Primary angiosarcoma of the breast

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CASE REPORT

An 18-year-old female presented with lump and pain of left breast for 6 months. There was a firm irregular mass of 10 × 8 cm in the center of the left breast with prominent cutaneous vasculature and multiple small nodules (Fig 1). There was ulceration of the areola and destruction of the nipple, but the axillae and right breast were normal.

Fine-needle aspiration cytology revealed only hemorrhagic material. Color Doppler ultrasonography demonstrated dilated vascular channels with minimal active flow, suggestive of cavernous hemangioma. Magnetic resonance imaging showed a resolving hematoma. The patient underwent lumpectomy amid diagnostic uncertainty.

Gross examination showed multiple hemorrhagic areas and 2 cystic areas of 2.5 cm and 1.5 cm filled with blood clots. Vascular channels were dilated and thrombosed with interspersed necrotic areas, mainly in the central quadrant. The vascular channels were lined by flattened or plump endothelial cells (Fig 2). Most of the endothelial cells were distributed in a single layer, but in some locations the cells formed papillae projecting into the lumen. Endothelial tufting was present occasionally. Immunohistochemistry for factor VIII was positive in the endothelial cells. The final histopathologic diagnosis was angiosarcoma of the breast. As this neoplasm often extends beyond its gross limits and simple excision is associated with a high rate of local recurrence, simple mastectomy was subsequently performed.

DISCUSSION

Primary angiosarcomas of the breast are extremely rare (0.05% of all breast cancers),1 occurring during the third and fourth decade of life and presenting as an ill-defined breast mass. Different
nomenclature used for angiosarcoma are heman-gioendothelioma, hemangioblastoma, hemangio-sarcoma, and metastasizing angioma. Metastases from breast angiosarcoma have been reported in lungs, skin, liver, bone, central nervous system, spleen, ovary, lymph nodes, and heart.²

Accurate preoperative diagnosis may be difficult, even with the aid of clinical examination, ultrasoundography, fine-needle aspiration cytology, and biopsy.³ Chen et al reported a 37% false-negative rate of biopsy. The differential diagnosis of this rare malignancy includes cystosarcoma phylloides, stromal sarcoma, metaplastic carcinoma, squamous cell carcinoma with sarcomatoid features, myoepithelioma, fibromatosis, fibrosarcoma, liposarcoma, and reactive spindle-cell proliferative lesions.² Ultrastructural examination of tumor cells may show intermediate filaments; for example, vimentin, cytokeratin. Weibel-Palade bodies, and pinocytic vesicles are associated in some cases. Immunostaining for factor VIII–related antigen is confirmatory for angiosarcoma of the breast.⁴

The treatment of angiosarcoma of the breast is early and complete surgical excision of the mass by simple mastectomy. Radical mastectomy is not advocated unless the deep fascia or pectoral muscles are involved. Axillary dissection is not indicated because nodal involvement is rare.⁵ The role of breast conservation therapy is limited because of the aggressive nature of the tumor and its detection usually occurs when the size is >5 cm. Extensive ulceration or chest wall involvement requires the strategy of chemotherapy (doxorubicin based) and radiotherapy followed by surgery. Adjuvant therapy is considered for multifocal lesions, locally invasive lesions, regional nodes, or skin involvement. Specific antibodies conjugated with an efficient cytotoxin (eg, ricin) or virus (eg, parvovirus) could be used to target the tumor vessel.⁶ Angiosarcoma has a high mortality rate with only 10% to 21% of patients being disease-free after 5 years.²

REFERENCES

Fig 2. Photomicrograph of angiosarcoma of the breast. (Hematoxylin-eosin stain; original magnification: ×50.)