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Angiosarcoma – A Case Report.

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ABSTRACT

Angiosarcoma is an aggressive malignant tumor of vascular origin. We present a case of secondary angiosarcoma in a 66 year old female patient, who presented with a history of left breast wide local excision with intraoperative brachytherapy. The only chance of curative treatment for secondary angiosarcoma is extensive surgery, preferably with resection of all irradiated tissue.

Keywords: Angiosarcoma, Metastasis.

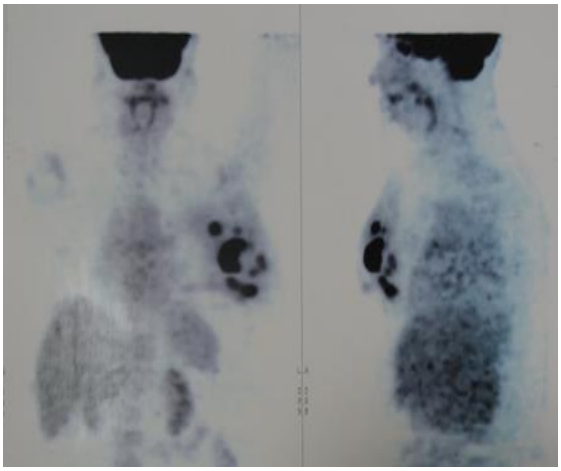
INTRODUCTION

Angiosarcoma is an aggressive malignant tumor of vascular origin, a rare entity in breast, accounting for 0.04% of malignant breast tumors. Primary angiosarcoma tends to occur in younger patients and mostly presents as a palpable mass. Secondary angiosarcoma usually occurs in older females as a sequelae of radiation, surgery or in chronic lymphedema, mostly in 6th decade. The median latency period is of 71 months.

CASE REPORT

We present a case of secondary angiosarcoma in a 66 year old female patient, who presented with a history of left breast wide local excision with intraoperative brachytherapy (15 Gy in 6 fractions). FNAC prior to the procedure was suggestive of adenocarcinoma but final HPE showed pleomorphic adenoma and she received no adjuvant treatment. Eight years later, she presented with complaints of multiple small nodules with erythematous patches over her left breast. There was a 2x2cm lump in the retro areolar region with left ecchymosis along with skin edema Her mammogram showed a speculated mass in the retroareaolar region and thickened subcutaneous plane with multiple densities. Subsequent MRIand PET CT breast showed an enhancing retroareolar lesion measuring 12 x 5, multiple small cystic areas, cutaneous deposits, diffuse thickening of the skin and avid stranding of parenchyma and muscle. An ultrasound-guided biopsy was performed which on histopathology along with IHC was suggestive of angiosarcoma. The tumor cells expressed vimentin, CD31 and were negative for CK, CK7, CK5/6, SMA, Desmin, CD34, S100, Melanin A. She underwent left simple mastectomy. Final histopathology showed a 7x6cm brown black lesion section with well-formed anastomosing vessels and solid sheets of high-grade epithelioid cells consistent with a high-grade epithelioid angiosarcoma.

She received hypo fractionated radiation therapy to chest wall to a dose of 50 Gy in 20 fractions by 6 & 15 MV photons followed by scar boost to a dose of 6 Gy in 3 fractions using 6 MeV electrons.The patient is in follow-up with no evidence of recurrence till date.



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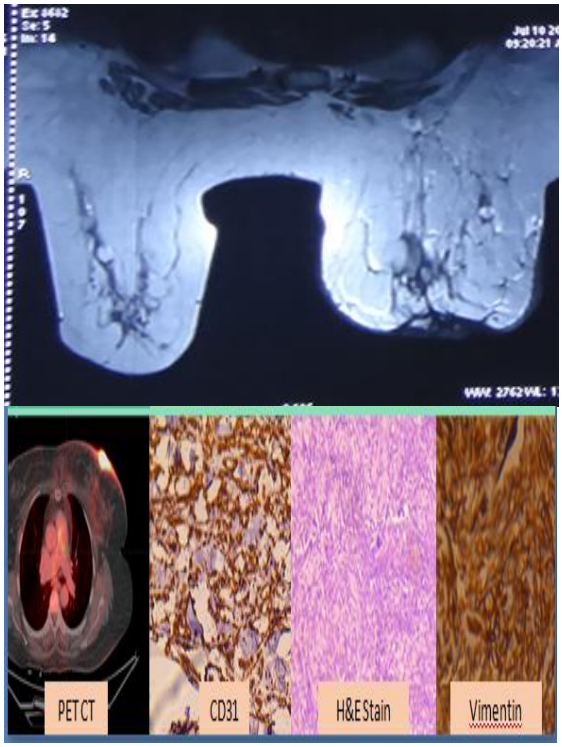

DISCUSSION & CONCLUSION

Angiosarcoma usually presents as a painless and rather quickly enlarging, palpable mass without tenderness. The tumor size is usually more than 4 cm in diameter. In 90% of cases, skin changes are noted (often confused with post-irradiation skin changes) consisting of breast edema, bruising associated with pain, hemorrhage, focal, elevated skin nodules, papules and vesicles.[1] Mammogram may not pick up an abnormality in close to 73% of cases. Sonography is useful for confirmation of a palpable mass. Color Doppler sonography may be useful in picking up hyper vascularity.[2] MRI does have a role in early detection. MRI findings of radiation associated angiosarcoma include rapidly enhancing dermal and intraparenchymal lesions, some of which are low signal on T2 weighted imaging.[3] The diagnostic test is a full thickness skin biopsy, which on IHC, is cytokeratin negative, positive for vimentin and factor VIII,positive for CD-1 and CD-34, expresses von Will brand factor, CD 34, CD 31, and vascular endothelial growth factor (VEGF) and CD117,absence of melanocytic markers (S100), human melanoma black-45, and melanoma antigen. The estimated probability of disease-free survival 5 years after initial treatment is about 76% for patients with grade 1 tumors, while it is about 15% for patients with grade 3 tumors.[4] Recurrence rates approach close to 70 percent. Disease recurrence is known to happen locally in the axilla. Metastasis to contralateral breast, to other sites, such as liver, lungs and bones is well known. The only chance of curative treatment for secondary angiosarcoma is extensive surgery, preferably with resection of all irradiated tissue. A report from Florida suggests that hyper fractionated irradiation may decrease rates of local recurrence and should be used in most favorable clinical situations.[5] In locally advanced cases- hyper fractionated therapy be given prior to surgical resection. There is limited data on value of chemotherapy.

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