Bilateral Primary Breast Angiosarcoma: A Case Report

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ABSTRACT

Background: Angiosarcoma can develop in all parts of the body containing blood vessels, including breast. Statistically, less than 10% of all angiosarcomas originate in the breast. Angiosarcoma accounts for less than 0.05% of breast primary cancers. Primary angiosarcoma develops without a history of treatment for breast cancer, whereas secondary angiosarcoma develops in patients who have already had treatments for other primary breast cancer.

Case presentation: In review of the literature, primary angiosarcoma, particularly bilateral, is rare. In this study, we present a patient, a young woman, with primary bilateral angiosarcoma.

Conclusion: Although breast angiosarcoma is rare, we should be aware of it, particularly in young women with breast mass that is hyperflow in color Doppler ultrasound.

Introduction

Malignant breast tumors that arise from stromal tissues are very rare, and angiosarcoma is a rare malignant tumor that arises from endothelial cells of vascular channels. Despite its rare occurrence, the breast is one of the most common sites in the body to develop angiosarcoma. Primary angiosarcoma of breast arises in patients who have never had history of treatment for breast cancer.

Secondary angiosarcoma occurs most frequently after breast conservation therapy with radiation therapy; the average latency is 5–10 years. Primary breast angiosarcomas, consisting of approximately 20% primary breast sarcomas, occur most commonly in women 20–40 years old. The incidence of primary breast angiosarcoma is approximately 17 new cases per million women. Secondary angiosarcomas usually occur in older women a few years after the treatment of breast cancer. Secondary angiosarcomas can arise on the chest wall following mastectomy or in the breast following breast conservation therapy and irradiation. Our patient was a young woman with metachronous bilateral breast angiosarcoma with no previous history of radiation.

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Case Report

A 26-year-old woman referred to us in the radiology department for sonographic evaluation of a new palpable lesion in the lateral part of her left breast. She had no positive family history of breast cancer. Last year, she noticed a red patch on her right breast during her pregnancy, which was diagnosed by her gynecologist as a dermatologic disease to be resolved after delivery. As the patch grew continuously, an excisional biopsy was made simultaneously during caesarian section, which proved to be low-grade angiosarcoma. Since she had no history of breast surgery or radiation, it was judged as primary breast angiosarcoma and modified radical mastectomy was pursued later. The patient was given adjuvant chemotherapy with anthracycline/ifosfamide-based regimen.

After 11 months, when she presented back to us, she felt a new palpable mass in the lateral part of her left breast without pain, nipple discharge, axillary swelling, and skin abnormality.

Using ultrasound, at the area of patient's complaint there was an isoechoic area 38*20*18mm without well-defined borders in the left breast at 3 o'clock location, which was significantly hyperflow using simultaneous color Doppler (Figure 1,2).

No apparent pathologic axillary lymph node was found by ultrasound. After consulting with the surgeon, core needle biopsy was performed. Samples were reviewed with an experienced pathologist, and again, low-grade angiosarcoma was reported.

Sections showed neoplastic tissue composed of interanastomosing vascular channels, some filled by RBC. The endothelial cells had hyperchromatic nuclei that bulged into lumen, or would lie along the wall of the vessel as thin elongated hyperchromatic threads. The vascular spaces are round to ovoid, or elongated with branches that anastomose with similar channels. Entrapment of fat, ducts, and lobules by neoplastic cells was noted. Mitosis and necrotic areas were absent. In Immunohistochemical studies, CK was negative but CD31 and CD34 were positive, and Ki67 was positive in 10% of tumoral cells on average (Figure 3).

We suspected of either bilateral primary breast angiosarcoma or metastatic angiosarcoma from contralateral breast. All her other metastatic work-ups including thoracic and abdominal CT scans were negative, and she was referred for left breast surgery. The patient underwent modified radical mastectomy of left breast and now this patient is under close follow-up.
Figure 3. Neoplastic tissue composed of interanastomosing vascular channels some filled with by RBC. The endothelial cells has hyperchromatic nuclei that bulge into lumen or lie along the wall of the vessel as thin elongated hyperchromatic threads.

Discussion

Breast sarcomas are a heterogeneous group of malignant neoplasms that arise from the mammary stroma. Angiosarcoma is one of the most common forms of breast sarcoma, which are developed from the endothelial cells of the blood vessels. Primary angiosarcomas are rare and account for 0.05% of all malignant breast tumors. Breast angiosarcoma can be observed as a primary neoplasm or as secondary following radiotherapy for breast carcinoma. Both primary and secondary breast angiosarcomas carry a prognosis worse than primary breast carcinoma. In the present paper, only primary angiosarcomas will be discussed.

Three grades of angiosarcoma have been described. Low-grade tumors have anastomosing vascular channels that invade the surrounding breast tissue. Intermediate-grade tumors have a more solid pattern of vascular growth and an increased mitotic rate. High-grade lesions have areas of hemorrhage and necrosis. Multiple grades may coexist in the same tumor, so grading of a tumor in core biopsy specimens may not be possible, and complete excision is needed to accurately determine the tumor grade.

Primary angiosarcoma arises in younger women, usually between 2–4 decades of life, unlike breast carcinomas that typically arise later in life. Between 6 and 12% of primary breast angiosarcomas are diagnosed during pregnancy or shortly after, like this patient whose right breast angiosarcoma was diagnosed in her late pregnancy, which can suggest a role of hormones. However, cases with reported positive estrogen receptors are so rare that a link between angiosarcomas and hormonal dependency is still not established. In our patient, IHC analysis only had CK, CD31, CD 34, Ki 67 and CD 31 without information about estrogen receptors.

The clinical presentation of primary angiosarcoma could be a palpable mass that may be growing rapidly, and bluish skin discoloration occurs in up to a third of patients and is thought to be attributable to the vascular nature of the tumor. In the series by Yang et al., the mean tumor size of the mass at presentation was 5.9 cm.

In imaging, breast angiosarcomas have no pathognomonic signs. Mammography is usually nonspecific. An ill-defined, non-calcified mass or focal asymmetry is the most common finding. Calcifications can also be seen.

It has been suggested that the echotexture of these lesions is highly variable and patients with higher-grade lesions at pathology are more likely to have abnormal mammographies.

Many women with primary breast angiosarcoma are young, and have dense breast parenchyma that is characteristic of their age, which may obscure visualization of a mass. Lieberman et al. reported 33% false negative in mammographies of angiosarcomas. Yang et al. reported 19% false negative reports in mammographic. The presence of fat in mammographic abnormalities has been reported, which suggests the differential diagnosis of hemangioma and angiolipoma. Sonography is very useful for evaluation of a lesion when a palpable abnormality is found. Masses may be circumscribed or ill-defined. Yang et al. reported mixed hyperechogenic and hypoechogenic regions without a discrete mass in 38% of patients in their series. Color Doppler sonography showed hypervascularity.

Differential diagnosis of this rare tumor includes phyllodes sarcoma, stromal sarcoma, benign hemangioma, squamous cell carcinoma with sarcomatoid features, and metaplastic carcinoma.
Immunohistochemical stains for endothelial markers (CD34 and CD31), epithelial markers (pancytokeratin), and other sarcoma markers help in making the correct diagnosis. Wide surgical resection is the mainstay of treatment for both forms of angiosarcoma. For small, grade I primary lesions, breast conservation therapy may be indicated. Chemotherapy may reduce the local recurrence rate. Patients with primary angiosarcoma of breast receiving Paclitaxel has been shown to have good responses in a number of studies.9, 13

The prognosis of angiosarcoma is thought to depend on the histologic grade and tumor size. Patients with a higher-grade lesion are more prone to develop a recurrence and have a lower survival rate than are those with a lower-grade lesion.9,14-17

In conclusion, solid-appearing breast mass in young women that are highly vascular should be considered malignant until proven otherwise. Breast angiosarcoma does not show skin discoloration in all cases. It is very useful to use color Doppler in sonographic evaluation of breast solid mass. Diagnosis of this entity is needed for appropriate early therapy.

References