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PRIMARY ANGIOSARCOMA BREAST- A CASE REPORT

General Surgery

Rahul M — Junior Resident, Department Of General Surgery, K.S Hegde Medical Academy, Deralakatte, Mangalore

Balakrishna N Shetty* — Professor, Department Of General Surgery, K.S Hegde Medical Academy, Deralakatte, Mangalore *Corresponding Author

ABSTRACT

Primary breast Angiosarcoma is a rare form of breast cancer, accounting for only 0.04% of all malignant breast tumors. we report a case report of 43 year old female presented with ulceration and skin discoloration of right breast, wedge biopsy shows features of Angiosarcoma, after metastatic work up patient underwent right Modified radical mastectomy following which she received adjuvant chemotherapy. At present patient is alive and doing well.

KEYWORDS

INTRODUCTION

Angiosarcoma is the most common sarcoma to occur in the breast, but is relatively rare. Angiosarcoma usually presents as a palpable mass, but 17% of cases may present with a bluish discoloration or bruising of the overlying skin. It arise from stromal tissues and extremely rare, accounting for <1% of all malignant breast tumors[1].

CASE REPORT

A 43-year old female, presented with history of Pain in the right breast since 1month, which was insidious in onset progressive in nature dull aching type, associated with discoloration and ulceration of the skin over right breast since 15 days. On examination 12 x10cm solitary lump involving all quadrants of right breast, tender, fixed to breast tissue and skin.3 x3 cm irregular ulcer present over the right breast lump, which bleeds on touch. Multiple right axillary lymph nodes palpable (central group). (Figure 1)

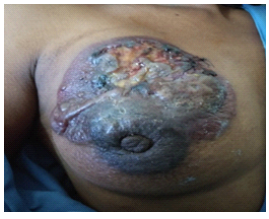
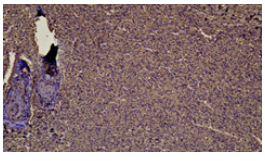


Figure 1

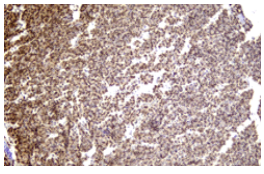
INVESTIGATIONS

CECT Thorax shows-10 x10 x 4cm well defined heterogeneously enhancing soft tissue dense lesion involving skin, subcutaneous plane and mammary parenchyma with multiple enhancing vascular channels and enhancing solid components features suggestive of angiosarcoma. Wedge biopsy of ulcer shows features of Angiosarcoma. Patient underwent Modified radical mastectomy. Histopathology examination of specimen shows well differentiated Angiosarcoms, No Lymphovascular invasion, No Metastatic deposits in lymph nodes. IHC-Positive for vimentin, CD31, CD34, FLI1 with Ki-67: 60%

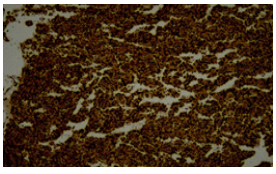


Vimentin V9 Positive

CD 31 positive



CD 34 Positive



FLI-1 Positive

CASE DISCUSSION

Angiosarcoma of breast can be primary or secondary. Primary angiosarcomas usually occur in young women (20–50 years)[2] .Angiosarcoma occurs almost exclusively in the female breast, rare in male. Bilateral tumors have been reported and several cases which have been diagnosed in the postmenopausal women. It often presents as discrete, palpable, non-tender lump associated with bluish discoloration of skin over breast. On mammograms, angiosarcomas appear as an ill-defined mass and lack the spiculations often seen in breast carcinomas. Rarely, the tumor may show a soap bubble appearance with a mean size of about 4.5 cm. They rarely manifest coarse non-branching microcalcifications[3]. 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. The differential diagnoses of this rare malignancy include benign haemangioma, cystosarcoma phyllodes, stromal sarcoma, metaplastic carinoma, fibrosarcoma, liposarcoma, and reactive spindle cell proliferative lesions[4].Based on histology primary angiosarcoma of breast is classified into 3 grades. Grade I-well differentiated tumor, Grade II- Moderately differentiated tumor, Grade III-Poorly differentiated with a sarcomatous spindle cell pattern. Factor VIII and CD31 immunostaining confirms the diagnosis. Primary angiosarcoma of the breast spread haematogenous to lungs, liver, bones, skin and to contralateral breast[5]. Available treatment options include surgery i.e gold-standard treatment consisting of modified radical mastectomy combined, when necessary, with axillary node dissection[6]. Chemotherapy (Paclitaxel) has been shown to produce excellent response in a number of studies in patients with primary angiosarcoma of breast. The role of radiation treatment is limited to the residual microscopic disease after surgery of the primary tumor. The potential impact of radiotherapy on local control and survival of patients with soft tissue sarcomas of other sites has been well documented. Adjuvant radiotherapy after surgery may have a beneficial effect for breast sarcomas, particularly for patients with microscopically positive margins. Agents such as Bevacizumab or Rapamycin which are anti VEGF factor may have a role in its treatment[7]. According to Rosen's study, the 5 years disease free survival rate for low grade tumours can be as high as 76% and up to 70% for intermediate grade tumours. Whereas 5 years survival rate for high grade tumours is about 15%[8].Our patient underwent modified radical mastectomy followed by 6 cycles chemotherapy and radiotherapy. Patient is now on regular follow up.

CONCLUSION

Primary Angiosarcoma of breast is a rare disease with no established standard treatment protocol. However surgery is considered as a primary treatment option. Chemotherapy, Radiotherapy and immunotherapy can be considered as adjuvant treatment modalities in its management.

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