Symptomatic Splenic Hamartoma: A Case Treated with Partial Splenectomy and Review of the Literature

Semptomatik Splenik Hamartom: Parsiyel Splenektomi ile Tedavi Edilen Bir Olgu ve Literatürün Gözden Geçirilmesi

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ABSTRACT Splenic hamartoma is a rarely diagnosed tumor in children. Most patients with splenic hamartomas are asymptomatic and a few have clinical findings. Most frequently reported symptoms include pancytopenia, anemia, and thrombocytopenia. Less commonly, fever, malaise, and weight loss have been reported. In the last decades, most of the splenic hamartomas were found incidentally during laparatomy or autopsy. The use of modern radiologic imaging techniques such as sonography, computed tomography, radionuclide scintigraphy, and magnetic resonance imaging showed that splenic hamartoma was not as uncommon as it was thought previously. We presented a 28-month-old girl who had a history of frequent infections, fever and anemia. On the physical examination, she had a palpable mass in the left upper quadrant of her abdomen. The sonography and computed tomography revealed that she had a mass of 5 x 4 x 4 cm in the lower pole of her spleen first thought to be a splenic hamartoma. We excised the splenic mass by partial splenectomy. The histopathological findings were compatible with splenic hamartoma. The reported patient is the third case of symptomatic splenic hamartoma in the English treated with partial splenectomy. Despite numerous imaging modalities available, it is difficult to base the a definitive diagnosis of splenic hamartoma solely on imaging. It is often necessary to obtain a tissue diagnosis to eliminate the possibility of malignancy. Spleen has important hematologic and immunologic functions especially in children, therefore, spleen-preserving surgery is an acceptable and desirable principle in the treatment of benign splenic lesions.

Key Words: Spleen, hamartoma, splenectomy, child

ÖZET Splenik hamartom, çocuklarda nadir olarak tanı alan bir tümördür. Hastaların çoğu asemptomatiktir ve çok azında klinik bulgu vardır. Bildirilen semptomlar sıklıkla pansitopeni, anemi ve trombositopenidir. Daha az sıklıkla, ateş, halsizlik ve kilo kaybı da görülebilir. Geçmiş yıllarda splenik hamartomların çoğunluğu rastlantısal olarak laparatomi ve otopsi sırasında saptanmaktaydı. Sonografi, bilgisayarlı tomografi, radyonüklit sintigrafi ve manyetik rezonans gibi modern radyolojik görüntüleme yöntemlerinin kullanımı, splenik hamartomların önceden düşünüldüğü kadar nadir olmadığını göstermiştir. Bu çalışmada, öyküsünde sık enfeksiyon, ateş ve anemi olan 28 aylık bir kız çocuğu sunuldu. Fizik muayenesinde, karın sol üst kadranında ele gelen kitlesi mevcut olan hastanın sonografik ve bilgisayarlı tomografi çalışmalarında dalak alt polünde 5 x 4 x 4 cm boyutlarında öncelikle splenik hamartom olduğu düşünülen kitle izlendi. Splenik kitle parsiyel splenektomi ile çıkarıldı. Histopatolojik bulgular splenik hamartom ile uyumluydu. Bildirilen hasta, İngilizce literatürde parsiyel splenektomi ile tedavi edilen üçüncü semptomatik splenik hamartomlu olgudur. Çok sayıda görüntüleme çalışması olmasına rağmen splenik hamartomun kesin tanısının yalnızca görüntüleme yöntemleri üzerine kurulması zordur. Malignite olasılığını elimine etmek amacıyla sıklıkla doku tanısına başvurmak gerekir. Dalağın özellikle de çocuklarda önemli hematolojik ve immünolojik görevleri vardır; bundan dolayı dalak-koruyucu cerrahi, selim splenik lezyonlar için uygun ve kabul edilebilir bir tekniktir.

Anahtar Kelimeler: Dalak, hamartom, parsiyel, çocuk

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Hamartomas of the spleen are unusual lesions. The first case of splenic hamartoma was described by Rokitansky in 1861.[1] Von Falkowski reported the first case of splenic hamartoma in a pediatric patient in 1914.[2] Before the use of modern radiological imaging techniques, splenic hemartomas were considered rare lesions and were only found incidentally during laparatomy or autopsy. One third of the cases reported in the past 135 years were diagnosed in the last decade after the employment of modern radiological techniques such as sonography, computed tomography (CT), radionuclide scintigraphy, and magnetic resonance imaging (MRI).

Most patients with splenic hamartomas are asymptomatic and only a few of them have clinical findings. In 1953, Videbaek was the first who reported the association of splenic hamartoma with hematologic disorders in a 30-year-old woman with the onset of her symptoms in childhood.[2] Since then, only 24 symptomatic cases with splenic hamartoma were reported in the pediatric patients and only 2 were treated with partial splenectomy. Here, we reported the third case with symptomatic splenic hemartoma who was treated with partial splenectomy.

CASE REPORT

In March 2005, a 28-month-old girl complaining of fever and rhinorrhea was admitted to our hospital. On physical examination, she had fever, signs of upper respiratory system infections, fever, anemia and she was under treatment with ferrum preparations for the last three months. Laboratory investigation was normal except for mild anemia (Hgb 9.7 g/dL) and leucocytosis (WBC 10000/mm^3). An abdominal ultrasound performed for the palpable mass on the upper quadrant revealed a heterogenic hypoechoic 5 x 4 x 4 cm mass with evident vascularization in the lower pole of the spleen. CT scan showed that the craniocaudal lenght of the spleen was 105 mm and the same mass, imaged at the same localization with the sonography thought to be a splenic hamartoma (Figure 1). We performed an elective partial splenectomy for both the definitive diagnosis and treatment of the mass (Figure 2). The histopathological work-up of the surgical specimen, including immunohistochemical investigations, confirmed the diagnosis of splenic hamartoma. The lesion was well demarcated from the adjacent normal spleen. There was hypercellular and paucicellular areas in the lesion. The cellular areas were composed of haphazardly arranged vascular spaces lined by plump endothelial cells with eosinophils, plasma cells, and macrophages simulating red pulp. There was no well-formed white pulp in the lesion (Figure 3). The rest of the spleen showed unremarkable histology of red and white pulps. Immunostaning of the lesion showed intense positivity for factor VIII of the well-formed vessels and sinusoidal lining cells.

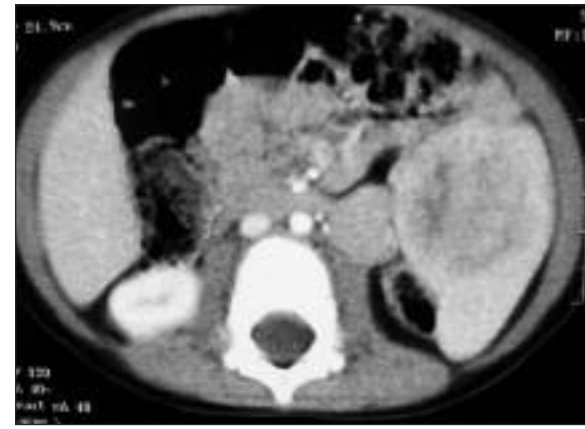

FIGURE 1: Abdominal CT image showing the splenic hamartoma in the lower pole of the spleen.

The patient's course was uneventful. She was seen in the outpatient clinic three months after the surgical intervention. Her physical examination and laboratory findings were normal (Hg 12.1 g/dL).

DISCUSSION

Primary nonlymphomatous tumors of the spleen are uncommon. Most are either cysts or hemangiomas. Hamartomas of the spleen are rarely reported entities. The incidence of the splenic hamartomas has been reported to be 3 in 200.000 splenectomies.[2] Reviews that accompany some case reports in the literature indicate that 20% of hamartomas occur in children. Synonyms for this tumor included splenomas, spleen within spleen, splenoadenoma, post-travmatic scars, fibrotic nodules, and hyperplastic nodules.

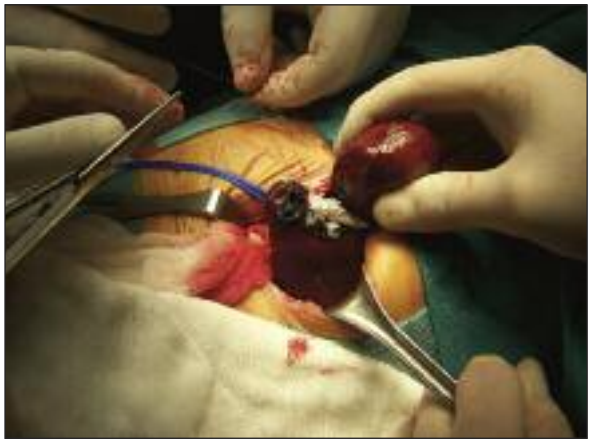

FIGURE 2: Operative photograph of the partial splenectomy procedure.

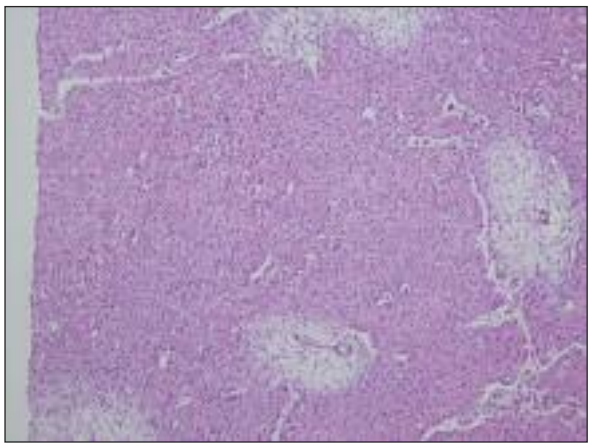

FIGURE 3: Splenic hamartoma with haphazardly arranged vascular spaces lined by plump endothelial cells with eosinophils, plasma cells and macrophages simulating the red pulp (HE, x200).

In 1907, Albrecht defined hamartomas as non-neoplastic tumors composed of an aberrant mixture of the normal tissue components of the organ in which they were found. Mordasini described two types of splenic hamartomas. White pulp hamartomas are composed entirely of lymphoid tissue, whereas red pulp hamartomas are formed by a complex of sinuses and structures equivalent to the red pulp of the spleen. We believe that histopathological classifying of splenic hamartomas as 'red pulp' or 'white pulp' is improper. Because; a review of previous reports indicates that a mixture of these types is common; so a rigid classification may not be warranted.[3]

Although splenic hamartomas are rare, their incidence increased with the use of modern radiologic imaging techniques in the last two decades. Hamartomas are hyperechoic when compared to the normal spleen on ultrasound, sometimes with cystic components. On CT, such masses usually create an irregularity in the contour of the spleen, and are almost isodense when compared to the normal tissue thus can be difficult to detect. Hamartomas are isointense relative to normal splenic tissue on T1-weighted MR images and heterogenously hyperintense on T2-weighted images, and they can demonstrate prolonged, heterogeneous enhancement after intravenous administration of contrast material.[4] Technetium-labeled sulphur colloid scan may demonstrate some uptake of radiotracer, but there is generally less accumulation in the hamartoma compared to the splenic parenchyma.[5] Despite numerous imaging modalities available, it is difficult to base the definitive diagnosis of a splenic hamartoma solely on imaging. The differential diagnosis of a splenic mass should also include epidermoid and simple cysts, granuloma, sarcoma, hemangioma, lymphoma, and leukemia. It is often necessary to obtain a histopathological diagnosis to eliminate the possibility of a malignancy.

Splenic hamartomas may be solitary or multiple and are usually well defined, however they are not encapsulated. While the multiple lesions may be treated with partial splenectomy as well. Preserving the spleen or conserving the splenic tissue is desirable in the surgical treatment of injuries to the spleen as well as of non-traumatic benign lesions. Spleen has important hematologic and immunologic functions especially in children. Post-splenectomy sepsis and overwhelming post-splenectomy infection (OPSI) syndrome are common after splenectomy.[6] Thus, splenic-tissue-conserving surgery is both an accepted and desirable principle in the surgical treatment of benign lesions of the spleen as mentioned in the current case.

Approximately 80% of splenic hamartomas produce no clinical symptoms; Twenty percent of the patients have symptoms, and the most common symptoms are associated with hematologic disorders such as anemia, thrombocytopenia or pancytopenia.[7] To our knowledge, only 24 pediatric

patients with symptomatic splenic hamartomas have been reported in the English Literature so far. Most of those patients had hematological disorders such as pancytopenia, thrombocytopenia, spherocytosis or various types of anemia.[8-14] An addition, most of these patients, had systemic, symptoms such as fever and lethargy.[7]

Apart from the 3 month-old infant who died after a spontaneous splenic rupture, all of the patients experienced a partial or complete improvement in their hematologic disorders after surgery.[1] The partial responders appear to be individuals with well-defined underlying disorders such as sickle cell anemia and hereditary spherocytosis.[7] We found only two cases with symptomatic splenic hemartoma who were treated with partial splenectomy in the English literature. We believe that the current case is the third case who was treated with partial splenectomy for a symptomatic splenic hemartoma.

The sequestration of blood elements within the lesion is proposed to be a major pathophysiologic mechanism of blood dyscrasia.[3] In the light of this information, removing the lesion by partial splenectomy should be adequate to alleviate the hematological disorders caused by the lesion itself. Therefore, we recommend performing a partial splenectomy for the removal of the lesion in selected patients, especially patients with solitary and the favorably located lesions.

In conclusion, splenic hamartomas should be included in the differential diagnosis of the splenic masses. Despite numerous imaging modalities available, a definitive diagnosis of a splenic hamartoma based solely on imaging techniques is not reliable. It is necessary to obtain a histopathological diagnosis to eliminate the possibility of a malignancy. In addition, spleen-preserving surgery is an acceptable and desirable principle in the treatment of benign splenic lesions.

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