Answer

Angiosarcoma of the Breast

At the time of mastectomy, a cutaneous nodule emerged (Figure 1). The magnetic resonance image shows enhancement of diffuse skin thickening with evidence of extension into the subareolar tissue (Figure 2). A total mastectomy with axillary lymph node dissection was performed (the latter was based on the erroneous preoperative diagnosis of ductal carcinoma). The final pathological results revealed poorly differentiated angiosarcoma both medial to the nipple and in the lower outer quadrant.

Angiosarcoma of the breast that arises in the absence of known risk factors, termed primary angiosarcoma, represents less than 1% of all primary breast malignancies and 3% to 10% of all breast sarcomas. In general, the disease is considered to be rare. It is more prevalent in younger populations and is most often found in women during their third to fourth decade of life. Secondary angiosarcoma of the breast is more common and is associated with known risk factors including extremity edema and chronic lymphedema. Angiosarcoma after a breast-conserving procedure is increasingly diagnosed in a small but significant portion of breast carcinoma survivors. Recently, multiple studies have established an association between radiation therapy and breast angiosarcoma. Cozen et al reviewed 264,444 cases of women in Los Angeles County, California, with previous invasive ductal carcinoma and found a significantly increased risk of angiosarcoma of the upper extremity (odds ratio = 59) and of the breast or chest wall (odds ratio = 11.6). This increased risk is thought to be related to persistent lymphedema or the radiation therapy itself. In a review of 194,798 cases of breast cancer from the Surveillance, Epidemiology, and End Results Program data, patients who had undergone breast conservation and radiotherapy had a relative risk of 15.9 of developing angiosarcoma. Lesions tend to appear 4 to 7 years after therapy and may be related to persistent lymphedema.

Definitive diagnosis of angiosarcoma of the breast can be difficult. Mammography may show a nonspecific mass or thickening of the skin that may be confused with a benign lesion. In fact, approximately one third of patients with breast angiosarcoma have no abnormalities on their mammograms. Fine-needle aspiration may also have a low diagnostic yield because of limited specificity. In this case, the fine-needle aspiration revealed only nonspecific tumor cells. As a result of this diagnostic difficulty, magnetic resonance imaging is becoming an increasingly popular diagnostic tool. Magnetic resonance imaging often reveals low signal intensity on T1-weighted images but higher signal intensity on T2-weighted images. With the use of gadolinium, images of lesions are enhanced on low-signal T1-weighting and high-signal T2 weighting.

Given the increasing use of radiation and breast-conserving therapy, physicians may see a rise in the incidence of secondary angiosarcoma of the breast. Furthermore, magnetic resonance imaging will likely prove useful in confirming suspicious lesions owing to the low yield of traditional imaging and the increasing incidence of angiosarcoma in these patients. However, the final diagnosis and treatment should be based on definitive pathological results. The recommended treatment for both primary and secondary angiosarcoma is simple mastectomy with an increasing role for adjuvant and neoadjuvant therapy. High-grade angiosarcoma shows 5-year survival rates approaching 10% to 21%.

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Correspondence: Stanley P. L. Leong, MD, Department of Surgery, University of California, San Francisco, Comprehensive Cancer Center at Mount Zion, 1600 Divisadero St, San Francisco, CA 94143-1674 (leongs@surgery.ucsf.edu).

REFERENCES


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