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Case Report

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Breast Angiosarcoma: Diagnostic Challenges and Management of a Rare Malignancy

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Abstract

Background: Primary angiosarcoma of the breast is an uncommon and aggressive vascular malignancy, representing less than 0.05% of all breast cancers. It often lacks distinct radiological features, which can delay diagnosis.

Case Summary: We present the case of a 67-year-old woman with no prior radiotherapy who developed a rapidly enlarging right breast mass. She underwent total mastectomy followed by adjuvant chemo-radiotherapy.

Conclusion: Due to its rarity and variable presentation, primary breast angiosarcoma poses diagnostic and therapeutic challenges. A multidisciplinary approach is essential for optimal patient outcomes.

Abbreviations: PBA: Primary Breast Angiosarcoma

Introduction

Primary breast sarcomas are rare malignant mesenchymal tumors with heterogeneous histology, including fibrosarcoma, angiosarcoma, malignant phyllodes tumor, and malignant fibrous histiocytoma **[1]**. Among these, primary breast angiosarcoma (PBA) accounts for roughly 1% of all breasts soft-tissue malignancies **[2]**.

PBA may arise de novo—more often in younger women—or occur secondary to radiotherapy for epithelial breast cancer after a latency of about a decade **[3-5]**. First described by Borrman in 1907 **[6]**, PBA is characterized by high vascularity, rapid growth, and poor prognosis. Histopathologically, three patterns are recognized.

- type I (well-formed vascular channels with minimal endothelial proliferation)
- type II (papillary endothelial structures)
- type III (solid endothelial proliferation with necrosis and hemorrhage) **[7]**. Because of its rarity, most publications are small series or case reports, and standardized treatment guidelines are lacking.

Case Report

A 67-year-old woman with a history of osteoporosis complicated by femoral neck fracture presented with progressive swelling of the right breast over 8 months, accompanied by mastodynia and skin changes.

Clinical Examination:

- A 10 cm soft, compressible, and tender mass involving the entire right breast.
- Overlying skin showed a purplish angiomatous

discoloration.

- No nipple discharge or palpable axillary nodes.
- Contralateral breast was normal.

Mammography demonstrated a large pseudo-nodular opacity of water-like density in the upper outer quadrant without microcalcifications. Ultrasonography showed diffuse infiltration and thickening of the subcutaneous fat with marked hypervascularity on Doppler; no discrete cystic or solid mass was identified. Core needle biopsy established a diagnosis of high-grade (type III) angiosarcoma. The patient underwent total mastectomy, followed by adjuvant chemotherapy and radiotherapy. Postoperative recovery was uneventful.

Discussion

Epidemiology & Presentation

PBA is an aggressive malignancy whose presentation and imaging are often non-specific, contributing to delayed diagnosis **[8]**. Mammography and ultrasound can appear benign, particularly in younger patients, while MRI more reliably suggests malignancy by demonstrating high T2 signal and rapid contrast uptake with subsequent washout **[2,8,9]**.

Diagnosis

Definitive diagnosis requires tissue sampling—preferably core needle biopsy—and immunohistochemistry, with typical positivity for endothelial markers such as CD31, factor VIII, and FLI1 **[9-12]**.

Surgery

Surgery remains the cornerstone of management. Modified radical mastectomy achieves the best local control, whereas axillary lymph node dissection is not routinely indicated because nodal spread is uncommon; it may be performed when needed to obtain clear margins **[8,9,13,14]**.

Chemotherapy

Given the high systemic relapse risk of soft-tissue sarcomas **[15]**, adjuvant chemotherapy is frequently considered. Meta-analyses and randomized data support anthracycline-based regimens (e.g., doxorubicin ± ifosfamide) for improved disease-free and overall survival in selected patients **[16-18]**, while taxane-based therapy has shown activity in metastatic angiosarcoma **[19]**.

Radiotherapy

Adjuvant radiotherapy can be considered for close/positive margins or high-grade disease, although its survival benefit remains uncertain; series report mixed results regarding local control and disease-free survival **[8,20-22]**.

Prognostic Factors

Prognosis is influenced by histologic grade, resection margin status, and tumor size **[1,9,23-25]**. In a large single-institution series, 5-year disease-free survival ranged from approximately 76% for low-grade tumors to about 15% for high-grade lesions **[8]**.

Conclusion

Primary breast angiosarcoma is an uncommon, highly vascular malignancy with limited evidence to guide care. Early recognition, radical surgical excision with negative margins, and multidisciplinary planning are key to optimizing outcomes. Further multi-institutional studies are needed to define the roles of adjuvant chemotherapy and radiotherapy **[1,8,9,20]**.

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