Primary Splenic Hemangioendothelioma: A Case Report and Literature Review

Primer Splenik Hemanjıyoendotelyom: Olgu Sunumu ve Literatürün Gözden Geçirilmesi

Nergis EKMEN, a Aysen HELVACI, b Meral GÜNALDI, c Naciye DEMIREL YILDIRIM, b Hadi SASANI d

aDepartment of Gastroenterology, İstanbul Bilim University Faculty of Medicine, bClinic of Haematology, Okmeydanı Training and Research Hospital, cClinic of Oncology, Bakırköy Dr. Sadi Konuk Training and Research Hospital, İstanbul 
dClinic of Radiology, Eğirdir Bone-Joint Diseases and Rehabilitation Hospital, Isparta

ABSTRACT Splenic tumors are rarely seen tumors. Primary non-lymphoid vascular tumors of spleen are hemangioma, lymphangioma, hamartoma, littoral cell angiomma, hemangioendothelioma, myoid angioendothelioma and angiosarcoma. Primary splenic epithelioid hemangioendothelioma is very rare tumor among this group. Hemangioendothelioma is generally considered to have a potentially intermediate-borderline malignancy. In the literature review, among splenic hemanjıyoendotelyoma cases, rarely of them were reported as primary splenic epithelioid hemangioendothelioma. Its frequency among splenic tumors is 0.3-14%. These tumors do not become in large sizes and have better prognosis. If they become large, they may cause symptoms such as pancytopenia and hemolytic anemia due to splenomegaly and hypersplenism. Early diagnosis and treatment may prevent conditions such as widespread metastasis, splenic rupture and operative complications. In this article, we present clinical and imaging findings of epithelioid splenic hemangioendothelioma with review of other reported cases in the light of literature.

Key Words: Hemangioendothelioma; spleen; splenectomy; tomography, spiral computed; magnetic resonance imaging


Anahtar Kelimeler: Hemanjıyoendotelyom; dalak; splenektomi; tomografi, spiral bilgisayarlı; manyetik rezonans görüntüleme


Splenic tumors are more rarely seen comparing to other visceral organ tumors. Primary tumors of spleen are divided into non-lymphoid and lymphoid groups. Vascular neoplasms of non-lymphoid tumors consist the vast majority part.1

doi: 10.5336/intermed.2015-44600

Copyright © 2016 by Türkiye Klinikleri

Türkiye Klinikleri J Intern Med 2016:1(1)
Hemangioma is the most common seen non-lymphoid vascular tumor of spleen and has benign characteristic. Its frequency among splenic tumors is 0.3-14%. These tumors do not become in large sizes and have better prognosis. If they become large, they may cause symptoms such as pancytopenia and hemolytic anemia due to splenomegaly and hypersplenism. Hemangiosarcoma is very rare non-lymphoid primary malign neoplasm of spleen. Hemangiosarcomas which may be in larger dimensions, are able to have very aggressive and fatal characteristic. Life expectancy of patients with hemangiosarcoma is less than one year. Widespread metastasis and catastrophic outcomes such as splenic rupture is possible. Among primary vascular tumors, epithelioid hemangioendotheliomas (HE) are rare intermediate vascular tumors. These tumors never perform the exact biological behavior. They may be seen just like hemangioma or hemangiosarcoma.

CASE REPORT

A 42-year-old male patient admitted to our clinic with complaint of left-side abdominal pain. His physical examination and all laboratory (biochemical tests, complete blood count) findings were found as normal. In abdominal ultrasonography examination, splenomegaly with multiple lesions (the largest size of 38x36 mm) in hypo-echoic appearance were observed (Figure 1). Blood flow velocities, flow patterns and directions were normal in Doppler ultrasonography. Abdominal computed tomography (CT) revealed multiple hyperdense nodular lesions in the splenic parenchyma (Figure 2). Characteristics of lesions were not compatible with metastasis in abdominal magnetic resonance imaging (MRI) (Figure 3). No imaging finding in the favor of metastasis was detected in thoracoabdominal CT study. Regarding to these findings, splenectomy was performed subsequently. In the pathological examination, many foci of HE were reported (Figure 4). Mild nuclear polymorphism, mitosis 1/50 (High Power Field-HPF), no necrosis, Ki 67 proliferation index less than 5% were observed in pathological examination. CD3, CD4 and factor 8 staining were positive. S-100, CD68, CD8, cytokeratin staining and immune reactivity were negative. After 2-year post-operation follow up, all of physical examination, laboratory and imaging findings were remaining stable.

DISCUSSION

HE is considered as an intermediate-borderline characterized malignancy. Generally, it can be seen in some regions including soft tissues, extremities, retroperitoneum, bones, liver, lung, brain, meninx, lymph nodes and skin. However, splenic involvement of HE is rarely seen condi-
In the current study, a literature review of splenic HE was conducted following a search of PubMed using the key words hemangioendothelioma and spleen or splenic hemangioendothelioma. Including seven cases in total, five of them were adults and two were in the pediatric age group. Non-specific symptoms such as weakness or abdominal swelling can be seen in patients with HE. In addition to these findings, palpable mass, hematologic abnormalities, hypo-hypersplenism, normochromic-normocytic anemia, trauma related consumption coagulopathy splenic rupture organ metastases and splenic rupture have been reported.1,2,7-10

In the literature, Kaw et al. reported a case of 48-year-old male patient who had been examined for chronic anemia and later, HE detected in the spleen. It was reported that the anemia was recovered during the follow up after the splenectomy.2 Another case of 58-year-old female patient who admitted with abdominal pain. In this case HE was detected after splenectomy.1 Fuchs et al. reported a 78-year-old female patient with splenomegaly, HE diagnosed after splenectomy for the risk of splenic rupture. No metastasis had been detected, however 6 months after splenectomy, the patient died.11 In another case, splenic HE was diagnosed in a newborn infant who had segmental splenectomy due to hypovolemic shock which had been caused by spontaneous rupture of the spleen; a year later, splenic functions were totally normal.12 By now, only two pediatric cases have been reported in literature; the first case was a 9-year-old male with HE of the liver and spleen who had received a partial splenectomy, however, he succumbed to consumptive coagulopathy.9 The second one, was the case of a 9-year-old female patient with splenic HE, who underwent a partial splenectomy. The patient showed no recurrence or complications during the 15-month follow up.13 In our case, the only presented symptom was left-side abdominal pain. Hematological parameters were completely normal. After the splenectomy, the patient was fol-
lowed up to 2 years revealed normal laboratory and physical examinations findings.

However definitive diagnosis cannot always be made by imaging. In general, radiological imaging in primary vascular tumors of spleen is not specific. In abdominal CT, the vascular composition of neoplasm can only be evaluated when the contrast material is given. It was very difficult to diagnose HE in our patient, because the images of US, MRI and CT were nonspecific for diagnosis as well as for metastasis. The patient was operated due to multiple lesions in the spleen and only after the pathological study, splenic epithelioid HE was diagnosed.

Eventhough, HE is the rarest tumor among non-lymphoid vascular splenic tumors; its clinical course is variable from benign hemangioma to malign hemangiosarcoma. Despite it is an intermediate-borderline tumor, rupture related to the growth of tumor or the possibility of metastasis can make this tumor as in malign characteristic. Excision of the mass before developing to be in the larger sizes, provides it to have good outcome results. Because early diagnosis and treatment may prevent conditions such as widespread metastasis, splenic rupture and operative complications. We strongly emphasize its clinical consideration and importance in differential diagnosis.

REFERENCES