.*

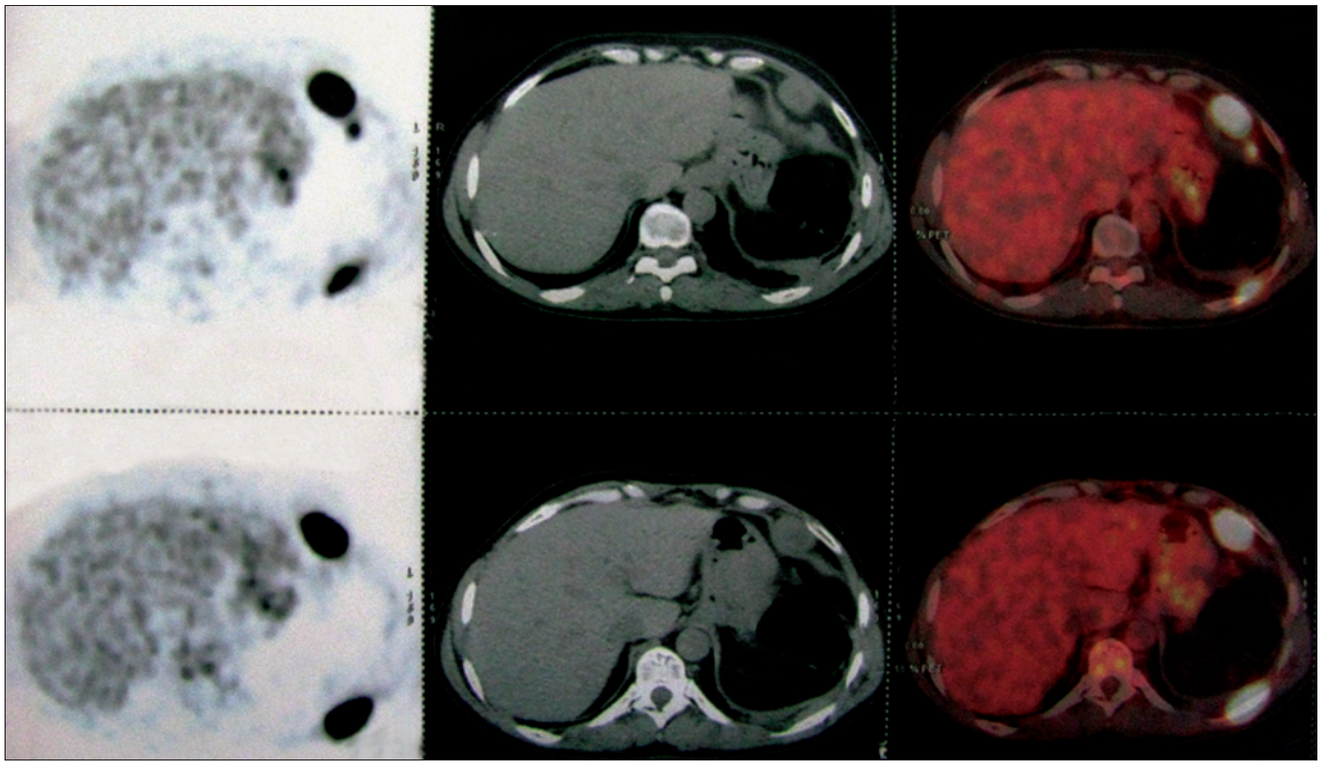

Figure 3. *PET scans. Axial plane images of the upper abdomen; a 3.5 cm soft tissue mass with smooth contours that infiltrates the left anterobasal costo-diafragmatic pleura is seen. The mass is hypermetabolic, and SUV value is 29. Additionally, in the left 10th costa, a mass that is considered to be metastasis with similar features is seen.*

heterogeneous enhancement and less enhanced areas due to necrotic degeneration can be seen. MR imaging of splenic angiosarcomas is important because MR imaging reflects the presence and nature of the hemorrhage within the tumor. A low signal indicates the presence of hemosiderin and siderotic nodules. In both T1- and T2-weighted images, the signal intensity of the mass changes from low to high based on the presence of the necrosis and blood products. Contrast-enhanced MR images show heterogeneous enhancement due to the presence of solid and necrotic areas.

Differential diagnoses of splenic angiosarcomas include lymphomas, metastases, hemangiomas, hamartomas, littoral cell angiomas, lymphangiomas, hemangiopericytomas, and hemangioendotheliomas [10].

Biopsy is avoided in cases of splenic angiosarcoma because of the high risk of rupture. Therefore, histological studies are usually done after splenectomy. For treatment, splenectomy is typically performed; chemotherapy has not been shown to be effective.

In conclusion, primary splenic angiosarcoma is a rare neoplasm with a high metastatic potential. There are many lesions included in the differential diagnosis, and many radiological methods must be used to examine patients with suspected primary splenic angiosarcoma. MR imaging provides some clues for the diagnosis, such as presence of blood product signals within the tumor, irregularity of the tumor margins and indistinctness between the tumor and adjacent organs. Additionally, increased FDG uptake visualized by PET-CT imaging can support the diagnosis preoperatively.

Conflict of interest statement: The authors declare that they have no conflict of interest to the publication of this article.

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