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Primary Angiosarcoma of Breast in Elderly Woman

Kanmani Velarasan .S, Deivanayagam Ramasundaram

Abstract: ***Introduction: Breast angiosarcoma is a rare tumor. It can be presented as primary or secondary type. Primary angiosarcoma arises spontaneously, while secondary angiosarcoma arises secondary to radiation. It presents as painless palpable masses clinically and confuse with common malignancies of the breast. Diagnosis with fine needle aspiration is difficult. Ultrasonography shows hypervascular mass. Immunohistochemical markers help in diagnosis. Surgery is the mainstay of treatment. Role of adjuvant chemotherapy and radiation therapy remains to be defined.***

Keywords: Angiosarcoma , breast, radiology, immunohistochemical markers, surgery, radiation therapy

Abbreviations: FLI 1 – Friend leukemia integration 1 transcription factor, ERG – Erythroblast transformation specific transcription factor related gene, CD 31 – Cluster of differentiation 31.

1. Introduction

Breastangiosarcoma is auncommon neoplasm. It accounts for about 0.05% of primary affecting breast. It affects quite younger patients. It arises fromcells lining the blood vessels of the breast. It has a moderate risk of local recurrence and high risk of metastases. We like to present a case of primary angiosarcoma of breast in a elderly woman.

Presentation and diagnosis
66 year old female presented with the complaints of lump in the left breast for five months duration. She was evaluated elsewhere initially. She underwent lumpectomy in December 2016. Histopathology specimen was reported as angiomatous neoplasm with pseudoangiomatous stromal hyperplasia. She did not undergo any adjuvant treatment. Lesion recurred in 6 months. Fine needle aspiration cytology of the lesion was suggestive of spindle cell lesion. Ultrasound of the breast showed mixed echoic mass lesion. Metastatic work up was found to be negative. She underwent Modified radical mastectomy.

Gross description
Mastectomy specimen measuring 16*14*12 cm overall and weighing 1220 gms . It has an overlying brown erythematous , elliptical portion of skin measuring 12*7 cm containing a 2.5*2.5 cm areola with 1.5 cm diameter averted nipple. The posterior surface is covered by fascia and inked out. An irregular grayish brown mass 6.5*5*4.5 cm with spongy areas seen, 1.5 cm from the deep margin and 2 cm from the skin. The mass is 2.5cm or more from all the other margins. Axillary pad of fat 4.5*3*2.5 cm with six nodes seen.

Microscopic description
Section studied shows breast tissue with a lesion composed of many irregular vascular spaces lined by attenuated endothelial cells with wide areas of hemorrhage and marked congestion. The dilated vascular channels and papillae are lined by scattered low cuboidal epithelium cells showing moderate cytoplasm and oval to spindle shaped cells with nuclear atypia and increase in mitosis.

Section from the residual breast shows features of fibrocystic lesion. Section from the skin above is free of tumor infiltration. The pectoralis muscle is adherent the tumor with foci of infiltration. Section from the nipple, areola and all the resected surface margins are free of tumor infiltration. Section from all the 6 nodes show reactive follicular hyperplasia with sinus and no metastatic deposits. Histopathology was reported as suggestive of angiomatous neoplasm.

Immunohistochemistry study was done. Markers FLI 1, ERG and CD31 was tested. FLI 1 and ERG showed diffuse nuclear positivity. CD 31 showed diffuse cytoplasmic positivity in 90% tumor cells. Diagnosis was reported as Angiosarcoma.

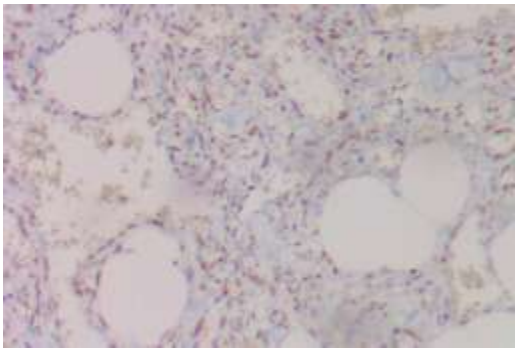
Figure showing positivity for FLI 1

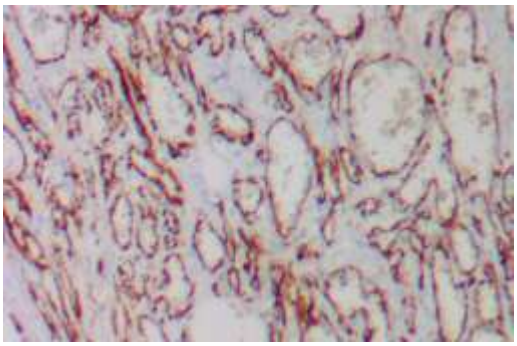
Figure showing positivity for CD 31

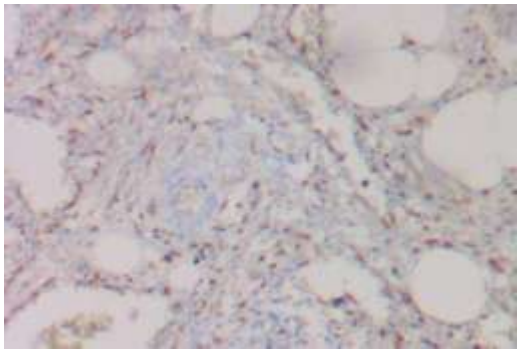

Figure showing positivity for ERG

Her clinical condition was discussed in our multidisciplinary tumor board. She was advised adjuvant radiation therapy.CT thorax was done before starting adjuvant treatment. There were no metastases in lungs. She was advised radiation therapy. Dose prescribed was 50 Gy in 25 fractions to chest wall alone.

2. Discussion

Mammary sarcomas are a mixed group of malignant neoplasms that arise from the mammary stroma (3). It arises from cells lining the blood vessels of the breast (4). Breast angiosarcoma can be primary neoplasm (arising spontaneously) or secondary angiosarcoma due to radiotherapy for breast carcinoma. Primary angiosarcoma of the breast is exceedingly rare, and accounts for approximately 0.04% of malignant breast neoplasms. Its incidence among breast sarcomas varies from 2.7% to 9.1%. The incidence of primary breast angiosarcoma is about

17 new cases per million women (2). Breast angiosarcoma is more frequent in young women (20 to 50 years).

Breast angiosarcoma presents as a painless, palpable massand rapidly growing in most of the documented cases. The right breast is more commonly involved than the left breast (9). In most cases, tumor size at diagnosis is bigger than 4 cm. In few patients, large tumorscan lead to thrombocytopenia and hemorrhagic manifestations (Kasabach–Merrittsyndrome) (5). It can involve contralateral breast even before dissemination, suggesting multifocal involvement (12).

Radiology:
Mammogram: They do not exhibit any pathognomonic features. They often appear as ill-defined masses on mammograms. Calcifications can be seen unlikefrom those seen in breast carcinomas (11). Ultrasound: Breast angiosarcomamay appear as solid lesion which can have well-defined or lobulated margins. They have both hypoechoic and hyperechoic appearance. There is often no acoustic shadowing (11). MRI: Magnetic resonance Imaging can show typical malignant signs (hyperintensity on T2 images and a rapid initial intense phase followed by washout). MRI is considered as helpful investigation (5).

Diagnosis prior to surgery, either by FNA (Fine needle aspiration) and biopsy is difficult(10). Pathologically, these tumors are subdivided into three groups according to the classification proposed by Donnel(6).

Grade I (well differentiated): Single layer of endothelial cells lining these channels dissect through the stroma causing distortion and little destruction of the preexisting lobules and ducts. The endothelial cells are usually flat. Nuclei may be hyperchromatic and contains small nucleoli. Solid and spindle cell foci, blood lakes and necrosis are not present.

Intermediate grade angiosarcoma differs from low-grade by containing additional cellular foci of papillary formations and/or solid and spindle cell proliferation.

Grade III: In these type, endothelial tufting and papillary formations are prominent. Conspicuous solid and spindle cell areasmostly devoid of vascular formations, are present as well. Mitoses may be brisk, mainly in more cellular areas. Areas of hemorrhage called as “blood lakes,” and necrosis are also seen.

Differential diagnosis of this rare tumor includes : 1.Benign hemangioma, 2.Phyllodes sarcoma, 3. Stromal sarcoma, 4.Metaplastic carcinoma, 5.Fibrosarcoma, 6.Liposarcoma, 7.Squamous cell carcinoma with sarcomatoid features, 8.Myoepithelioma, 9.Fibromatosis, 10.Reactive spindle cell proliferative lesion, and 11.High-grade mammary carcinoma (10).

Immunohistochemical stains for epithelial markers (pancytokeratin), endothelial markers (CD34 and CD31), factor VIII and CD31 immunostaining (11)and other sarcoma markers should help in making the accurate diagnosis.

Surgery is the primary mode of treatment .It consists of total mastectomy (1). Bosquet et al showed the axillary lymph node involvement is less than 10% (8). Axillary dissection is not necessary. Hematogenous dissemination is the main mode of metastases (11).

Chemotherapy: There is no consensus regarding the type of regimen to be used. Chemotherapy appears to be beneficial in high-grade lesions and in the metastatic setting.Hirata et al showed an improvement in overall survival rate in patients with metastatic angiosarcoma treated with taxane regimens(7).

Radiotherapy: Adjuvant radiotherapy may be beneficial for patients with microscopically positive margins (5).In some series, adjuvant radiation was offered based on tumor features(8) . Due to small number of patients in the studies, statistical significant correlation between radiation and overall survival is difficult to comment (5).

Prognostic factors: Prognosis does seem to be influenced by the grade. Well differentiated (grade I) tumors have a better prognosis than poorly differentiated (grade III). Low grade tumors have lower local recurrencesand distant metastases (1)

3. Conclusion

The diverse features, rarity, diagnosing and treatment approachof this tumor needs to be stressed. Multi

disciplinary approach involving onco-surgeon, pathologist ,radiologist, radiation and medical oncologist is necessary formanagement.Awareness of clinical and pathological features of this tumor is essential to not to miss the diagnosis and delaying of definitive treatment.

Conflicts of interest: None

Acknowledgements: None

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