Current Problems in Diagnostic Radiology 45 (2016) 284–287

Current Problems in Diagnostic Radiology

journal homepage: www.cpdrjournal.com

Primary Splenic Angiosarcoma: Clinical and Imaging Manifestations of This Rare Aggressive Neoplasm

Ali Batouli, MD[a,*], Samuel W. Fairbrother, MD[a], Jan F. Silverman, MD[b], Maria De Los Angeles Muniz, MD[b], Kevin B. Taylor, MD[c], Mark A. Welnick, MD[a], Sheri A. Mancini, MD[d], Matthew S. Hartman, MD[a]

[a] Department of Radiology, Allegheny Health Network, Pittsburgh, PA
[b] Department of Pathology, Allegheny Health Network, Pittsburgh, PA
[c] Department of Radiology, University of Pittsburgh Medical Center, Pittsburgh, PA
[d] Department of Surgery, Allegheny Health Network, Pittsburgh, PA

Primary splenic angiosarcoma is a rare and fatal neoplasm arising from vascular endothelial cells within the spleen. With an incidence of 2 cases per 10 million people worldwide, the diagnosis and treatment of this rare entity is unfamiliar and challenging. We describe the case of a previously healthy 45-year-old woman who presented with vague upper-abdominal pain and was found to have a splenic mass on computed tomography. The patient underwent laparoscopic splenectomy and was found to have splenic angiosarcoma on microscopic evaluation. Although specific radiologic diagnosis is not possible, bringing the possibility of primary splenic angiosarcoma to the ordering clinician's attention has the potential to hasten treatment and improve patient outcomes. This case highlights the importance for radiologists to be aware of this rare neoplasm and to consider it in the differential when encountering a heterogeneously enhancing splenic mass.

Introduction

We present the case of a 45-year-old woman with primary angiosarcoma of the spleen, an extremely rare and lethal disease. First described by Langhans in 1879,[1] splenic angiosarcoma is one of the least common neoplasms, with an incidence of approximately 0.2 cases per million. Despite its rarity, the disease is the most common nonlymphoid primary splenic neoplasm. The diagnosis carries a poor prognosis and often presents in late stage with metastatic disease. Splenectomy is the mainstay of therapy, and chemotherapy and radiation therapy are ineffective in achieving a cure in this deadly disease.[2]

Case

A 45 year old woman with a past medical history significant only for nephrolithiasis presented with left upper quadrant and left flank pain. Social, family and past surgical histories were unremarkable. Laboratory studies, including complete blood count, basic metabolic panel, and erythrocyte sedimentation rate were within normal limits.

Unenhanced computed tomography (CT) of the abdomen and pelvis performed as part of the initial workup revealed splenomegaly with a large heterogeneous splenic mass as well as nonobstructive nephrolithiasis (Fig 1). Although the latter finding was present on a CT performed 9 years earlier for flank pain, the splenic mass was new. A contrast-enhanced CT of the abdomen was ordered for further characterization, which revealed irregular heterogeneous and nodular peripheral enhancement with a large centrally hypoattenuating area consistent with necrosis or non-contrast filled vascular spaces (Fig 2). The mass measured 5.7 × 9.2 cm and did not invade through the splenic capsule. Although there was no associated lymphadenopathy, multiple sub-5 mm peripherally enhancing and centrally hypoattenuating lesions were noted within the liver concerning for possible metastatic disease. Differential considerations for the findings included lymphoma, metastatic disease, vascular neoplasm, and abscess. Hemangioma and other benign vascular lesion were deemed less likely owing to the aggressive appearance of the splenic lesion as well as the associated hepatic lesions. Given the associated hepatic lesions and the lack of laboratory and vital sign abnormalities, splenic abscess was also thought unlikely.

Magnetic resonance imaging (MRI) of the abdomen performed 1 month later redemonstrated the peripherally enhancing, heterogeneous splenic mass, which now measured 5.7 × 10 cm. The mass contained large areas of T1 hypointensity and associated T2 hyperintensity (Fig 3A-C). The lesion enhanced heterogeneously with a peripheral predominance, likely secondary to central necrosis and central vascular spaces that had not yet filled with contrast (Fig 3D and E). A small area of T1 hyperintensity consistent with hemorrhage was noted centrally (Fig 3C). Multiple

* Reprint requests: Ali Batouli, MD, Allegheny Health Network, 320 E. North St. Pittsburgh, PA 15214
E-mail address: abatouli@wpahs.org (A. Batouli).

http://dx.doi.org/10.1067/j.cpradiol.2015.07.004
0363-0188/

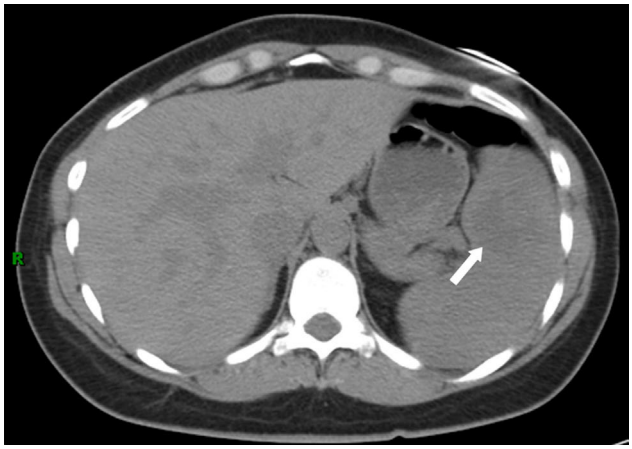

Fig. 1. Axial unenhanced CT of the abdomen and pelvis revealed splenomegaly with a large heterogeneous splenic mass (white arrow). (Color version of figure is available online.)

T2 hyperintense, peripherally enhancing masses were diffusely scattered throughout the liver (Fig 3A-E). Given the irregular peripheral predominant enhancement pattern, angiosarcoma was considered higher on the differential, although lymphoma or other neoplastic processes were still favored statistically. A contrast-enhanced CT of the chest performed as part of the metastatic workup did not demonstrate disease in the chest.

The patient underwent an open splenectomy and wedge biopsy of the liver 7 weeks after the initial CT scan. In addition to the splenic mass and multiple liver lesions documented on diagnostic imaging, a single omental nodule was noted intraoperatively. There was no visible peritoneal disease or free intraperitoneal fluid. Gross pathology revealed an enlarged spleen with a nodular tan-pink surface to purple-gray surface and intact capsule (Fig 4). Innumerable soft and firm surface nodules were present. Sectioning revealed the vast majority of the splenic parenchyma to be involved in the neoplastic process with multinodular round and irregularly shaped lesions. Microscopic examination established the diagnosis of a splenic angiosarcoma with metastatic spread to the liver and omentum. Fig 5 demonstrates microscopic images of the tumor with hematoxylin and eosin stain showing vascular channels lined by atypical endothelial cells with enlarged nuclei. This appearance was consistent with the diagnosis of angiosarcoma. The liver lesions and omental nodules also demonstrated a similar appearance.

The patient's clinical status continued to deteriorate postsplenectomy. She was started on paclitaxel one month after surgery, with a chemotherapeutic course complicated by neutropenia and multiple infections. A repeat CT of the abdomen and pelvis done shortly after the start of chemotherapy demonstrated a substantial increase in hepatic metastatic burden (Fig 6). The patient continued to deteriorate and passed away approximately 5 months after her initial CT examination.

Discussion

Despite being known as the most common nonlymphoid primary splenic neoplasm, splenic angiosarcoma is an extremely rare tumor that arises from vascular endothelial cells within the spleen. Its annual incidence is approximately 0.2 cases per million individuals.[2] As such, it is often near the bottom of the differential diagnosis of radiologists encountering an aggressive splenic lesion. Just as in this case, the tumor most commonly presents in middle aged and older adults, though pediatric cases have also been reported.[2-5] There is no known risk factor for this rare neoplasm.[2] Although associations with prior chemotherapy and radiation therapy have been proposed, most cases of the disease are not associated with either. Unlike hepatic angiosarcoma, no statistical link has been found between splenic angiosarcoma and exposure to chemicals such as thorium dioxide, vinyl chloride, or arsenic.[2] Our patient had no specific risk factors for angiosarcoma or neoplasm in general.

Nonspecific upper-abdominal pain with or without fever, weight loss, and fatigue are the most common presenting symptoms.[2,5,6] Physical examination findings of splenomegaly and palpable left upper quadrant mass are common. Owing to invasion through the capsule and significant mass effect, splenic rupture, which was not present in our case, has been reported in 13%-32% of patients.[2-4] Laboratory abnormalities include anemia (reported in up to 81% of cases), thrombocytopenia (reported in up to 40% of cases), and less commonly, leukocytosis.[5-7] However, in our patient, the initial laboratory values were within normal limits.

Splenic angiosarcoma has high rates of regional and distant metastatic disease and local recurrence and is therefore most often fatal. Similar to our patient, approximately 70% of patients present initially with disease involvement of the liver, which is the most common site of metastasis. The overall rate of metastatic disease is high, with up to 90% of patients developing metastases to the liver. Lower but still substantial rates of metastasis are found in the lungs, lymph nodes, osseous structures, and brain.[2,8] The disease is most often fatal, with the vast majority of patients dying within 1 year of diagnosis, with or without metastatic disease at presentation.[2,3]

Radiologically, splenic angiosarcomas present as a large multinodular mass with areas of central necrosis, infarct, and hemorrhage as seen in our case. Imaging and pathologic findings of necrosis are seen in approximately 83% of patients.[9] On ultrasound, splenic angiosarcomas present as mixed cystic and solid masses with heterogeneous echogenicity.[9-11] CT also shows a heterogeneous mass with generally low central attenuation except for areas of acute hemorrhage, which demonstrate high attenuation.[4,9,11] On MRI, the masses are generally hypointense to normal splenic tissue on T1 and T2. However, foci of internal necrosis and subacute hemorrhage, which may involve most of the mass, show high T2 signal intensity. Additionally, areas of very low signal on

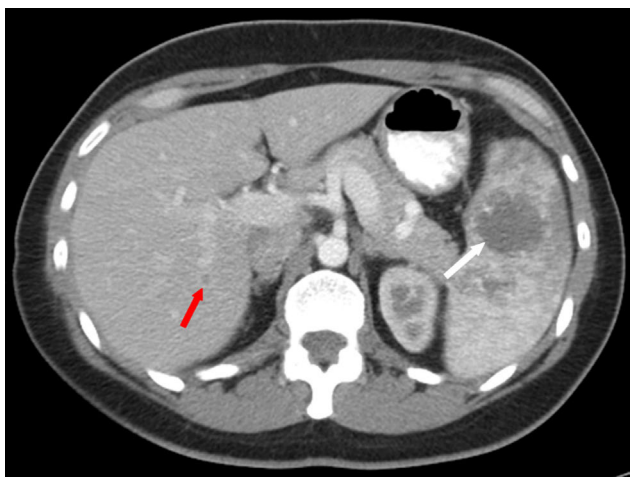

Fig. 2. Axial enhanced CT of the abdomen and pelvis performed shortly after the initial CT showed a large splenic mass (white arrow) with irregular, heterogeneous and nodular peripheral enhancement as well large centrally hypoattenuating areas consistent with necrosis or noncontrast filled vascular spaces. Multiple enhancing and centrally hypoattenuating lesions consistent with metastases were also noted in the liver (red arrow). (Color version of figure is available online.)