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Cystic splenic hamartoma with prominent endothelial proliferation mimicking malignancy: a case report

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SUMMARY

Splenic hamartoma is an uncommon benign lesion and usually determined incidentally. A 44-year-old female patient admitted with abdominal pain. There was not any history of trauma. During the ultrasonographic examination a cystic mass was seen in the upper pole of the spleen. Only anemia was detected biochemically, and all other laboratory findings were normal. The patient underent splenectomy. Grossly, a cystic mass including solid component was seen in the upper pole of the spleen. Histopathologically, cystic component did not have lining epithelium. Solid component included sinus like clefts, broad cordons and areas of prominent endothelial proliferation. The lesion was diagnosed as hamartoma. In this report, the clinical and histopathological features of a case of splenic hamartoma with prominent endothelial proliferation that may be easily confused with malignancy is presented.

Key words: *Cystic splenic hamartoma, endothelial proliferation*

ÖZET

Maligniteyi taklit eden belirgin endotelyal proliferasyon gösteren kistik splenik hamartoma: olgu sunumu

Splenik hamartoma nadir görülen benign bir lezyondur ve genellikle rastlantısal olarak saptanır. Kırk dört yaşında kadın hasta karın ağrısı şikayeti ile başvurdu. Travma öyküsü yoktu. Batın ultrasonografisi sırasında dalağın üst kutbunda kistik kitle görüldü. Biyokimyasal incelemede sadece anemi saptanmış olup, diğer tüm laboratuvar incelemeleri normaldi. Hastaya splenektomi yapıldı. Makroskobik olarak, dalağın üst kutbunda solid komponent de içeren kistik kitle görüldü. Histopatolojik olarak kistik komponentin döşeyici epitelinin bulunmadığı görüldü. Solid komponent, sinüs benzeri yarıklar, geniş kordonlar ve belirgin endotelyal proliferasyon alanları içermekteydi. Lezyon hamartoma olarak rapor edildi. Bu makalede maligniteler ile karıştırılabilecek, belirgin endotelyal proliferasyon içeren bir splenik hamartoma olgusunun klinik ve histopatolojik özellikleri sunulmuştur.

Anahtar kelimeler: *Kistik splenik hamartoma, endotelyal proliferasyon*

Introduction

Splenic hamartoma is a benign uncommon lesion, first described by Rokitansky in 1861 (1-5). There have been cases over 100 so far (1). It has been also named as splenoma and splenadenoma, hamartoma, hemangioma, lymphangioendothelioma, angiomatosis, hemangiomatosis, hymphangioma, hyperplastic nodule and tumor-like congenital malformation (1-6). It has been claimed that there have been several pathogenetic mechanisms such as congenital, neoplastic, posttraumatic and hamartomatous according to the pathogenesis of the splenic hamartoma (1-6). It has been previously considered as splenoma and splenadenoma. Later it has been claimed that the splenic hamartoma may be a special form of a hemangioma/lymphangioma. These suggestions have supported that this lesion can be neoplastic. But, the most favorite suggestion related to the pathogenesis of the splenic hamartoma is tumor-like congenital malformation that occurs as result of the abnormal organization of the normal splenic red pulp elements (1-3).

Although the splenic hamartoma is seen at every age, it is frequently detected in older people (1-2). It is usually asymptomatic and incidentally determined during the autopsy, radiological studies and surgical staging of Hodgkin disease.

Some complications like trombocytopenia and anemia can be shown due to sequestration of the hemapoetic cells in some patients with splenic hamartoma. Sometimes, the splenic hamartoma can undergo cystic degeneration, in turn, constituting pseudocyst. Pseudocyst appears as a complication in the splenic hamartoma. The association of the pseudocyst and the splenic hamartoma is rather uncommon. It may hemorrhage inside the lumen and even rupture outside the lumen in some cases. Moreover, the prominent endothelial proliferations in the splenic hamartoma may constitute atypical features. So, these cases can be misdiagnosed as malignant.

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Case Report

A 44-year-old female patient had been suffering from nonspecific abdominal pain, weakness and fatigue for one year. There was not any trauma except for the right nephrolitotomy operation, which was performed 14 years ago. The routine laboratory findings were normal except for the anemia (hemoglobin: 9.0 gr/dL, hematocrit: 27.4%). The cystic mass that was 11 cm in diameter was determined in the upper pole of the spleen during the abdominal ultrasonographic and tomographic examinations (Figure 1). In turn, splenectomy was performed. She was followed for 8 years and maintained her life healthy.

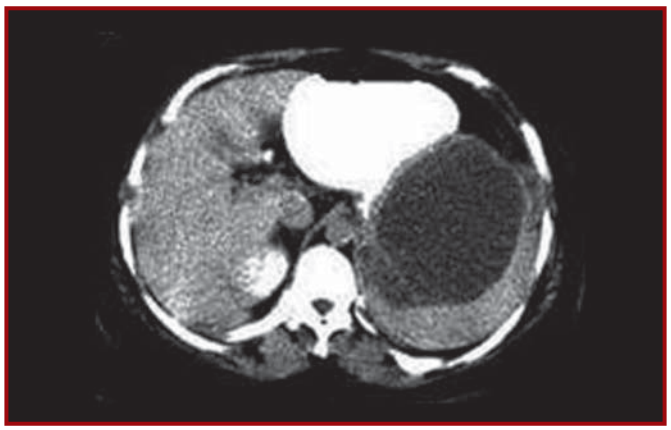

Figure 1. The magnetic resonance imaging appearance of the cystic splenic hamartoma

Grossly, the spleen was 20x17x5 cm in size. There was cystic mass in the upper pole of the spleen. It was 11x10x10 cm in size and its wall thickness ranged between 0.4 and 0.6 cm. In one side of the cystic mass a solid brownish hamartomatous component was noticed protruding towards the cystic lumen. This component was 6x2.5x2 cm in size, and it contained focal microcystic areas. There was abundant brownish granular material within the cystic lumen. The spleen was normal at the other sites (Figure 2).

Histopathologically, the cyst with thick fibrous wall was not lining epithelium. The normal splenic border and the luminal surface of the hamartoma were surrounded with broad fibrous capsule. It consisted of the sinus like clefts and the cordons like structures mimicking the normal spleen red pulp. While lymphoid follicules were seen in the normal spleen, they were not present in the hamartoma (Figure 3A, 3B). The lymphocyte and macrophage clusters were present inside the sinusoid like structures of the hamartoma (Figure 4). The cordons of the hamartoma consisted of the polygonal cells with eosinophilic cytoplasm and oval-round nucleus

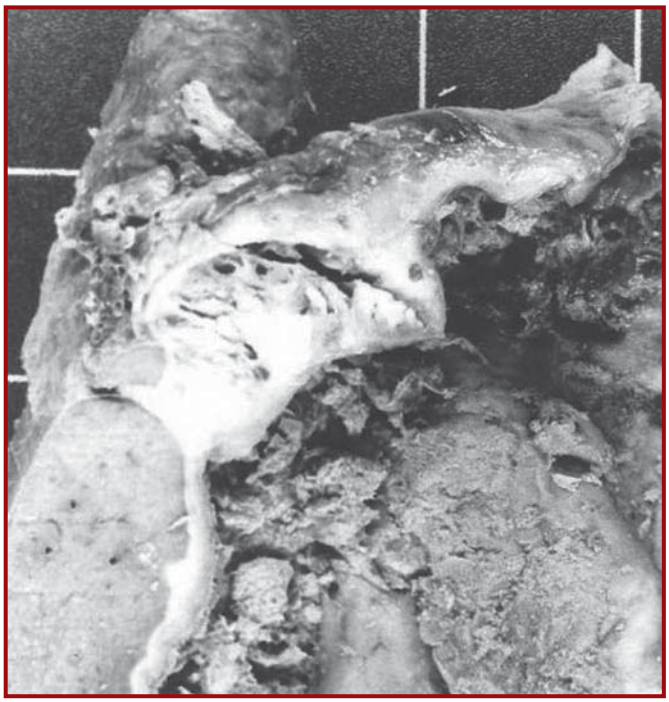

Figure 2. The macroscopic appearance of the cystic splenic hamartoma. Cystic lumen (in the right upper side), microcystic and solid hamartomatous area (in the left side) and normal splenic paranchyma (in the right bottom side)

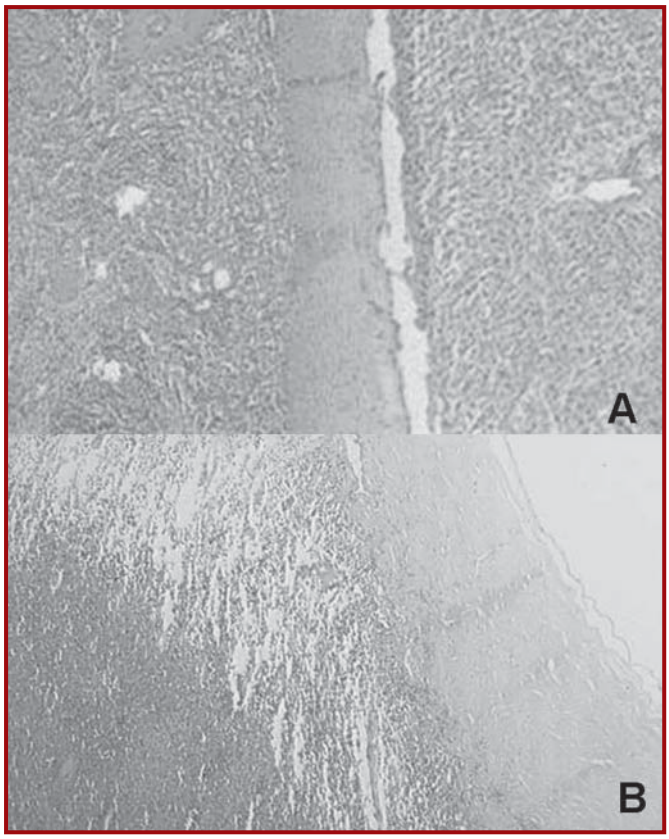

Figure 3 A. Normal splenic paranchyma (in the left side), broad fibrous capsule (in the middle area) and the splenic hamartoma (in the right side) (H&E, x100) **B.** Splenic hamartoma (in the left side) and cystic lumen (in the right side) (H&E, x100)

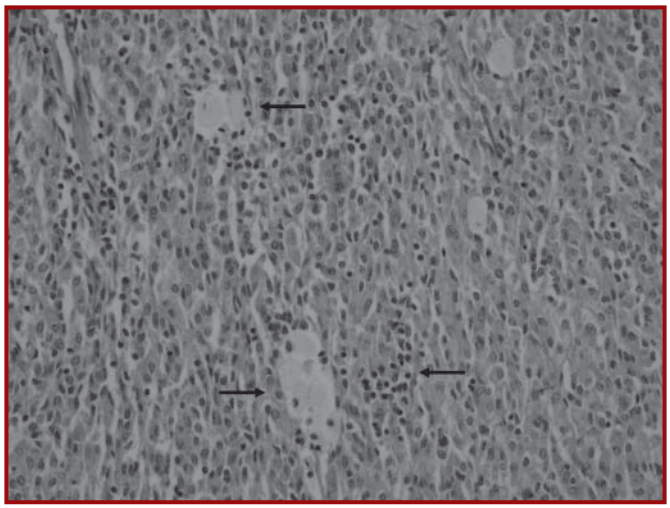

Figure 4. Lymphocyte and foamy macrophage clusters are shown inside sinusoid like structures (H&E, x200)

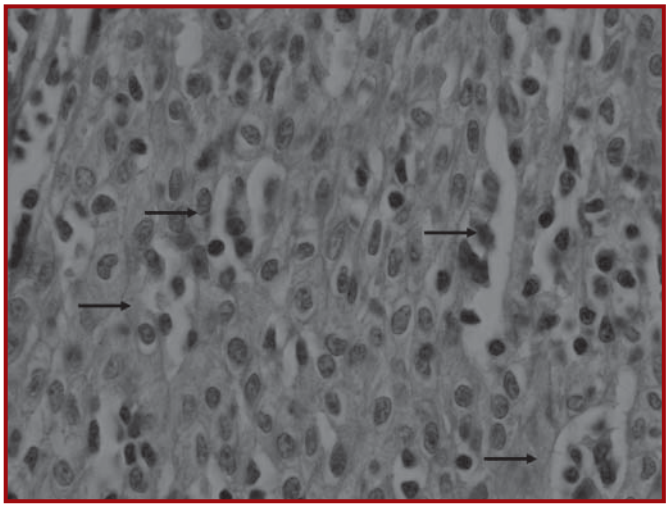

Figure 5. Polygonal cells with eosinophilic cytoplasm and sinusoid like clefts in the hamartomatous area (H&E, x400)

(Figure 5). There was slight pleomorphism in the polygonal cells. But, there was not mitotic activity. The sinusoid like structures were lined by proliferating endothelial cells with pleomorphic features. These structures can demonstrate cystic enlargements in some areas (Figure 6). These endothelial cells can demonstrate the reactive multinuclear features due to degenerative changes. Therefore, these endothelial cell proliferations can be misinterpreted as a malignant lesion (like undifferentiated carcinoma of the spleen), especially, in the complicated cases. There were focal hemosiderin pigment accumulations in the hamartoma.

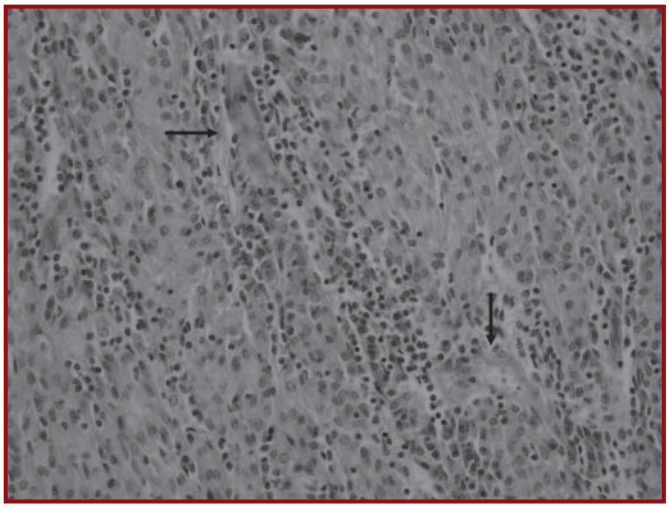

Figure 6. Sinusoid like structures lined proliferating endothelial cells with slight pleomorphic appearance and cystic enlargement (H&E, x200)

Discussion

Splenic hamartoma is an uncommon benign lesion that has been named in different ways. Splenic hamartoma and hemangioma have been previously considered as the same lesions due to the similarity of the clinic and morphologic features (1-3). But, the last studies have proven that these lesions are different entities. They have to be distinguished from each other (4). These lesions demonstrate the macroscopic and microscopic differences. Splenic hamartoma is homogen dark brownish lesion, while the hemangiomas are white lesions that are usually satellite scar in their center. The hemangioma consists of the small vascular spaces lined by flattened endothelial cells and is surrounded by thin fibrous tissue. Splenic hamartoma usually consists of red pulp like cordons and sinus like clefts. The vascular spaces in the splenic hamartoma are uncommon than the hemangioma. In addition to these differential features, while the big vascular structures are found in the hemangioma, they are not seen in the splenic hamartoma. The lymphocyte clusters in the hamartoma cannot be seen in the hemangioma. Sometimes, these morphologic features may not be helpful in the differential diagnosis of the problematic lesions. CD8, which immunohistochemically stains splenic endothelial cells may be useful in the differential diagnosis (4). The pseudocystic change can also be seen in the hemangioma as result of the cystic degeneration (1). Our case is clear and can be easily differentiated from the hemangioma. The giant cell formations and pleomorphic features of the endothelial proliferations in some areas may lead to misdiagnosis. Therefore, these lesions have to be closely followed because of the malignancy possibility. Our case was consulted. But, it was interpreted as the undifferentiated carcinoma of the spleen. We interpreted as the splenic hamartoma showing the pseudocystic changes. Afterwards, we have closely followed the patient for 108 months. The patient

has continued her life in a healthy way without any problem.

Splenic hamartoma is uncommon, and should be distinguished from the primer hemangioma and malignant lesions of the spleen. The association of the pseudocyst and splenic hamartoma is very uncommon as well (1). We considered that it emerged as a complication in the background of the splenic hamartoma. It is necessary to be careful because the prominent endothelial proliferations in the sinusoid like clefts and the slight pleomorphic polygonal cells in the red pulp like cordons of the splenic hamartoma can be confused with malignancy by mistake. These histopathologic features should be evaluated in detail, and these patients should be closely followed.

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