Hamartoma – A Rare Benign Tumor of the Spleen: A Report of Four Cases

Dalağın Nadir Görülen Benign Tümörü, Hamartom: Dört Olgu Sunumu

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Abstract

Splenic hamartoma is an uncommon benign tumor. We retrospectively analyzed all patients who underwent splenectomy between May 2000 and June 2006 and four cases of splenic hamartoma were encountered. Three patients presented with mild abdominal pain. Abdominal ultrasonography was the first diagnostic step, which revealed a splenic mass. Doppler ultrasonography, computed tomography, and magnetic resonance were among the other diagnostic methods. An elective splenectomy was performed for all of the cases. Splenic hamartoma must be considered in the differential diagnosis of splenic masses unrelated to any other malignancy. Elective splenectomy is indicated due to hematological disorders or symptoms of pain and appreciation of the mass.

Keywords: Splenic mass, Hamartoma, Splenectomy

Anahtar Kelimeler: Dalak kitlesi, Hamartom, Splenektomi

Özet

Introduction

One pathological condition of the spleen is benign splenic tumors, which are classified into the categories of cyst, hemangioma, and hamartoma. Of these, splenic hamartoma is a rare, benign tumor, which usually presents asymptptomatically. In 1861, splenic hamartoma was first described by Rokitansky [1]. Since then, 170 to 190 cases have been reported in the literature. Hamartoma, which means ‘scribble error’, is a tumor-like proliferation of both the cells and tissues of the normal spleen. It affects all age groups, mostly the elderly population. The most common solid or complex cystic splenic mass in an asymptomatic patient is the splenic hemangioma. These types of tumors are usually detected accidentally or through post-mortem examination. Nevertheless, recent advances in imaging techniques have allowed the preoperative detection of splenic tumors [2].

We retrospectively reviewed all the patients seen at the Ege University School of Medicine, Department of General Surgery during a five year period and detected four cases of splenic hamartoma. The purpose of this study was to evaluate the optimal surgical approach and efficacy of treatment by analyzing demographic variables, clinical features, diagnostic methods, surgical technique, and follow-up.

Case Report

Clinical features of the patients are summarized in Table 1. There were two male and two female patients with a mean age of 55.7 years (range: 50-64 years). In three patients, the symptoms were hypochondriac pain and a feeling of mass-like distension in the left upper quadrant. In the other patient, a splenic mass was detected during whole-body imaging before adjuvant therapy to explore whether any metastasis had occurred. Laboratory findings revealed normal values for both complete blood count tests and blood chemistry tests in all patients. Abdominal ultrasonography (US) was the initial radiological study performed in all patients. Splenic masses were found at multiple locations in only one patient, whereas they were located in the inferior pole in two patients and in the superior pole in the other patient. In one patient who had a clinical history of larynx cancer, the splenic mass was determined to be hypervascular based on computed tomography (CT), and it was therefore considered to be a malignant tumor, mostly from metastasis. In one patient, magnetic resonance revealed benign characteristics. In only one patient was the splenic mass evaluated with Doppler ultrasonography for further radiological study; and it showed hypovascular tumoral mass and thrombosis of the splenic vein, accompanied by the findings of portal hypertension. Splenomegaly was found in two cases. The mean size of the tumors was 5.2 cm and ranged from 4 to 5.7 cm. None of the patients in this study were subjected to fine needle aspiration biopsy (FNAB) to determine the tumor histology as part of a preoperative assessment due to the risk of hemorrhage and inadequate evaluation.

An elective splenectomy procedure was performed in all cases. Pathological examination of the tumors confirmed the splenic hamartoma for all the patients.

The recovery period was uneventful and the mean length of the hospital stay was 6.2 days (range: 5-8 days). In the follow-up period, which ranged from 18 to 76 months, abdominal ultrasonography was performed for imaging studies. Fortunately, no recurrence has been observed thus far.

Discussion

Splenic hamartoma is an uncommon non-neoplastic lesion composed of an abnormal mixture of white and red pulp. Although most of the reports in the literature consist of adult patients of an average age of 40–50 years, some of the reports indicate that 20% of hamartomas occur in children. No reliable data exist on the incidence of splenic hamartoma. In autopsy studies, an incidence of 0.024-0.13% has been reported, while an incidence of 0.015%-2.7% was noted among the patients that had undergone a splenectomy [3]. In our study, the incidence of 1.2% is well correlated with available literature data.

The presenting symptoms are often dependent on the size of the hamartoma. Hematological disorders are particularly prevalent for children. The major symptoms are anemia and/or thrombocytopenia, as well as frequent infections [3]. In the present study, all of the patients were admitted to the hospital with nonspecific abdominal pain and a feeling of a bulging on the left side with no underlying disorders. Only one patient was diagnosed during total body imaging for the possibility of metastasis due to a previous history of larynx cancer. Laboratory studies revealed no hematological abnormalities or elevated tumor markers.

The association of splenic hamartomas with other malignant
cies is quite high. It has been shown that splenic hamartomas may arise from an acquired proliferative process. Therefore, if any newly recognized splenic mass is diagnosed in patients with a previous malignancy, hamartoma should be taken into account as a differential diagnosis. In our series, one patient had a previous diagnosis of larynx cancer [3].

An exact preoperative diagnosis is rarely established. Because the malignant counterpart of splenic hamartoma and splenic hemangioma are virtually identical, as well as the inflammatory pseudotumor and follicular dendritic cell sarcoma, the diagnosis must be confirmed by histopathological findings [4]. However, recent progress in imaging procedures has helped us detect splenic tumors and possibly appropriate treatments. These lesions may appear as single or multiple masses and ultrasonography is usually the first radiological method. In our series, only one patient was diagnosed with multiple masses. Doppler US, CT, and magnetic resonance (MR) are advanced methods for diagnosis. Doppler US may help to distinguish splenic hamartomas from other relatively common splenic tumors, such as hemangiomas or metastases [5]. A diagnosis of splenic hamartoma may be suggested when findings of increased blood flow on color Doppler images are seen in association with a homogeneous solid echogenic mass [3]. Nakanishi et al [6] reported that CT and MR are not useful, whereas US can be used to diagnose splenic hamartoma without the administration of a contrast material. Nevertheless, there are some reviews that advocate MR as an excellent imaging method for the diagnosis of splenic hamartoma [7]. In the present series, MR was performed on only one patient with suspicion of malignancy due to a hypervascular and calcified splenic mass and it was confirmed as a relatively benign mass. Recent studies have suggested that the diagnosis of hamartoma is feasible with the use of radioisotopes and radionuclide imaging. Serial nuclear scintigraphy is not helpful in the diagnosis, but both increased radioisotope and radionuclide uptake and no uptake have been reported thus far [8]. FNAB sampling has also been considered as a preoperative diagnostic method. However, Zissin et al. reported a patient with a missed diagnosis of hamartoma because it was confirmed as malign melanoma by preoperative FNAB [9]. FNAB can be considered dangerous both because of the risk of bleeding and because it is an ineffective diagnostic method for splenic hamartomas. In the present study, all of the patients were initially subjected to US; CT was performed on three patients and MR and DUS were each used on only one patient. FNAB was not performed on the patients due to the hypervascular findings in radiological studies. Arterial vascularization, calcification, and irregular margins of these masses, hamartomas, can conduct the clinician to the malignant tumors.

Whether the solid tumors of the spleen are benign or malignant, imaging studies may be helpful in predicting the diagnosis, but exact diagnosis often requires pathological examination of the mass. Surgical procedures for splenic hamartoma include splenectomy, partial splenectomy, laparoscopic splenectomy, and hand-assisted laparoscopic splenectomy [6]. In recent series, particularly in children, partial splenectomy has become the more popular surgical treatment of splenic masses due to the immunologic role of the spleen. However, the application of laparoscopic splenectomy for splenic hamartoma is still controversial because the final histological diagnosis may be affected if the spleen is fragmented during the retrieval of specimens. In our series, all of the patients were subjected to elective complete splenectomy. In three of them, partial splenectomy was not possible due to multiple lesions in one patient, malignancy history in another, and advanced age, hypervascularization, and calcification in the third patient. Due to thrombosis of the splenic vein, case four was not an appropriate candidate for partial splenectomy. However, in case of suspicion of malignancy, the spleen needs to be removed intact to avoid peritoneal dissemination of tumor cells. No accessory spleen tissue was detected through operative observations. This observation is important because continued hematological disturbances may be encountered in patients with previous hematological disorders via spillage of splenic tissue into the peritoneal cavity [10].

During the follow-up period, laboratory findings and USG are required to evaluate whether or not any intra-abdominal collection or recurrence is present. In our study, US control for all patients was performed during the follow-up period of 20 to 61 months (mean: 39 months). However, because of the benign pattern of these tumors, adjuvant therapy is usually not required.

In conclusion, if a suspicious splenic mass is detected, either incidentally or symptomatically, the most important part of the treatment procedure is to make a definitive diagnosis between benign and malignant mass. If splenic hamartoma is supported by radiological findings and clinical evaluation, an elective splenectomy should be performed to make a definitive diagnosis by histological examination.

<table>
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<th>Table 1. Clinical data of the patients.</th>
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US: ultrasonography, CT: computed tomography, MR: magnetic resonance

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