



## CASE REPORT

### DIAGNOSIS, TREATMENT, RADIOLOGIC AND PATHOLOGIC FINDINGS OF SPLENIC ANGIOSARCOMA: A CASE REPORT

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#### ABSTRACT

Although primary splenic angiosarcomas are the most common malignant nonhematopoietic tumors of the spleen, they are extremely rare tumors with very poor prognosis. These tumors are highly aggressive and lethal, present with widespread metastasis or splenic rupture. Definitive diagnosis and treatment is surgical and survival is significantly improved when splenectomy is performed prior to rupture. Diagnosis, radiologic and pathologic findings and treatment of a patient with primary splenic angiosarcoma are presented.

**Keywords:** Angiosarcoma, Spleen, Immunohistochemistry, Imaging

### SPLENİK ANJİOSARKOMANIN TANISI, TEDAVİSİ, RADYOLOJİK VE PATOLOJİK BULGULARI: OLGU SUNUMU

#### ÖZET

Primer splenik anjiosarkomalar dalağın hematopoetik sistemle ilişkili olmayan en sık malign tümörleri olmasına karşın oldukça nadir görülmekte ve prognozu son derece kötüdür. Bu tümörlerin seyri oldukça agresif ve ölümcül olup yaygın metastazlar veya dalak rüptürü ile karşımıza çıkarlar. Kesin tanı ve tedavi cerrahi ile olur ve rüptür öncesi uygulanan splenektomi yaşam süresini anlamlı şekilde uzatır. Primer splenik anjiosarkomalı bir hastanın tanı, radyolojik patolojik bulgular ve tedavisini sunuyoruz.

**Anahtar Kelimeler:** Anjiosarkoma, Dalak, İmmünohistokimya, Görüntüleme

#### INTRODUCTION

Primary malignant splenic mesenchymal tumors are extremely rare<sup>1</sup>. These tumors, the most common being angiosarcoma, are highly aggressive<sup>2</sup>. Patients present with widespread metastasis or splenic rupture, the latter being lethal<sup>2</sup>. Significant improvement of survival with splenectomy prior to rupture makes early diagnosis a necessity. Symptoms, signs, radiologic and pathologic findings of primary splenic angiosarcoma are presented.

#### CASE REPORT

A 50 year-old male patient was admitted with left upper quadrant pain, fatigue and weight loss of 7 kg in 2 months. His physical examination revealed a painful, palpable spleen 6 cm below the left costal margin on the mid axillary line. Peripheral blood count revealed a hemoglobin level of 9.8 g/dl, hematocrit level of 30.1%, a platelet count of 129000/mm<sup>3</sup> and a white blood cell count of

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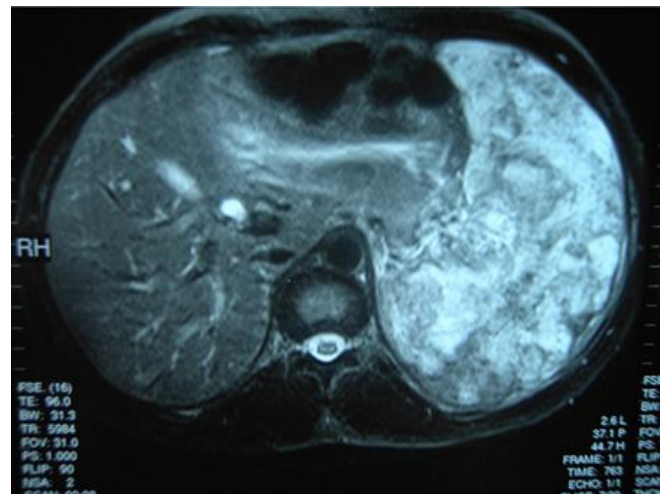


7300/mm<sup>3</sup>. Liver function tests were within normal limits except for mildly elevated LDH levels. Peripheral blood smear revealed schistocytes and thrombocytopenia which were suggestive of microangiopathic hemolytic anemia. Bone marrow aspiration biopsy and core biopsy were normocellular. On ultrasonography(USG), the spleen was extremely enlarged and filled with multiple irregular contoured nodular lesions, the largest being 55x38 mm in size, located on the lower pole of the spleen. These lesions were thought to be metastatic. On the abdominal computerized tomography(CT) the spleen was extremely enlarged, the longest axis being 17 cm and filled with diffuse hypodense lesions, which were thought to be secondary to a lymphoproliferative disease like lymphoma or to a primary splenic malignant tumor(Figure 1). On abdominal magnetic resonance imaging(MRI), the spleen was extremely enlarged, containing multiple hemorrhagic and necrotic lesions which were suggestive of primary splenic angiosarcoma(Figure 2). Multiple bone lesions were observed which were thought to be metastatic. Fine needle aspiration

biopsy(FNAB) revealed a few atypical spindle cells suggestive of splenic angiosarcoma. Splenectomy was performed for definitive diagnosis and treatment. The splenectomy specimen was 22x16x7 cm in size, and had a smooth capsula. On the cut surface, a white tumoral lesion of 10 cm in diameter was observed with uneven sharp borders which completely replaced splenic parenchyme. The rest of the parenchyme was cystic, hemorrhagic and necrotic. Histopathologic examination revealed hemorrhagic infarcts and solid tumoral lesions which completely replaced the splenic parenchyme. Atypical spindle cells and pleomorphic cells were observed in these solid lesions. These cells had a hyperchromatic nucleus and an irregular nuclear membrane. The tumoral lesions consisted of freely anastomosing papillary and classical, characteristic vascular channels. On immunohistochemical examination, these lesions stained positive for factor VIII, CD34, CD31, vimentin, and CD 68, which presented the diagnosis of angiosarcoma. The Ki proliferation index was less than 25%.



**Figure 1:** Abdominal CT: extremely enlarged spleen; the longest axis being 17 cm.



**Figure 2:** Abdominal MRI showing extremely enlarged spleen with multiple hemorrhagic and necrotic nodules:



## DISCUSSION

Primary splenic angiosarcomas are the most common malignant nonhematopoietic tumors of the spleen<sup>2</sup>. They are tumors of vascular origin, characterized by masses of endothelial cells displaying cellular atypia and anaplasia, characteristic of malignancy<sup>3</sup>. The incidence is only 0.14-0.25 cases per million<sup>4</sup>. The age group for this malignancy is 50-59 years, with a slight male predominance<sup>5</sup>. Patients usually present nonspecific symptoms like left upper quadrant pain and fatigue<sup>5</sup>. Physical examination reveals a left upper quadrant mass and tenderness and microangiopathic hemolytic anemia is observed on laboratory examinations<sup>2</sup>. Although presenting symptoms are nonspecific and are often confused with lymphoma, considerations for treatment and prognosis is quite different<sup>6</sup> and surgery provides the definitive diagnosis and treatment<sup>6</sup>. These tumors are highly aggressive and lethal, present with widespread metastasis or splenic rupture<sup>2</sup>. There is no correlation between spontaneous splenic rupture and the age or sex of patients, the size of spleen and degree of anemia<sup>6</sup>. Improvement of survival by splenectomy before development of splenic rupture makes early diagnosis a necessity<sup>7</sup>. USG determines the cystic or solid nature of the tumoral mass and is the first investigation method to diagnose malignancy<sup>8</sup>. CT provides the basic information about this organ<sup>9</sup>. MRI, by virtue of its superb soft tissue contrast and lesion characterization, is used for splenic lesions in which differential diagnosis was not reached by CT<sup>9</sup>. FNAB alone cannot make but supports the diagnosis of angiosarcoma if it is clinically suspected. Definitive diagnosis is made by histopathologic and immunohistochemical examination of the spleen. Microscopical examinations reveal freely anastomosing papillary and classical characteristic vascular channels lined by masses of endothelial cells displaying cellular atypia and anaplasia, characteristic of malignancy<sup>3</sup>. Immunohistochemically, at least 2 vascular proliferation markers (CD34, FVIII<sub>Ag</sub>, VEGF3 and CD31) and at least 1

histiocytic differentiation marker (CD68 and/or lysosyme) makes the diagnosis<sup>10</sup>. Histologic angiosarcoma studies must include mitotic ratio, cellularity, cellular pleomorphysim, dimension of necrosis and vascular differentiation. Only the mitotic ratio, KI 67 proliferation index, determines prognosis. Because of the rarity of this tumor, no specific regimen of chemotherapy has been employed in enough cases to enable the drawing of conclusion to the effect on survival<sup>6</sup>. However, empiric use of multiagent chemotherapy can be offered to patients with metastatic disease<sup>6</sup>.

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