Case Report

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An Elusive Diagnosis: Primary Breast Angiosarcoma with Metastatic Spread-A Case Report and Literature Review

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Abstract

Breast angiosarcoma is an extremely rare disease that is more aggressive than mammary carcinomas. A 38-year-old woman was admitted to the hospital with breast edema. Mastitis was suspected, and biopsies were done, but no cancer was found. Her follow-up revealed a lesion with axillary involvement. She underwent neoadjuvant chemotherapy. Her mastectomy was scheduled when lung metastases were found during treatment. The pathology report indicated high-grade angiosarcoma. She underwent adjuvant chemotherapy. Brain metastases were identified, and she died during her follow-up. Radiological imaging may potentially miss it or misidentify it as a benign lesion. The literature has identified several treatment approaches, but due to the small number of patients, no specific guidelines exist. This case demonstrated that the disease is challenging to diagnose, and that magnetic resonance imaging can serve as an effective diagnostic instrument.

Keywords: Breast cancer, breast, primary breast angiosarcoma, metastasis

Introduction

Angiosarcomas, which are uncommon breast tumors of soft tissue, can manifest in both primary and secondary forms. The primary form lacks a known precursor, while the secondary form records radiation exposure to the breast tissue. Primary mammary angiosarcomas are aggressive tumors with a high propensity for local recurrence and distant metastasis, as well as a high risk of mortality due to the tumor (1,2).

This article presents a 38-year-old patient with metastatic high-grade angiosarcoma in the right breast. She underwent a mastectomy and had pulmonary metastases prior to and after the procedure. She later developed brain metastases and lesions on her tongue, identified as lobular capillary hemangiomas. She died of her disease.

Case Report

Our case represents a patient with metastatic primary breast angiosarcoma (BA) and all necessary informed consents were obtained from the patient prior to the operation. In 2019, a 38-year-old female patient presented with right breast edema and pain at a rural hospital. The ultrasound of the breast indicated a nodule and mastitis. A trucut biopsy reveals no cancer in this tumor. A 5 cm lesion and purulent discharge on her right breast led to her hospitalization one month later. The prior biopsy raised suspicion of granulomatous mastitis; however, no granuloma was detected, and malignancy could not be excluded.

She received a mammography as part of her routine national breast screening program follow-ups, which identified a BIRADS-4 lesion. Following this, she

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***This case was presented as a poster presentation in the 23rd Turkish National Surgical Congress at Antalya (April 2024) and the abstract was added to the congress book.**

conducted a biopsy on the lesion, leading to the diagnosis of an angiosarcoma. The positron emission tomography/computed tomography (CT) scan identified a 70x63 mm tumor in her right breast with increased fluorodeoxyglucose (FDG) uptake (SUV_{max}=8.2), along with right axillary lymph nodes at level 1, the largest of which measured 14 mm in diameter and showed increased FDG uptake (SUV_{max}=3).

She attended our hospital's oncology clinic and received paclitaxel as neoadjuvant chemotherapy. A CT scan of the chest and abdomen was performed following three cycles of therapy. Several metastatic lesions in her lungs, the largest measuring 8 mm were identified.

Immediately after the discovery of her lung metastases, the oncology clinical meeting reviewed the case and scheduled a mastectomy. The final pathology indicated a high-grade angiosarcoma with significant vascular cavity invasion. Eight lymph nodes were identified as reactive. The immunohistochemistry examination demonstrated widespread endothelial staining with CD31, CD34, and factor 8. Mammoglobin, GCDFP15, and HHV8 are negative. Her Ki-67 index is 70-75%. The lesion measures 7x5x3.5 cm (American Joint Commitee on Cancer-pT3N0M1). The tumor displays significant hemorrhagic regions, fibrin accumulation, and necrosis, alongside endothelial arrangement around the vascular space, solid cellular proliferation, endothelial cell budding, papillary formations, mild atypia, nucleoli, and mitotic activity. The vascular space is extensively invaded. No tumor is present in the sections of breast skin, nipple, or areola. The tumor extends 0.2 cm from the operative margin of the fascia (Figures 1-4).

On postoperative day 13, CT scans of the thorax demonstrated numerous nodules in both lungs, some exhibiting contrast enhancement. The patient is then assigned an *ifosfamide, mesna, adriamycin*-IMA chemotherapy regimen for palliative treatment. A control thoracic CT was conducted on her after three months and three cycles. Her CT images demonstrated several metastatic nodules, the largest measuring 6 millimeters in diameter. The quantity, dimensions, and density of the nodules markedly diminished in comparison to the patient's prior thoracic CT scan.

Following the conclusion of the fourth cycle, an magnetic resonance imaging (MRI) of her brain indicated no signs of metastases. She then finished three more cycles, resulting in a total of six therapy cycles. After completing her chemotherapy program, she developed brain metastases and lesions on her tongue, later diagnosed as lobular capillary hemangiomas. She subsequently died from her illness.

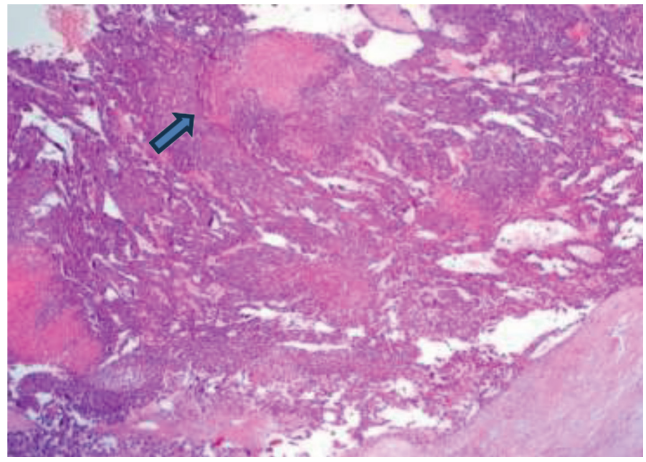

Figure 1. The tumor consists of papillary structures and vascular formations, with areas of necrosis observed (x40)

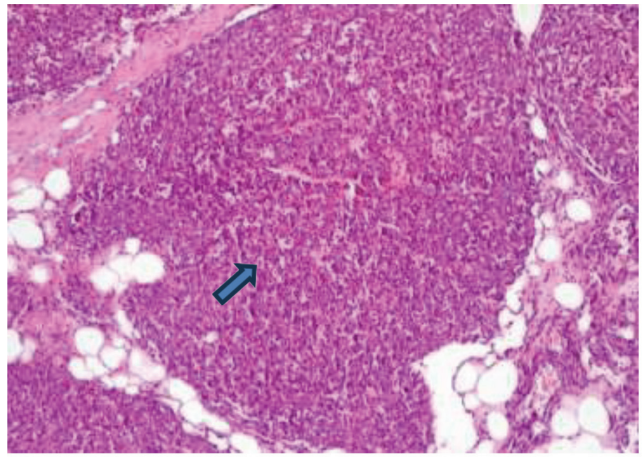

Figure 2. Solid areas with spindle cell morphology and limited vascular structures are observed (x100)

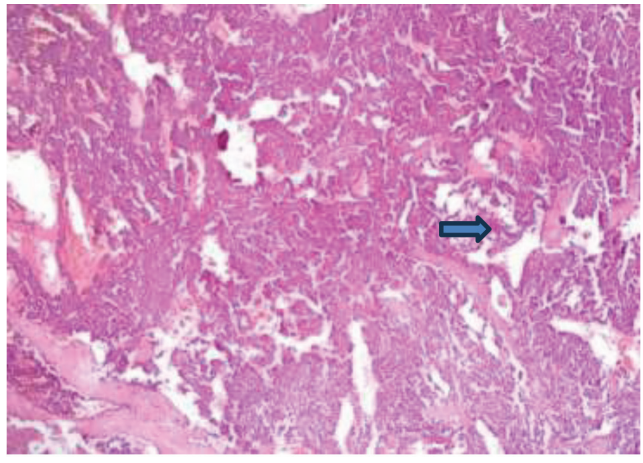

Figure 3. Extensive hemorrhagic areas are present between the papillary formations (x40)

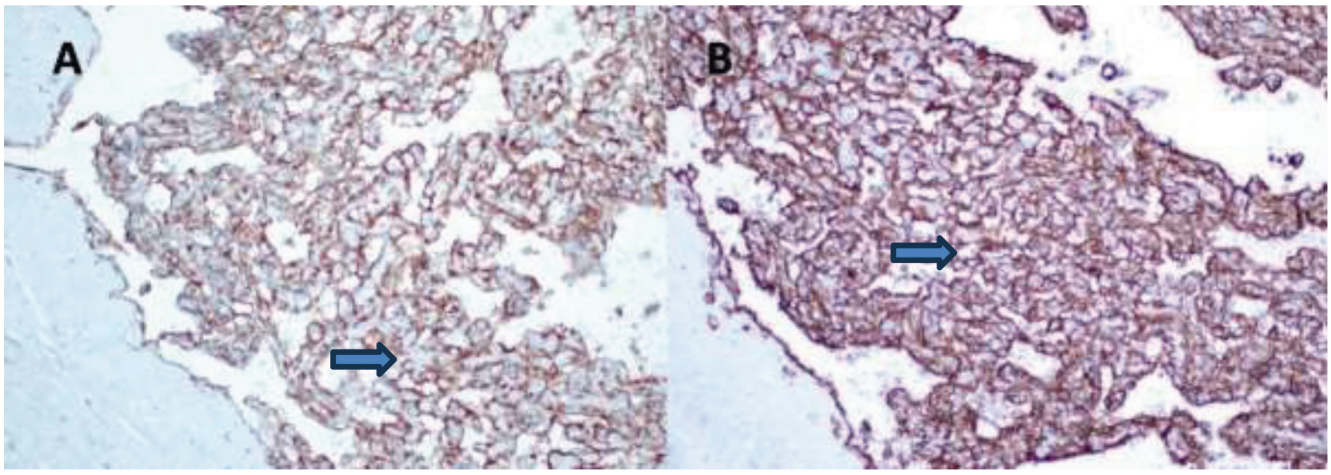

Figure 4. In the immunohistochemical study, tumor cells show diffuse positivity with CD31 (Panel A) and CD34 (Panel B) (x200)

Discussion

Breast cancer is the most common type of cancer and the leading cause of mortality among women worldwide (3-6). The World Health Organization classification categorizes invasive breast cancer into 21 unique histological categories based on cellular shape, growth characteristics, and structural patterns (7). Breast angiosarcoma is an extremely rare disease with an incidence rate of less than 0.05% of all breast cancers (2).

The appearance of BA's on mammograms is non-specific. Young women's high breast density can easily obscure findings such as focal asymmetry or an ill-defined non-calcified lesion (8,9). Yang et al. (10) analyzed BA patients and found that 1,9% of angiosarcomas are not visible on mammograms but are detectable with ultrasound imaging. Kaklamanos et al. (11) also pointed out that imaging methods like mammography and ultrasound can misdiagnose these lesions as benign since they don't show the typical signs of adenocarcinoma, especially in younger women. Breast MRI is frequently useful for detecting these types of lesions (11).

Sebastian et al. (12) revealed that the lesions may be overlooked in imaging studies. Despite their patients undergoing wide local excision, they recommended mastectomy as the primary treatment choice. Magnetic resonance imaging should be the primary modality, and vascular markers like CD31 and CD34 can assist in pathological diagnosis. In our case, we also used these markers for diagnosis (12).

Borrman (13) reported the first case in 1907, and due to the rarity of these cases, therapeutic recommendations and prognostic factors are not well-established. According to several studies, the surgery of choice for the majority of patients with angiosarcomas and primary breast sarcomas is total mastectomy, or in some cases, total mastectomy and axillary dissection. Kaklamanos et al. (11) suggested that smaller lesions could potentially benefit from breast-conserving surgery as a treatment option. There are also articles suggesting that axillary dissection is unnecessary because angiosarcomas metastasize via the vascular system.

Iacoponi et al. (14) reported performing a nipple-sparing mastectomy and breast reconstruction on a patient with a grade 1 angiosarcoma that completely occupied the right breast, followed by adjuvant chemoradiotherapy. They indicate that the patient is asymptomatic and disease-free thirteen months after the primary diagnosis.

Killoran and Dissanayake (15) reported a case of a 79-year-old woman diagnosed with primary BA. The patient had wide local excision, removal of muscle and fascia, and reconstruction of the chest wall using the latissimus dorsi flap. During her follow-up, the patient developed lung metastases and received palliative chemotherapy similar to our patient. They indicated that the impact of neoadjuvant treatment on the prognosis is unknown. They also observed that early MRI assists in the diagnosis (15).

Vohra et al. (16) documented a case of an 87-year-old patient diagnosed with primary angiosarcoma, with no metastases at the initial assessment. She underwent a mastectomy. They utilized CD34 markers to validate the final pathology specimen, similar to our approach. After a follow-up regimen of three months, the patient experienced widespread metastases, which ultimately led to her death from the disease. They recommended complete mastectomy as the surgical procedure and advised against axillary surgery unless axillary disease is confirmed (16).

Due to the small number of known patients with primary BA, there are no studies examining the effects

of adjuvant chemotherapy or radiotherapy. Some retrospective studies suggest that patients at high risk for recurrence (tumors of high grade and size) may benefit from adjuvant therapy. According to some authors, this tumor may be chemosensitive, and the administration of chemotherapy for specific patients can increase survival (11).

Our patient was a 38-year-old woman who presented to the hospital with right breast pain and swelling. The inability to make a definitive diagnosis in a rural hospital may be due to the absence of an MRI. This is because this type of tumor often does not display its characteristic features on mammograms and ultrasounds, especially in young women. She received neoadjuvant chemotherapy, during which she developed pulmonary metastases. Then she had a salvage mastectomy. The metastasis in her lungs regressed after the addition of palliative chemotherapy to her treatment plan. But finally, she developed brain metastasis and died of her condition. Due to the limited number of patients with primary BA, there is no specific guideline for determining surgery and adjuvant/neoadjuvant treatment plans.

Conclusion

This case demonstrated that the disease is challenging to diagnose and that MRI can serve as an effective diagnostic instrument. Our case demonstrated that, despite undergoing a salvage mastectomy and supplementary palliative care, a patient with lung metastases can experience disease progression. By analyzing our patient and the limited cases documented in the literature, clinicians can formulate a treatment strategy. Further research is necessary to establish particular guidelines for this kind of breast cancer.

Ethics

Informed Consent: All necessary informed consents were obtained from the participants

Footnotes

Authorship Contributions

Surgical and Medical Practices: M.B.A., K.C., S.K., A.Y., Concept: M.B.A., K.C., Design: M.B.A., K.C., Data Collection or Processing: M.B.A., K.C., S.K., Analysis or Interpretation: M.B.A., K.C., Literature Search: M.B.A., K.C., Writing: M.B.A., K.C., S.K., A.Y.

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