Case Report

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Angiosarcoma of the Breast and Spleen: A Case Report with Imaging and Pathologic Findings

유방 및 비장의 혈관육종: 영상 소견 및 병리 소견을 포함한 증례 보고

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Angiosarcoma of the breast is a rare malignant sarcoma of endovascular origin that presents with various radiological findings. We encountered a case of angiosarcoma of the breast presenting as an irregular indistinct hyperechoic mass on ultrasonography, with a large single angiosarcoma of the spleen in a previously healthy 36-year-old female. Herein, we report the imaging and pathologic findings and review the literature.

Index terms Angiosarcoma; Breast; Spleen; Ultrasonography; Multidetector Computed Tomography

INTRODUCTION

Angiosarcomas are the most common sarcomas of the breast, but are still extremely rare, accounting for < 0.05% of all breast tumors (1). They are highly aggressive and characterized by extensively infiltrative growth and rapid proliferation (2). They are divided between primary angiosarcoma (PAS) and secondary angiosarcoma (SAS). Of the two, PAS is the greater diagnostic challenge because of its rarity and nonspecific clinical and imaging findings in young population. To our knowledge, little studies of angiosarcoma which were presented as a single mass in each breast and spleen at the time of initial diagnosis have been reported. Herein, we report a rare case of angiosarcoma of the breast which presented as a hyperechoic mass on ultrasonography (US) with a large angiosarcoma of the spleen. A brief review of the relevant literature is also provided.

CASE REPORT

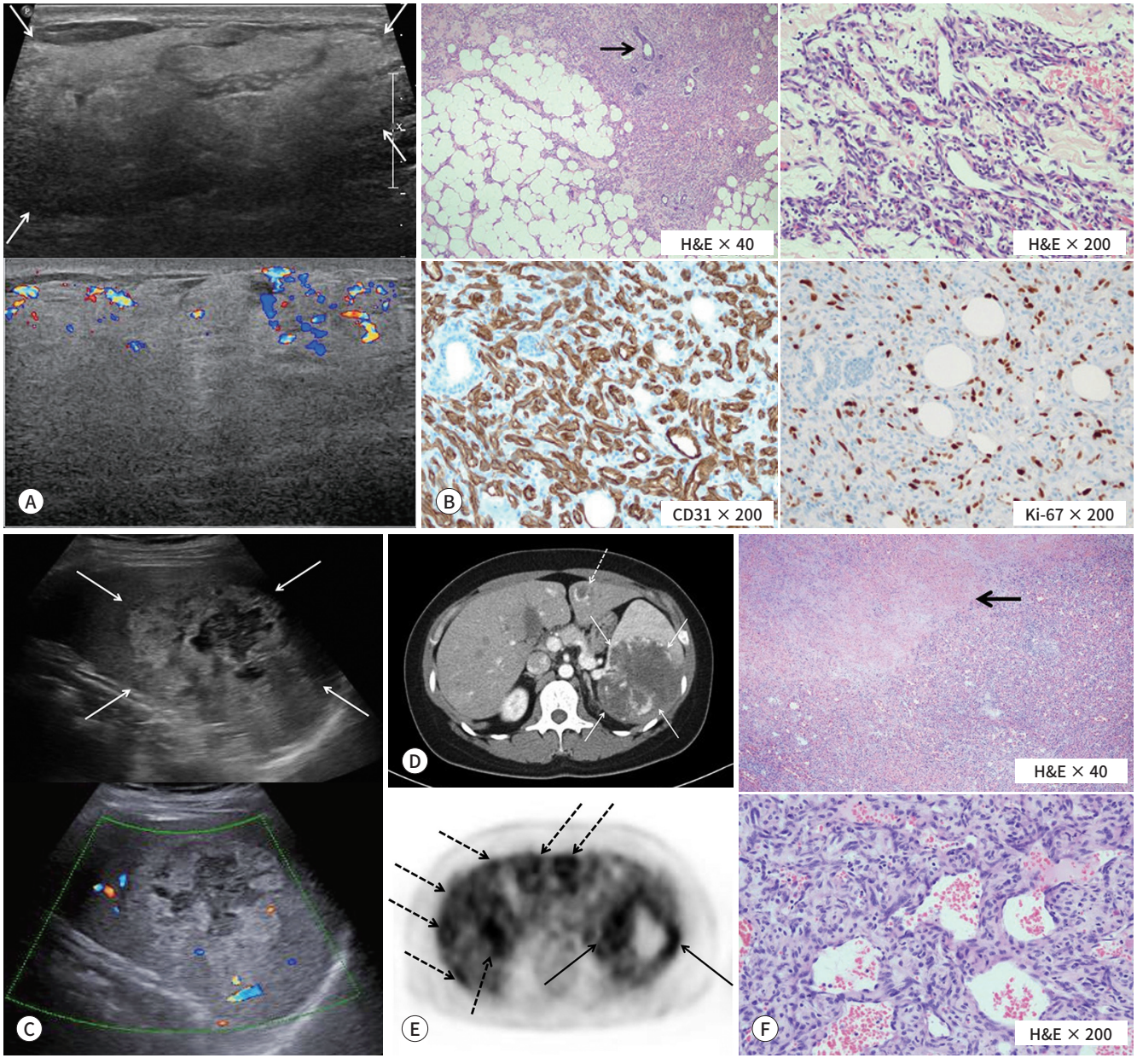
A 36-year-old female presented with a 3-month history of a palpable mass in the left breast. She had no personal or family history of breast cancer, breast surgery or radiation therapy. The mass was biopsied at another center, and hemangioma or angiolipoma was suspected as likely diagnosis. After the needle biopsy, the patient was referred to our hospital for surgery. On physical examination, a 7 cm mass was palpated in the upper outer quadrant of the patient's left breast without any skin change. US showed an irregular mass with indistinct margins and an internal heterogeneous hyperechogenicity. Color Doppler image showed increased internal and rim vascularity of the mass (Fig. 1A). We assessed the mass as category 4A, low suspicion for malignancy according to American College of Radiology Breast Imaging Reporting and Data System. Subsequently, she underwent a wide local excision of the breast mass. On the gross examination a 5.5 cm sized, poorly defined reddish gray lesions within the breast fat tissue was revealed. Microscopically, the tumor was comprised of proliferations of anastomosing vascular channels which were diffusely infiltrating into the adjacent adipose tissue (Fig. 1B). The endothelial cells lining each of the vascular spaces were mildly pleomorphic and sometimes showed multi-layering mitoses were observed around 5 in 10 high power fields. Immunohistochemical stains of CD31 and CD34 endothelial makers expressed diffuse positivity and Ki-67 proliferation index was increased up to 30%. The mass was diagnosed as a spindle cell type angiosarcoma developed in the breast parenchyma. The mass size was 5.5 cm × 4.0 cm and Fédération Nationale des Centres de Lutte Contre Le Cancer (FNCLCC) grade was 1. The surgical margin was focally positive for tumor involvement. Ten days after surgery, she complained of left upper quadrant abdominal pain. Abdominal US showed an oval mass with heterogeneous echogenicity and increased peripheral vascularity in the spleen (Fig. 1C). Multiple hyperechoic masses in the liver were also observed. Contrast-enhanced CT showed an 8 cm low-density mass in the spleen with peripheral heterogeneous enhancement (Fig. 1D). Multiple low-density masses with rim enhancement were also seen in the liver. ^{18}F-fluorodeoxyglucose (FDG)-PET showed multifocal uptakes in the spleen and the liver with standardized uptake values of 4.7 and 5.7, respectively (Fig. 1E). The patient underwent a splenectomy and the mass showed similar histology to breast with extensive necrosis and it was diagnosed as angiosarcoma with a size of 8.0 cm × 7.0 cm and FNCLCC grade 2 (Fig. 1F). She was treated with chemotherapy and was followed up with regular CT studies. Follow-up abdominal and pelvic CT showed aggravated hepatic metastasis. Four months after the initial diagnosis of angiosarcoma of the breast, she died due to hepatic failure.

This study was approved by the Institutional Review Board of our institution (IRB No. 2020-10-001). Informed consent was waived due to the retrospective nature of the study.

DISCUSSION

Breast angiosarcoma is a rare malignancy of endovascular origin and is characterized by aggressive behavior and poor prognosis due to a high local recurrence and incidence of distant metastasis (3, 4). PAS is an extremely rare malignancy of the breast parenchyma; no

Fig. 1. Image findings of angiosarcoma of the breast and spleen in a 36-year-old female.
A. Longitudinal US shows an irregular heterogeneous hyperechogenic mass with indistinct margins in the upper outer quadrant of the left breast (arrows). Color Doppler image shows increased internal and rim vascularity of the mass.
B. Light microscopic findings of the breast mass show a tumoral component diffusely infiltrating the surrounding adipose tissue. Normal breast ducts are entrapped within the tumor (arrow) (H&E, × 40). Proliferating dilated and angulated vascular channels lined by multilayered atypical endothelial cells can also be observed (H&E, × 200). CD31 immunohistochemical staining, an endothelial marker shows a strong membranous expression (CD31, × 200). The proliferation index based on the Ki-67 stain expression is increased (Ki-67, × 200).
C. Abdominal US shows a heterogeneous echoic mass with an oval shape and an indistinct margin of the spleen (arrows). Color Doppler image shows increased peripheral vascularity of the mass.
D. Contrast-enhanced abdominal CT scan shows an 8-cm low-density mass with peripheral heterogeneous enhancement in the spleen (arrows) and a rim-enhanced mass in the liver (dashed arrow).
E. ^{18}F-fluorodeoxyglucose-PET shows multifocal uptakes in the spleen (arrows) and the liver (dashed arrows) with standardized uptake values of 4.7 and 5.7, respectively.
F. Light microscopic findings of the mass in the spleen are indicative of a tumor composed of proliferating vascular spaces with adjacent massive necrosis (arrow) (H&E, × 40). High-power view shows anastomosing dilated vascular channels lined by atypical endothelial cells (H&E, × 200).
H&E = hematoxylin and eosin, US = ultrasonography



known risk factors identified. It generally arises in relatively young female aged 30–50 years (5). By contrast, SAS is much more common, developing in the dermis of the skin and subcutaneous layers, typically after a period of 5–10 years following radiotherapy or chronic lymphedema (Stewart Treves Syndrome). It arises in older female aged primarily between 60–70 years (5). Patients with PAS typically present with a rapidly growing palpable mass, fullness or swelling of the affected breast, and occasionally purplish skin discoloration local to the malignancy (5). Rapidly growing breast lesions in young female such as malignant phyllodes tumor, juvenile fibroadenoma, and metaplastic carcinoma should be considered as differential diagnosis. On the other hand, SAS presents as painless purplish discoloration, hematoma-like swelling, eczematous rash, and diffuse breast swelling which may be difficult to distinguish from post-irradiation change (5, 6). Radiologic diagnosis is difficult because of the nonspecific findings. Mammography may appear normal with or without subtle skin thickening (7). Previously reported findings have included a mass with round, oval or irregular shape and circumscribed or indistinct margins, or focal asymmetry. Coarse calcifications may also be present (3, 7). US may show hypoechoic, hyperechoic or heterogeneous echoic masses, or only parenchymal heterogeneity without mass formation (8). Color Doppler study usually shows hypervascularity (8). On MRI, low signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and heterogeneous enhancement with rapid enhancement and washout kinetics have been reported (8, 9). On ^{18}F-FDG PET, avid FDG uptakes with variable standardized uptake values have been observed (10). Pathologic diagnosis using fine needle aspiration and needle biopsy may often be non-diagnostic; therefore, incisional or excisional biopsies should be considered for conclusive diagnosis (3). Total mastectomy or breast conserving surgery is the preferred initial treatment. The necessity for axillary nodal dissection is unclear because axillary nodal metastasis is not common. (5). Adjuvant therapies including chemotherapy and radiation therapy may be used depending on the locoregional tumor extent and incidence of distant metastasis (5).

Like other sarcomas, the prognosis of angiosarcoma depends on tumor size, histologic grade, and margin status (5). Specifically, low histologic grades (i.e., bland, well-formed vascular channels which invade the breast parenchyma) have a better survival rate than high grades (i.e., sarcomatous changes with areas of necrosis, hemorrhage and infarction). As such, 5-year survival rates of 76% for low grade, 70% for intermediate grade (i.e., solid neoplastic vascular growth with an increased mitotic rate), and 15% for high grade angiosarcoma have been reported (1, 7). Distant metastasis have been thought to be mainly hematogeneous and reported in the lung, skin, liver, spleen, bone, central nervous system, ovary, lymph nodes, omentum, adrenal gland, heart, gastointestinal tract, tonsil, oropharynx, and even the psoas muscle (3, 5).

In summary, angiosarcoma of breast is rare malignant tumor that can be often misdiagnosed radiologically owing to the nonspecific features. In the present paper, we describe a rare case of angiosarcoma of the breast and spleen in a young patient and report the findings of US, CT, and ^{18}F-FDG PET. Although hyperechoic masses on the US are usually benign, and diagnosis should be made by considering clinical and other imaging findings.

Author Contributions

Conceptualization, J.H.K.; investigation, K.J.; supervision, J.H.K.; visualization, K.W.; writing—original draft, K.J.; and writing—review & editing, J.H.K., K.W.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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유방 및 비장의 혈관육종: 영상 소견 및 병리 소견을 포함한 증례 보고

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유방의 혈관육종은 매우 드문 악성 육종으로, 영상 소견이 비특이적이며 지금까지의 보고가 제한적이다. 또한 진단 당시 유방 및 비장 각각에서 하나의 큰 병변으로 보인 혈관육종은 거의 보고되지 않았다. 이에 저자들은 과거력이 없는 건강한 젊은 여성의 유방 및 비장에서 함께 관찰된 혈관육종 증례를 영상 및 병리학적 소견과 함께 보고하고자 한다.

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