Primary Angiosarcoma of the Breast

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Abstract

A 21-year woman presented with right breast mass of ten months duration, she did not complain of breast pain or tenderness. On physical examination a firm non tender mass was palpable at right upper quadrant region. Fine needle aspiration cytology (FNAC) showed predominantly blood, few endothelial like cells mimicking vascular tumor and it was confirmed as angiosarcoma after biopsy and histopathological examination.

Key words: Angiosarcoma, Rare Breast disease, Bluish skin color lesion, Lump breast.

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Introduction

Angiosarcoma of the breast is an uncommon, extremely hostile neoplasm of vascular origin. Two hundred and nineteen cases have been described [1] since the first case reported by Schmidt [2] in 1887. The frequency of this rare tumor is 0.04% of primary mammary tumors[3] and approximately 8% of mammary sarcomas.[4] Several reports have been published with different names for this malignant condition, such as hemangioendothelioma, haemangioblastoma ,haemangiosarcoma, and metastasizing angioma . This neoplasm carries a very poor prognosis, with a five-year survival of 850%. [5] Metastases derived from mammary angiosarcomas have been reported in lung, skin, liver, bone, CNS, spleen, ovary, lymph nodes and heart . [6,7]

Case report

A 21 year old female presented with left breast mass of ten months duration, she did not complain of breast pain or tenderness.

On physical examination a firm non tender mass was palpable at right upper quadrant region. It was about (10x10) cm in diameter and was not associated with lymph node enlargement. The lump was not fixed to the chest wall, not associated with nipple retraction or discharge, nor skin tethering and was associated with bluish skin color lesion.

There was no history of previous breast surgery,breast irradiation or family history of breast cancer. Ultrasound findings showed a heterogeneous hyperechogenic mass with associated architectural distortion in the upper outer region of the right breast with a diagnosis suggestive of angiosarcoma.

Routine laboratory tests on blood and urinanalysis were normal.

Fine needle aspiration cytology (FNAC) showed predominantly blood, few endothelial like cells mimicking vascular tumor, as FNAC has a false-negative rate of 1% to 35%, so it was confirmed by biopsy which was diagnostic for the angiosarcoma. Right mastectomy was performed for her, and a ill circumscribed lesion measuring (10x10x8) cm with hemorrhagic and nodular cut surface was resected. Histopathological examination confirmed it as high grade angiosarcoma (Fig.1).

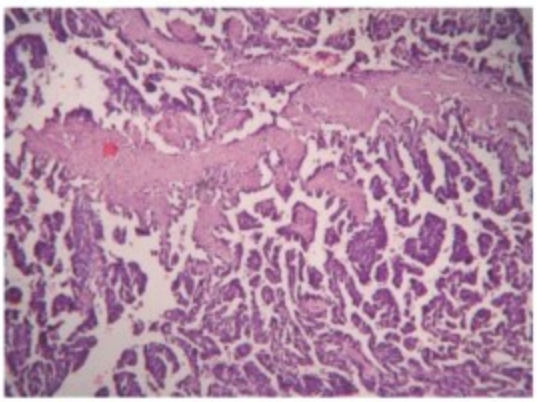

Fig.1: Histological section showing anastomosing vascular channels that invade the surrounding breast tissue (H&E X 400).

Discussion

Primary breast angiosarcoma is a connective tissue tumor that commences in the breast itself. It can be a sporadic event or related to a prior therapy. With the increasing use of breast conservation therapy for breast cancer, reports of post irradiation angiosarcoma have increased. Both primary and secondary angiosarcomas may present with bruise like

skin discoloration. Primary breast angiosarcoma is a rare type of tumor and accounts for 0.04% of all malignant breast tumors.[3]The annual incidence of mammary angiosarcoma is 5.8 per 10million women.[8] Primary angiosarcoma is diagnosed in patients who have never had breast cancer. Secondary angiosarcomas that are similar tumors occurring after breast treatment have also been reported. They occur after radical mastectomy or lumpectomy and radiation. The incidence of post irradiation angiosarcoma appears to be low, ranging from 0.090.16%.The tumor might be felt like a hard, painless mass in the breast; skin may be blue or red over the mass,and there may be some swelling and lumpiness in the area. Some patients reports feeling a tender or even painful mass, depending on the size of the tumor. This type of breast cancer is aggressive and the prognosis depends on the stage at the time of diagnosis. It can spread through blood stream to the lungs, liver, bones, and skin and to the other breast. This type of breast cancer has a high rate of recurrence. Patients with breast angiosarcoma usually have a poor prognosis, and only 10% to 27% of patients will survive this cancer about 10 year [9]. Pretreatment duration of the lesion and primary tumor size were not significantly related to the risk of recurrence or to survival. Total mastectomy is recommended for most mammary angiosarcomas [5] and surgical extirpation remains the only effective treatment.[6]

Three grades of angiosarcoma are described. Low-grade tumors consist of anastomosing vascular channels that invade the surrounding breast tissue.Intermediate-grade tumors have more solid neoplastic vascular growth and an increased mitotic rate. High-grade lesions, as in our case (Fig.-1), have frankly sarcomatous areas, as well as areas of necrosis, hemorrhage, and infarction. More than one grades may exist in the same tumor. The differential diagnosis of angiosarcoma includes metaplastic carcinoma, the acantholytic variant of squamous cell carcinoma, hemangioma and pseudoangiomatous stromal hyperplasia. Breast angiosarcoma is frequently advanced at diagnosis and has a tendency for local regional recurrence.[10]

Conclusion

The importance of this case report is that primary breast angiosarcoma is a rare disease which can often be missed or produce diagnostic difficulty or dilemma unless there is a preconception about it. Strong suspicion, related clinical findings and use of invasive and noninvasive diagnostic modalities with high level of interpretation are crucial to reach a diagnosis.

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