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PRIMARY BREAST ANGIOSARCOMA: A CASE REPORT

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ABSTRACT

Less than 1% of all breast cancers are classified as primary mammary angiosarcomas, which are highly uncommon and aggressive tumors arising from the inner layer of blood and lymph vessels. These tumors typically have a poor prognosis and exhibit a notable tendency for metastatic spread.

The present report outlines a case involving a 57-year-old woman who discovered a concerning mass in her left breast during a routine check-up. Initially, she underwent a lumpectomy, followed by a prophylactic mastectomy a year later. The mass was identified as a well-differentiated primary breast angiosarcoma. The patient is presently under careful observation and may need further treatment options, which could include reconstructive surgery and adjuvant therapy.

This case highlights the importance of prompt intervention and early detection in enhancing the outcomes for patients with primary breast angiosarcomas. Despite their rarity, swift and comprehensive treatment is essential to reduce the risk of metastasis and relapse.

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INTRODUCTION

Angiosarcomas are uncommon tumors of mesenchymal origin that arise from the inner surface of blood and lymphatic vessels, exhibiting significant metastatic and aggressive behavior (other terms include haemangiosarcoma and lymphangiosarcoma) (1). These tumors can develop in any organ, but they are predominantly found in the skin and soft tissues. Notably, primary angiosarcoma of the breast is a malignant endothelial tumor originating in mammary tissue, typically unrelated to radiation exposure. These cases are exceedingly rare, accounting for less than 1% of all breast cancer diagnoses (2). Angiosarcomas can be categorized into primary and secondary types, with secondary angiosarcomas being the most commonly seen. Secondary angiosarcomas, develop as a complication of radiation therapy after breast conservation or from chronic lymphedema. Primary angiosarcomas of the breast account for just 0.04% of malignant breast tumors (3). The exact pathophysiological mechanisms behind primary angiosarcomas remain largely unclear (4). Nevertheless, mutations in PLCG1 and KDR, which are part of the VEGFR2 signaling pathway, have been noted. Additionally, a few exceptionally rare instances suggest amplification of the MYC gene. In contrast, secondary angiosarcomas in the breast are generally linked to prior radiation exposure in the chest region and its vicinity.

CASE PRESENTATION

A 57-year-old woman presents with a medical background of hypothyroidism and a previous cesarean section. She reports no allergies, does not smoke, and does not consume alcohol. She has been vaccinated against COVID-19 with three doses and has experienced two infections.

The patient was scheduled for admission to the hospital on 24th November 2023 for surgical treatment of a left breast mass, identified one month prior during a routine check-up. An MRI conducted on 30th October 2023 revealed a sizable mass measuring 7 x 6 x 5.2 cm in the upper central area of the left breast, showing atypical histological features categorized as BIRADS 4, warranting additional histological evaluation. Histological analysis of breast gland tissue samples taken on 18th October 2023 revealed primary atrophy of the duct-lobular units and moderate stromal fibrosis accompanied by a significant chronic inflammatory response. No signs of malignancy were found in the analyzed specimens.

The patient underwent a lumpectomy on the left breast at the 12th hour. The procedure was conducted with the patient in the supine position and under general anesthesia. The patient was discharged on 28/11/2023 with detailed instructions and a pain management plan. A biopsy performed on 27/11/2023 indicated a well-differentiated primary breast angiosarcoma. Due to this biopsy finding, the patient was admitted to the clinic on 29/2/2024 for a planned prophylactic mastectomy of the left breast. On 1/3/2024, the patient had a radical left breast mastectomy via PATTEY 1. The surgery was done with the patient in the supine position, the left arm in abduction and external rotation, under general anesthesia. The surgical specimen was sent for further biopsy analysis. The patient was released on 3/3/2024 with REDOVAC and a pain management plan in place, pending the biopsy results. Future reconstructive surgery may be necessary for the patient.

DISCUSSION

Patients diagnosed with breast angiosarcomas typically have a median age of 40 years, in contrast to the 70-year median age seen in secondary angiosarcoma (3). Most individuals exhibit a rapidly enlarging mass which is sometimes accompanied by purplish-blue skin discoloration or breast asymmetry (5). On mammography, it generally appears as a non-calcified mass or asymmetry (1). Ultrasonography may show either a hyperechoic mass or a combination of hyper- and hypoechogenic areas accompanied by architectural distortion. Dynamic contrast-enhanced MRI displays characteristic malignant enhancement patterns. Fine-needle aspiration (FNA) has a false negative rate of 40%, which establishes core needle biopsy as the preferred diagnostic method. Approximately 20% of patients are found to have regional disease at diagnosis. The most common metastatic sites include the lungs, liver, bones, and central nervous system (CNS).

The tumors present as hemorrhagic, diffuse, or multinodular formations, ranging from 0.7 to 25 cm in size (average: 6.7 cm, median: 5 cm). Typically, they are found in parenchymal tissue rather than on the skin (6). Their margins can often be indistinct. Tumors that are better differentiated exhibit a spongy, hemorrhagic aspect, while poorly differentiated tumors appear more solid, characterized by dense grayish-white tissue interspersed with necrotic areas. These less differentiated tumors may also contain regions of cystic, spongy, hemorrhagic vascular tissue in their vicinity. Angiosarcomas are generally found deep within the breast tissue and may or may not involve the skin, exhibiting infiltrative or indistinct borders. A wide morphological spectrum is encountered, generally with a predominant vasoformative or solid component identified. Unlike secondary angiosarcomas, primary variants more frequently display well-defined, small to medium anastomosing vessels that penetrate fibroadipose tissue. Well-differentiated angiosarcomas consist of well-structured vessels lined by flattened endothelial cells showing little atypical features (7). The presence of irregularly shaped vascular channels dissecting through adipose tissue and mammary lobules suggests a malignant nature. A lobular-type growth pattern, typically seen in haemangioma, is not present. Lesions with an intermediate appearance show endothelial multilayering, hobnailing, or papillary-like projections. Poorly differentiated angiosarcomas exhibit solid, cellular regions consisting of sheets of spindled to epithelioid cells interspersed with variably developed anastomosing vascular channels, accompanied by notable blood lakes, mitotic activity, and areas of necrosis (8). Epithelioid angiosarcomas present a solid structure, characterized by sheets of large atypical epithelioid to polygonal cells that contain ovoid vesicular nuclei, prominent nucleoli, and relatively ample cytoplasm. Due to potentially limited vasoformation, epithelioid angiosarcoma can be mistaken for carcinoma. Frequent mitotic figures and necrosis are typically observed. Angiosarcomas express endothelial markers, with strong, membranous CD31 staining and nuclear ERG immunoexpression noted. Aberrant expression of KIT, synaptophysin, chromogranin, and CD30 can be seen. Most tumours lack expression of MYC protein. Staging holds no clinical significance.

DISCUSSION

The primary treatment for angiosarcoma is total mastectomy, with or without radiation therapy (9,10). Nevertheless, local recurrences are noted in about 50% of patients. The choice of local treatment largely depends on the ratio of tumor size to breast size (11). If the tumor is small and well-defined, it may be possible to excise it surgically with clear margins and opt for cosmetic reconstruction, making breast-conserving surgery a viable option. For patients with larger tumors, mastectomy is necessary. Axillary lymph node dissection is typically not performed unless there is pathological evidence of nodal involvement, given the low rates of nodal metastasis. The role of radiation following resection is not clearly defined (4). Various studies often combine primary breast angiosarcomas with secondary (radiation-associated) angiosarcomas, employing differing approaches of radiation therapy and systemic treatment in either the adjuvant or neoadjuvant setting (12). The primary chemotherapy treatment for breast angiosarcomas includes regimens based on taxanes and anthracyclines. In patients with locally advanced or metastatic angiosarcoma, the rate of complete or partial response was 25% (compared to 21% for those with other types of sarcomas). Enhanced progression-free and overall survival rates were observed with the combination of doxorubicin and ifosfamide, as opposed to using anthracycline chemotherapy alone.

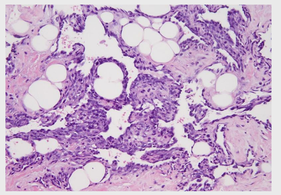

Figure 1: Primary breast angiosarcoma

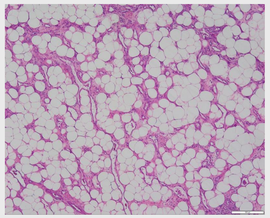

Figure 2: Primary breast angiosarcoma

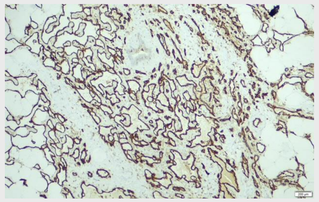

Figure 3: Primary breast angiosarcoma

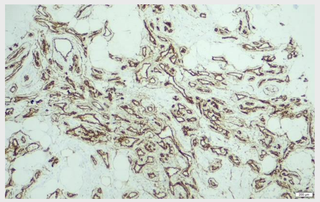

Figure 4: Primary breast angiosarcoma

CONCLUSIONS

Primary mammary angiosarcomas are exceptionally rare malignant tumors known for their poor prognosis and high likelihood of metastasis. Although they represent only a tiny fraction of breast cancer cases, they present substantial difficulties in both diagnosis and treatment. Total mastectomy remains the primary approach for treatment due to their aggressive nature and tendency for early spread. However, even with surgical intervention, there is still a notable risk of recurrence, and adjunct therapies like chemotherapy and radiation often yield limited success. This underscores the importance of diligent post-operative monitoring and the need for advancements in treatment methods. Management of this rare but aggressive tumor type requires meticulous treatment planning right from the outset.

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