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PRIMARY ANGIOSARCOMA OF BREAST: A RARE CASE REPORT

Pathology

Dr. Shikha Sharma Senior Resident, Department of Pathology, IGMC SHIMLA
Dr. Gauri Nakra Junior Resident, Department of Pathology, IGMC SHIMLA
Dr. Anchana Gulati* Professor, Department of Pathology, IGMC SHIMLA *Corresponding Author

ABSTRACT

Primary breast angiosarcoma is an exceedingly rare malignancy representing less than 0.04% of total breast malignancies and approximately 8% of breast sarcomas. It presents as a rapidly growing painless palpable mass in third to fourth decade of life. We present the case of a 43 years old female who appeared to have chief complains of pain and a palpable mass in the right breast for 2 months. On palpation, there was a lump of size 6×5 cm, in the upper outer quadrant of right breast. No enlarged lymph nodes were palpated. FNAC of the breast lump suggested a preliminary diagnosis of Primary Angiosarcoma, which was further confirmed on histopathology and immunohistochemistry. Being a rare and aggressive entity, a quick diagnosis and robust treatment regime is required. Neo-adjuvant and adjuvant chemotherapy and radiotherapy are helpful in patients with high risk of recurrence.

KEYWORDS

Primary Angiosarcoma, Breast, Aggressive Malignancy.

INTRODUCTION

Primary breast angiosarcoma is an exceedingly rare malignancy representing less than 0.04% of total breast malignancies and approximately 8% of breast sarcomas (1). Breast angiosarcomas have been classified as primary, that arises de novo, and secondary, as consequence of chronic lymphoedema or breast irradiation after breast conserving surgery (2,3). Primary angiosarcomas of the breast generally evolve during the third and the fourth decades of life (median age 35), although cases have been noted in postmenopausal women as well (4,5). Patient usually presents with a rapidly growing painless and palpable mass (≥4 cm) with a sensation of fullness, and an exponential growth is seen within the breast, infrequently associated with purple blue discoloration of skin.

Although the etiology is unknown, the various risk factors associated with primary angiosarcoma breast are trauma, radiation and lymphedema (after treatment for breast cancer. It is a highly aggressive tumor with poor prognosis and a high risk of recurrence as well as metastasis. Primary angiosarcoma diagnosis is confirmed by core biopsy as fine-needle aspiration associated with nearly 40% of false negative results (6). Surgery is the main treatment modality with chemotherapy and radiotherapy used in case of recurrence.

Case Report

A 43 years old female presented with complains of pain and a palpable mass in the right breast of 2 months duration. On palpation, the left breast was unremarkable and in the right breast, a lump of size 6×5 cm, in the upper outer quadrant was noted with nipple retraction.

The lump was tender, firm in consistency, mobile and not fixed to the chest wall as well as to the overlying skin. No lymph nodes were palpable in bilateral axilla. The patient was then sent for routine laboratory examinations and was advised to go for bilateral mammography and FNAC of the breast lump.

Bilateral mammography revealed a large soft tissue density mass measuring 6.5×3cm with irregular margins in the right breast in outer quadrant in peri-areolar region with nipple inversion. No microcalcifications were noted and an impression of BIRADS IV was given.

FNAC of the right breast lump revealed cellular smears with isolated as well as loose syncytial aggregates of pleomorphic spindle to epithelioid tumor cells with hyperchromatic nuclei and fine chromatin, focally conspicuous nucleolus and moderate amount of finely vacuolated basophilic cytoplasm.

Intracytoplasmic eosinophilic material was seen focally along with the fragments of mature adipose tissue infiltrated by atypical tumor cells in a hemorrhagic background (Figure 1A &B). A preliminary diagnosis of Angiosarcoma right breast, was given.

The tru-cut biopsy of the breast mass was received as 3 grey-white to grey-brown biopsy cores, ranging in length from 0.5-1.5 cm. Microscopy revealed multiple biopsy cores comprising fibro-collagenous and mature adipose tissue along with foci of necrosis and hemorrhage devoid of breast lobules. Foci of viable tissue revealing anastomosing vascular channels, infiltrating the stroma, lined by pleomorphic atypical spindle to epithelioid endothelial cells having hyperchromatic nuclei with fine chromatin and eosinophilic cytoplasm along with scattered mitosis were seen. Solid foci of atypical spindle cells with foci of slit like spaces were also noted (Figure 2).

A focus revealed small buds/papillary fronds of endothelial cells with hyalinized stroma projecting into vascular space. Keeping in view the above findings,possibility of Angiosarcoma Right Breast, Intermediate grade was suggested and the patient was planned for radical mastectomy. MRM specimen was received and was examined.

Gross examination revealed right breast measuring 25×23×10 cm. Overlying skin (22×9cm), nipple-areola complex appeared unremarkable. On cut-section, there was a friable tan-brown tumor measuring 6.5×5.5×5 cm, in the retro areolar region.

The tumor revealed focal cystic, hemorrhagic and necrotic areas as well (Figure 3). The tumor was 2 cm away from the deep resection plane and 1 cm away from the overlying skin.

All the resection margins were grossly uninvolved. 15 axillary lymph nodes were palpated in the axillary tail ranging in diameter from1mm to 1.2 cm. Microscopy of the sections from the tumor revealed marked areas of necrosis and large blood lakes along with foci of tumor cells infiltrating the adipose tissue and fibrous breast parenchyma. The tumor cells were slender to plump spindle to epithelioid, having hyperchromatic nuclei with fine chromatin, inconspicuous nucleoli and eosinophilic cytoplasm (Figure 4A).

These tumor cells were lining the vascular channels separated by thin fibrous stroma with extravasated RBCs. Focal papillary projections (endothelial tufting) were seen intraluminally(Figure 4B). Mitosis was seen focally. All the resection margins were free from tumor. Sections from skin, nipple, areola were unremarkable. All the 15 lymph nodes were free from tumor invasion. Based on the above findings the final diagnosis of Malignant Vascular Tumor with morphology favouring high grade Angiosarcoma, right breast was suggested.

Routine IHC for breast tumors which included ER, PR and Her 2 neu was done in our department which came out to be negative. Further IHC including vascular markers were advised but patient refused due to financial constraints. She was put on chemotherapy regimen and on follow up , she developed paraplegia with widespread metastasis and breathed her last after one and half years of diagnosis.

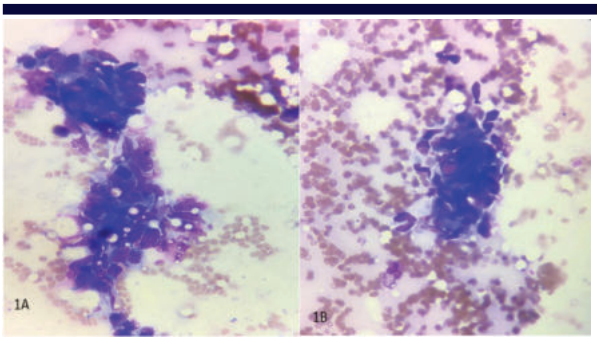

Figure 1A & B: FNA of breast lump revealing aggregates of pleomorphic epitheliod to spindled cells with eosinophilic secretions and hemorrhage in the background.(Giemsa, 400X)

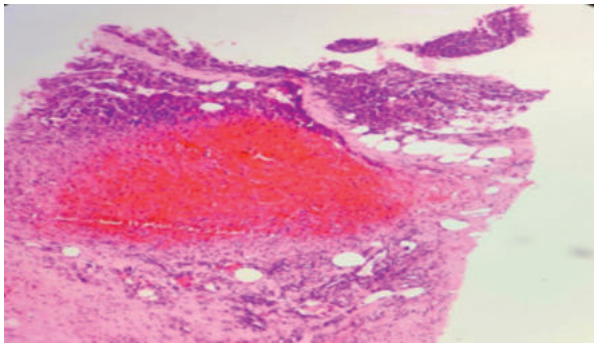

Figure 2 : Trucut biopsy revealing fibrocollagenous stroma with foci of hemorrhage and anastomosing vascular channels and spaces lined by atypical spindled cells(H&E,100X)

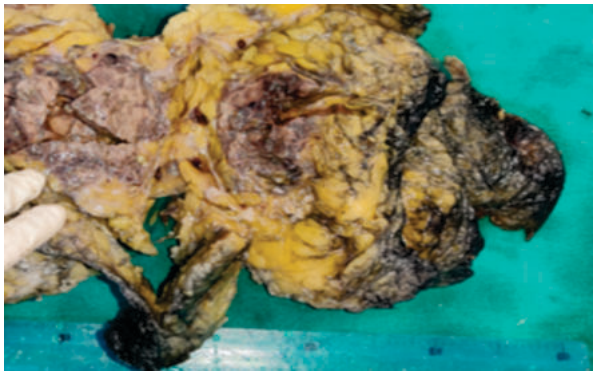

Figure 3: Gross image of mastectomy specimen revealing tan brown tumor with areas of hemorrhage

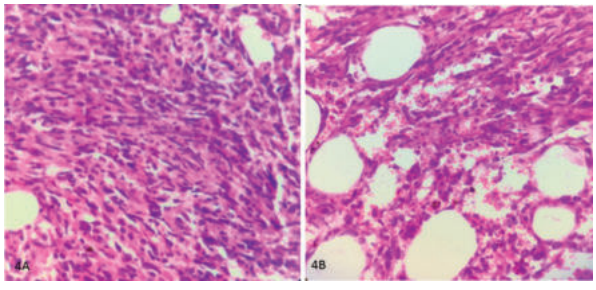

Figure 4A& 4B: High power view of tumor comprised of plump spindled to epitheliod cells infiltrating the fibrous beast parenchyma in 4A and are lining the vascular channels with extravasated RBCs and endothelial tufting as seen in 4B(H&E,400X)

DISCUSSION

Angiosarcoma is the most common sarcoma to occur in the breast, but is relatively rare. Angiosarcoma occurs almost exclusively in the female breast, with only five cases of male breast angiosarcoma reported in the literature. Preoperative diagnosis, by FNAC and biopsy, may be difficult task for a pathologist if the specimen size is small. Large-core biopsies are usually helpful to make the correct diagnosis as they provide a larger sample, but because of the vascular nature of these tumors, such a macrobiopsy is often difficult to perform. Three main histopathological patterns have been described in literature: grade 1- characterized by vascular channels invading the breast tissue with scarce endothelial proliferation, grade 2- comprising papillary endothelial components, grade 3 reveals necrosis and hemorrhage addition. (7) Before diagnosing primary angiosarcoma of breast, other rare histologies like liposarcoma, fibrosarcoma, osteosarcoma,metaplastic carcinoma, and stromal sarcoma, are excluded as part of a differential diagnosis (4). Presence of CD31 and factor VIII in immunohistochemical assessment are also contributory in diagnosing primary angiosarcoma and the vascular origin of the tumour(8).

On mammograms, angiosarcomas appear as an ill-defined mass and lack the spiculations often seen in breast carcinomas as in our case . Sonography usually shows a solid mass that may have well-defined or lobulated margins, with both hypoechoic and hyperechoic appearance. Magnetic resonance imaging (MRI) of angiosarcoma shows a mass with low signal intensity on T1-weighted images, but high signal intensity on heavily T2-weighted images. The latter suggests the presence of vascular channels containing slow flowing blood.

Surgical resection is the mainstay of treatment. Total mastectomy, sparing the axillary lymph nodes is usually preferred, unless the lymph nodes are palpable. After the resection, adjuvant chemotherapy is often given, as tumor free margins are very difficult to attain in these cases and many patients present with local recurrence. As vascular involvement is the most common route of metastasis for the primary angiosarcoma, local lymph-node involvement is extremely rare; thus, making a routine local lymphadenectomy not necessary (9). Most often the enlarged lymph nodes, in the course of primary angiosarcomas, result from the local compression of the large tumor, and are reactive only.

According to Rosen's study, the 5 years disease free survival rate for low grade tumors can be as high as 76% and up to 70% for intermediate grade tumors. Whereas 5 years survival rate for high grade tumors is about 15% (10).

CONCLUSION

Primary breast angiosarcoma is an exceedingly rare and aggressive malignancy that requires a robust treatment regime. A definitive diagnosis should always be made on core biopsy. The role of neo-adjuvant and adjuvant chemotherapy and radiotherapy is still ill-defined; however, these should be considered in patients with high risk of recurrence. Because of the rarity of the disease, patients with primary angiosarcoma should be managed at highly specialized medical units.

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