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Case Report
Olgu Sunumu

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Angiosarcoma of the Breast: Case Report

Memenin Anjiyosarkomu: Olgu Sunumu

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Abstract: Angiosarcoma is a rare and aggressive tumor of the breast arising from endothelial cells. It is usually associated with chronic lymphedema or radiation therapy. The diagnosis is made by biopsy and there is no specific finding on mammography. In this study, we report a 59-year-old woman with a history of radiotherapy for invasive ductal carcinoma who subsequently developed primary angiosarcoma of the breast. Angiosarcoma of the breast was first documented in 1887 and is classified as primary and secondary. While surgical R0 resection is recommended for the treatment of these tumors with unclear etiology, the significance of axillary lymph node dissection remains uncertain. Adjuvant radiotherapy has been shown to reduce the local recurrence rate, but adjuvant chemotherapy and radiotherapy have no significant effect on 5-year survival. Although there is no consensus on treatment regimens, it is concluded that more comprehensive studies should be conducted in a larger sample.
Keywords: Breast, angiosarcoma, radiotherapy

Özet: Anjiyosarkom, memede nadir görülen ve endotel hücrelerinden kaynaklanan agresif bir tümördür. Genellikle kronik lenfödem veya radyasyon tedavisi ile ilişkilidir. Tanı biyopsi ile konur ve mamografide spesifik bir bulgusu yoktur. Bu çalışmada, invaziv duktal karsinom nedenli radyoterapi öyküsü olan ve sonrasında primer meme anjiyosarkomu gelişen 59 yaşındaki kadın hasta sunulmaktadır. Memenin anjiyosarkomu, ilk olarak 1887 yılında belgelenmiş olup, primer ve sekonder olarak sınıflandırılır. Etiyolojisi tam olarak açıklanamamış olan bu tümörlerin tedavisi için cerrahi R0 rezeksiyon önerilirken, aksiller lenf nodu diseksiyonunun önemi belirsizdir. Adjuvan radyoterapinin lokal rekürrens oranını azalttığı, ancak adjuvan kemoterapi ve radyoterapinin 5 yıllık sağ kalım üzerinde belirgin bir etkisi olmadığı görülmüştür. Tedavi rejimleri konusunda fikir birliği olmamakla birlikte, daha geniş örneklemde daha kapsamlı çalışmalar yapılması gerektiği sonucuna varılmıştır.
Anahtar Kelimeler: Meme, anjiyosarkom, radyoterapi

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1. Introduction

Angiosarcoma is an aggressive tumor originating from endothelial cells. Although most commonly found in the breast, it is a rare mesenchymal tumor that can also occur in the heart, pericardium, liver, skin, bone, and soft tissues. Breast angiosarcoma constitutes approximately 0.04% of all malignant breast tumors (1). Its etiology can be spontaneous or associated with factors such as chronic lymphedema or radiation therapy. However, these types of angiosarcomas account for about 50% of all breast sarcomas and are linked to radiation therapy used in the treatment of primary invasive breast cancer (2, 3). The latency period for radiation-induced breast sarcoma development can range from 3 to 20 years. The incidence has been reported to be 0.3% at ten year and 0.5% at fifteen year (4). Angiosarcoma does not have specific findings on mammography, and diagnosis is generally confirmed through biopsy (5).

This case presentation discusses a 59-year-old female patient with primary breast angiosarcoma, supported by current literature.

2. Case Report

A 59-year-old female patient presented to our clinic with a raised lesion approximately 2 cm in diameter adjacent to the areola on her right breast. She has a medical history significant for diabetes mellitus, hypertension, and hypothyroidism. Her past medical records revealed a history of left modified radical mastectomy in 2010 due to carcinoma in situ of the left breast, which was diagnosed as high-grade comedo-type carcinoma in situ with microinvasive foci on final pathology, prompting clinical follow-up. During her second year of follow-up, a 1 cm lesion was detected in the right breast, confirmed as invasive ductal carcinoma on stereotactic excisional biopsy. Subsequently, she underwent segmental mastectomy and axillary dissection of the right breast, with no residual tumor found on pathology. She received chemotherapy and radiotherapy to the right breast as part of her oncological treatment. In her sixth year of follow-up, she presented with the 2 cm raised lesion near the areola of the right breast, which was biopsied and pathologically diagnosed as an atypical vascular tumor. After obtaining informed consent, she underwent right mastectomy (Figure 1). Histopathological evaluation of the specimen revealed tumor cells diffusely positive for CD31 (Figure 2). Additionally, at 200x magnification, numerous irregular and anastomosing vascular structures containing atypical endothelial cells were observed (Figure 3). Based on these findings, she was diagnosed with breast angiosarcoma and referred to medical oncology for continuation of oncological management.

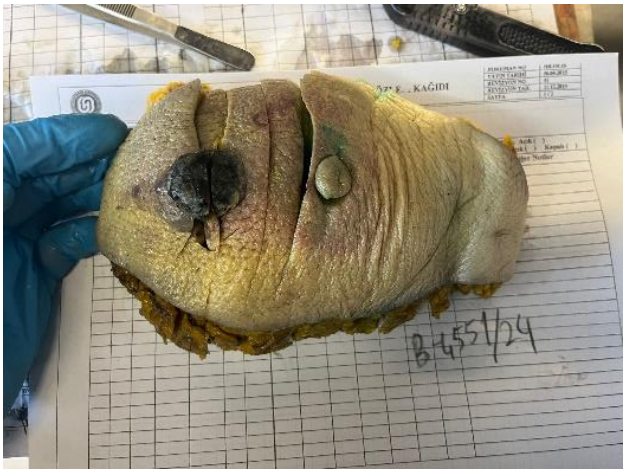

Figure 1. Right breast mastectomy material

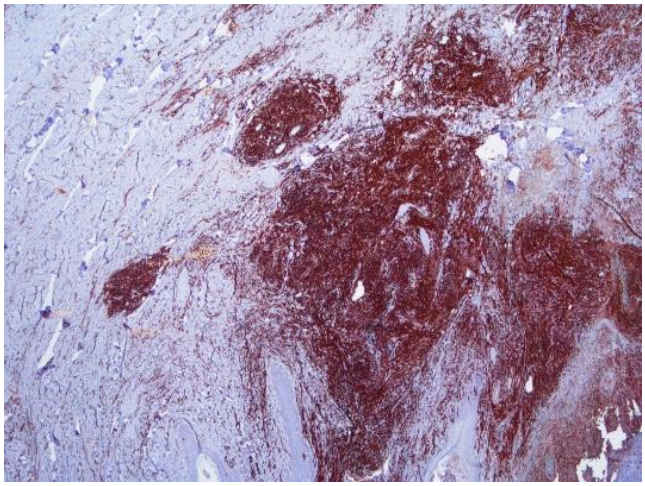

Figure 2. Tumor cells showing diffusely strong positivity for CD31

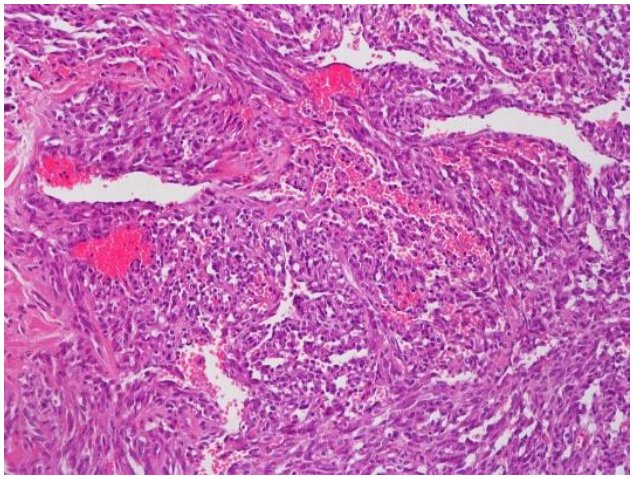

Figure 3. Tumorous lesion consisting of numerous irregular and anastomosing vascular structures containing atypical endothelial cells at 200 magnification

3. Discussion

Breast angiosarcoma was first documented by Schmit in 1887 (6). These malignant tumors originate from the endothelium of vascular structures surrounding the lobules of the breast. They are classified into primary and secondary types etiologically. Primary breast angiosarcoma tends to have an aggressive course with a generally poor prognosis, more commonly affecting women aged 30-50 years without any significant medical history or risk factors (7). The etiology of secondary breast angiosarcoma remains unclear; however, chronic lymphedema, exposure to chemical agents, ionizing radiation, chronic inflammation, and trauma are proposed etiological factors (8). Additionally, 6-12% of breast angiosarcomas occur in women during pregnancy and lactation, suggesting a potential role of high estrogen levels in etiology (9).

In a retrospective study by Kim et al. (10) involving 15 patients, 73.3% presented with masses larger than 5 cm, and the 5-year survival rate was 28.3% for tumors larger than 5 cm compared to 66.7% for tumors 5 cm or smaller, though statistically significant difference was not demonstrated (p=0.096).

There is no standardized treatment regimen for breast angiosarcoma, but surgical R0 resection is generally recommended as the cornerstone of treatment for all angiosarcomas (10). Toesca et al. (11) showed in their study

that total mastectomy is not superior to breast-conserving surgery, emphasizing the primary goal of surgery is to achieve negative surgical margins. In our case, a total mastectomy was performed considering the previous history of segmental mastectomy and the loss of nipple during negative surgical margins.

The role of axillary lymph node dissection in breast angiosarcoma is uncertain because angiosarcoma primarily metastasizes hematogenously (12). Merino et al. (13) in their study of 13 patients diagnosed with breast angiosarcoma, did not find axillary lymph node involvement. Therefore, in the absence of palpable axillary lymph nodes preoperatively, axillary lymph node dissection is not recommended (13). Axillary dissection was not performed because the patient had a history of axillary dissection and no positive axillary lymph nodes.

Although there is no consensus on adjuvant and neoadjuvant treatment protocol, studies have shown that adjuvant radiotherapy reduces local recurrence rates (14). Another study indicated a positive impact of adjuvant radiotherapy on recurrence-free survival when evaluating both primary and secondary angiosarcomas together. Concerns about angiosarcomas developing secondary to radiation exposure and complications related to radiation exist; however, this study suggests that patients who receive postoperative radiotherapy have lower rates of local recurrence (15). Conversely, Kim et al. (10) stated in their study that adjuvant chemotherapy and radiotherapy did not significantly affect 5-year survival ($p>0.05$).

In our patient with a history of radiotherapy after right segmental mastectomy and axillary dissection, radiation oncology did not recommend radiotherapy due to T1N0 and surgical margin negative angiosarcoma in the pathology after mastectomy, but adjuvant chemotherapy (Taxol) was started by medical oncology. Her treatment is still ongoing.

In conclusion, there is no consensus on the treatment of breast angiosarcomas, but R0 surgical resection is the recommended initial approach. Further comprehensive studies with larger sample sizes are needed to reach consensus on chemotherapy and radiotherapy regimens.

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