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Case Report

Angiosarcoma in Spleen - A Case Report

Md. Mustafizur Rahma[1], Afroza Kutubi[2], Dilip Kumar Ghosh[3], Abdur Rahim[4], Jahangir Alam[5], AZM Mostaque Hossain[6]

Abstract

Primary angiosarcoma of the spleen is a rare neoplasm with a very poor prognosis. We present here computed tomography and magnetic resonance imaging of a 65-year-old man with primary splenic angiosarcoma The literature on primary splenic angiosarcoma is reviewed and imaging variability of this rare tumor is discussed.

Mr. Hazi Abul Kalam, 65 years non-diabetic normotensive got admitted in Shaheed Suhrawardy medical college hospital with the complaints of anorexia with marked weight loss for last 1 year and feeling of a lump in left upper abdomen for last 10 months. On abdominal examination spleen was enlarged in long axis 10 cm from left costal margin, non tender, margin regular, firm in consistency and all others examinations revealed normal.

Ultrasonography reveals cholelithiasis with space occupying lesion in spleen otherwise normal study. FNAC of spleen no malignant cell or caseation necrosis but TB can not be excluded. Computed tomography (CT) scan of abdomen and CT guided FNAC shows adeno carcinoma of spleen. After consultation with oncologist laparotomy & spleenectomy done. Histopathology shows angiosarcoma. Patient were referred to oncologist for further management.

Keywords: *Spleen, Angiosarcoma, Computed Tomography, Magnetic Resonance Imaging.*

Introduction

Angiosarcoma is a rare malignant neoplasm of the endothelial cell, which represents less than 2% of all soft tissue sarcomas. Although primary involvement of the spleen is extremely rare, angiosarcoma is the most common primary nonlymphoid malignant lesion of the spleen. Since the first description of primary splenic angiosarcoma more than 100 cases have been reported[1]. It is a very aggressive neoplasm with a high metastatic rate and poor prognosis.

Metastasis is typically involves the liver, lungs, bone, bone marrow and lymphatic system. We present the computed tomography (CT) and magnetic resonance (MR) imaging findings of a patient with primary splenic angiosarcoma. The literature is reviewed and imaging variability of this rare neoplasm is discussed.

1. Asst. Prof. Surgery, ShSMC
2. Asst. Prof. Gynae & Obst, SSMC
3. Asst. Prof. Gastroenterology, ShSMC
4. Asst. Regt. Medicine, ShSMC
5. Asst. Prof. Surgery, ShSMC
6. Assoc. Prof. Surgery, ShSMC

Correspondence : Md. Mustafizur Rahman, Asst. Prof. Surgery, ShSMC, E-mail : drmustafiz@hotmail.com

Case Presentation

A 65-year-old man was admitted in Shaheed Suhrawardi Medical College Hospital with left upper quadrant pain and fatigue & feeling of a lump in left upper abdomen. There was no history of trauma or chemical agent exposure. Physical examination revealed splenomegaly. Anemia and thrombocytopenia were present. An abdominal ultrasonography (US) revealed space occupying lesion in spleen with cholelithiasis.

USG showed parenchymatous masses with heterogeneous echogenicity in the enlarged spleen. The liver was normal. Subsequent CT and MR examinations of the whole abdomen confirmed the sonographic findings. The variable-sized (1-7 cm in diameter) masses of the spleen were hypodense on CT scans. Calcification was not present The

splenic parenchyma was heterogeneous and hypointense on T1-weighted images and hyperintense on T2-weighted images compared to the liver parenchyma. MR imaging revealed ill-defined nodular lesions with low- or high-signal intensity on both T1- and T2-weighted images depending on the age of blood products or presence of necrosis The splenic parenchyma as well as the masses were enhanced heterogeneously after the administration of contrast material The differential diagnosis from CT and MR examinations included vascular splenic tumors or lymphoma.

The patient underwent splenectomy. The specimen measured 22x16x7 cm. Histopathologic examination disclosed primary angiosarcoma. The patient received two courses of multidrug regimen consisting of ifosfamide, vincristine and actinomycin D. Follow-up CT and abdominal ultrasonography at seven months after surgery revealed multiple metastatic lesions in the liver & upper abdomen. The patient died due to metastatic disease within eight days after second admission.

Discussion

Primary angiosarcoma of the spleen is an extremely rare neoplasm, with a reported incidence of only 0.14-0.25 cases per million[2]. It may develop at any age. The mean age at presentation is 59 years with a range between 14 months and 89 years[2-4]. There have been eight reported pediatric cases[5,6]. There is a slight predominance in men[2,4]. Clinical presentation of splenic angiosarcoma is variable. Patients usually present with left upper abdominal pain, fatigue, weight loss, and anorexia. Left upper abdominal pain is the most common symptom. It occurs in 75% to 83% of patients with splenic angiosarcoma[4,7]. Splenomegaly is the most common finding (in 68% of cases) at physical examination[4]. Hepatomegaly and left upper quadrant mass are the other common findings. The major complication is splenic rupture, which often leads to fatal hemoperitoneum[8].

Cytopenia (91%), leukocytosis (20%), thrombocytosis (5%) and elevated erythrocyte sedimentation rate (15%) are the abnormal laboratory findings[2]. Anemia and thrombocytopenia are detected in 75% to 81% and 14% to 55% of reported cases, respectively.

The pathogenesis of primary splenic angiosarcoma is still unknown. Exposure to some chemical agents, such as thorium dioxide, vinyl chloride, and arsenic have been implicated because of their association with hepatic angiosarcomas; however, no such association of these agents was found for splenic angiosarcomas in the large series of Neuhauser et al.[4] and Falk et al.[2]. Other reported causes of primary splenic angiosarcoma are exposure to ionizing radiation or chemotherapy for lymphoma[9,10]. Moreover, some authors claim that these tumors develop from preexisting benign counterparts, such as hemangioma or hemangioendothelioma[11,12]. No such preexisting tumor or risk factors were present in our patient. Primary splenic angiosarcomas are very aggressive neoplasms, with a median survival of five months irrespective of treatment. Neuhauser et al.[4] reported that 93% of patients died with disseminated tumor within 29 months. However, early diagnosis with splenectomy has a favorable survival rate[2,8]. Hsu et al.[5] reported that a patient in their study remained disease-free 162 months after splenectomy. This patient is the longest survivor of splenic angiosarcoma in the literature.

High incidence of metastasis (in 69%-100% of cases) was reported in previous series[2,4,13]. Metastasis occurs early and most often involves the liver. Neuhauser et al.[4] found that patients had metastasis to the liver (89%), lung (78%), lymph nodes (56%), and bone (44%). Bone marrow, gastrointestinal tract, brain, adrenal gland, omentum, and peritoneum metastases were also reported in the literature[14,15]. Metastasis can exist after initial therapy. Liver metastases occurred in three of six patients after splenectomy in the study of Hsu et al.5. The intervals between the operation and metastasis were 3, 33 and 56 months. In our case, liver metastasis occurred seven months after splenectomy.

Biopsy is contraindicated in splenic angiosarcoma because of high risk of rupture. Therefore, histologic studies can only be made after splenectomy. The histologic findings of splenic angiosarcoma are similar to those of angiosarcomas seen in other parts of the body. It arises from the endothelial lining of the splenic blood vessels[3]. Cut specimens usually

reveal poorly defined nodular masses. Diffuse involvement of the spleen is common, and replacement of the entire splenic parenchyma by the tumor may be seen. Solitary mass is a less common finding. Hemorrhage and necrosis are frequently seen within the tumor.

The radiologic appearances of splenic angiosarcoma are variable and nonspecific. The most useful imaging studies for diagnosis are US, CT and MR imaging. US findings are nonspecific in many cases16. The most common findings are splenomegaly and ill-defined solid and cystic masses with heterogeneous echotexture[17,18]. The areas of hemorrhage and necrosis within the tumors are frequently seen as cystic areas. Increased blood flow may be detected in the solid part of the masses on color Doppler images.

CT may reveal an enlarged spleen with hypo- or hyperattenuating areas on nonenhanced scans[19]. Areas of hyperattenuation are likely to reflect acute hemorrhage or hemosiderin deposits. Scattered punctate or massive calcification can be seen on nonenhanced scans20. On contrast-enhanced CT scans, the tumors may exhibit peripheral or heterogeneous contrast enhancement similar to that of hepatic cavernous hemangiomas[21,22].

Ill-defined nodular lesions with low- or high-signal intensity may be seen on both T1- and T2-weighted images depending on the age of blood products and presence of necrosis. High-signal intensity on both T1- and T2-weighted images is related to subacute hemorrhage or tumor necrosis, and low-signal intensity is related to chronic hemorrhage or fibrosis within the tumor. The appearance of splenic angiosarcoma on contrast-enhanced T1-weighted images consists of hyperintense masses with focal areas of nonenhancement depending on the hemorrhage and necrosis within the tumor.

The differential diagnosis should include lymphoma, metastatic tumors, and other splenic vascular lesions such as hemangioma23. Differentiation between angiosarcoma and these tumors is difficult for radiologists because of overlapping imaging findings. Therefore, a definitive diagnosis can only be made by histopathologic examinations after splenectomy.

Conclusion

Primary splenic angiosarcoma is a rare and aggressive neoplasm that is difficult to diagnose. The radiologic appearance varies from one to several nodular, well-defined masses to large ill-defined areas of hemorrhage and necrosis. Many radiological methods are available; however, MR imaging seems to be more effective in the overall assessment and staging of splenic angiosarcoma. Although rare, the possibility of hemangiosarcoma should be kept in mind in assessing splenic tumors.

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