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RADIOGENIC ANGIOSARCOMA OF THE BREAST (RASB): A CASE AND IMAGING FEATURES

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Abstract

Radiogenic angiosarcoma of the breast (RASB) is a rare late sequela of breast irradiation following breast-conserving surgery for invasive breast cancer and carries a poor prognosis; it is a rare finding and the clinical presentation is uncharacteristic; often present as multiple, distinct lesions.
Diagnosis is made by an abnormal mammogram, ultrasound, or MRI resulting in a core biopsy [1]; mammography may not have any suspicious findings, and ultrasound findings are nonspecific in one-third of patients [2].Surgery is the mainstay of treatment for localized disease while systemic chemotherapy and re-irradiation are appropriate for women with disseminated or recurrent RASB with a good long-term outcome in selected cases [6].
We present the case of a 82 yo woman with RASB, poorly indifferentiated angiosarcoma of the left breast, after a history of invasive ductal cancer of the left breast, treated in 2007 with quadrantectomy, linphoadenectomy and radiotherapy.

Keywords: *sarcoma, breast cancer, angiosarcoma, rasb*

Introduction

Radiogenic angiosarcoma of the breast (RASB) is a rare late sequela of breast irradiation following breast-conserving surgery for invasive breast cancer and carries a poor prognosis; it is a rare finding and the clinical presentation is uncharacteristic; often present as multiple, distinct lesions.
Angiosarcoma is an high grade tumor of endothelial origin and the most common subtype of the primary breast sarcomas; the other subtypes of primary breast sarcomas are named based on their type of cell origin and include fibrosarcoma, leiomyosarcoma, osteosarcoma, liposarcoma, chondrosarcoma, malignant histiocytoma, and Kaposi sarcoma. Angiosarcomas are graded by the appearance of histologic behavior of the endothelial cells based on nuclear atypia, mitotic activity, papillary formations, and presence of blood lakes.
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Due to the rarity of RASB, there is no standardized therapy regimen for women with this disease. Radical surgery of the tumor either by local resection or mastectomy is the most commonly cited treatment [3-5] and complete tumor resection is associated with an improved prognosis.
Surgery is the mainstay of treatment for localized disease while systemic chemotherapy and re-irradiation are appropriate for women with disseminated or recurrent RASB with a good long-term outcome in selected cases [6].

Case report

We present the case of a 82 yo woman with RASB, poorly indifferentiated angiosarcoma of the left breast, after a history of invasive ductal cancer of the left breast, treated in 2007 with quadrantectomy, linphoadenectomy and radiotherapy.
In August, a mass of 4.3 cm was diagnosed in the left breast with subcutaneous thickening. RASB was diagnosed by core biopsy with three samples obtained with a 14G needle and the patient undwerent chemoterapy treatment.
After 5 months, a disease progression was diagnosed with an enlargement of the lesion (6 cm diameter) and a controlateral lesion in upper outer quadrant appeared (22x10 mm).
In September 2020, a mass of gastric body (6 cm diameter) was diagnosed during and abdomen ultrasonography and histological report of an epithelioid angiosarcoma.
A total body CT was performed on Jan 2021; the exam showed: encefalic secondary lesions, pulmonary metastases with pleural effusion, right adrenal metastases and abdominal lymphonode (Image).
Figure shows the US presentation of the RASB.
The tumour presented positivity for ERG,CD31,CD34 and myc but negativity for cytocheratin, (pan, 5/6,8/18), EMA, CD45-LCA, Er and PgR receptors and HER2). Proliferation index (Ki67) was 60%.

Discussion

RABS has no pathognomonic imaging features (also because is a rare neoplasm) and can mimic an invasive breast carcinoma. Clinically, a painless breast mass can occur.
On mammography it can present with a architectural distortion and/or noncalcified irregular or oval breast masses with indistinct margins; they are not as circumscribed as other breast sarcomas and grow as ill-defined hemorrhagic mass lesions with less cellular components widely dispersed around the main portion of the tumor, with consequent architectural distortion [7].
In our case, no mammography should be done because of the dimension of the mass and the difficulty to execute the exam; only clinical evaluation and ultrasonography was done [Fig. 1]
Total body CT was performed to evaluate secondary lesions and in thorax acquisition a good evaluation of the breast was done [Fig.2]

Ultrasonography shows hypoechoic, irregular masses with indistinct or spiculated margins. Masses may be circumscribed or ill-defined and hypoechoic or hyperechoic; it can also present as diffuse, mixed echotexture regions without a discrete mass, as was observed in our presented case [8]. Color doppler may show hypervascularity .
Calcifications may be present due to phleboliths [4].

MRI can show irregular lesions with spiculated margins and a broad spectrum of internal enhancement characteristic; it presents a rapid initial signal increase and delayed washout in the kinetic analysis [9].
Tumor dimensions are an important prognostic factor; sarcomas with a diameter less than 5 cm is associated with a better outcome; margin status of resected tumors is a major factor in recurrence [3].
In our case, the patient suffer from claustrophobia and she refused Magnetic Resonance; only CT gave us other features of the lesion.
A high index of suspicion and careful attention to radiology-pathology concordance is essential to ensure that one does not overlook and delay the diagnosis of this aggressive malignancy [4]. Immunohistochemistry studies of the tissue are also essential, not only for distinguishing sarcomas from carcinomas but also for classifying sarcomas further into the array of different subtypes.
The most common treatment is radical surgical excision without lymph node dissection. As most patients who develop radiation- associated angiosarcoma have undergone breast conserving therapy, total mastectomy with radical resection of all previously radiated skin is the recommended surgical treatment. This includes radiated skin even outside the borders of the breast [7].

Patients who undergo total mastectomy who have a previous history of radiation are also considered to be candidates for immediate reconstruction.
The most common site of distant metastasis was the lungs and pleura, followed by liver, bone, and superficial soft tissues.

Conclusion

Our study shows that the imaging findings of primary breast sarcoma are not pathognomonic and can mimic those of breast carcinoma; a rapidly growing lesion and in case of a positive anamnesis for breast surgery and irradiation should make think about a RABS hypothesis.
In literature, there are very few studies on this topic and searching the terms "radiogenic breast sarcoma" on Pubmed, only two results are available in the last ten years [6, 10].
Very few information are available for imaging detection and characterization, but a multidiscipinary evaluation can guide the correct diagnosis.
In conclusion, we found that RASB is a rare late complication of breast irradiation. The prognosis of women with RASB is poor. Surgery is the mainstay of treatment for localized disease while systemic chemotherapy and re-irradiation are appropriate for women with disseminated or recurrent RASB.

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Figure 1. shows ultrasonographic appearance (a, b) of a case of radiogenic angiosarcoma of left breast in trapezoidal and classical image of exam conducted with multifrequency linear probe.
c) shows a contralateral lesion with suspicious features

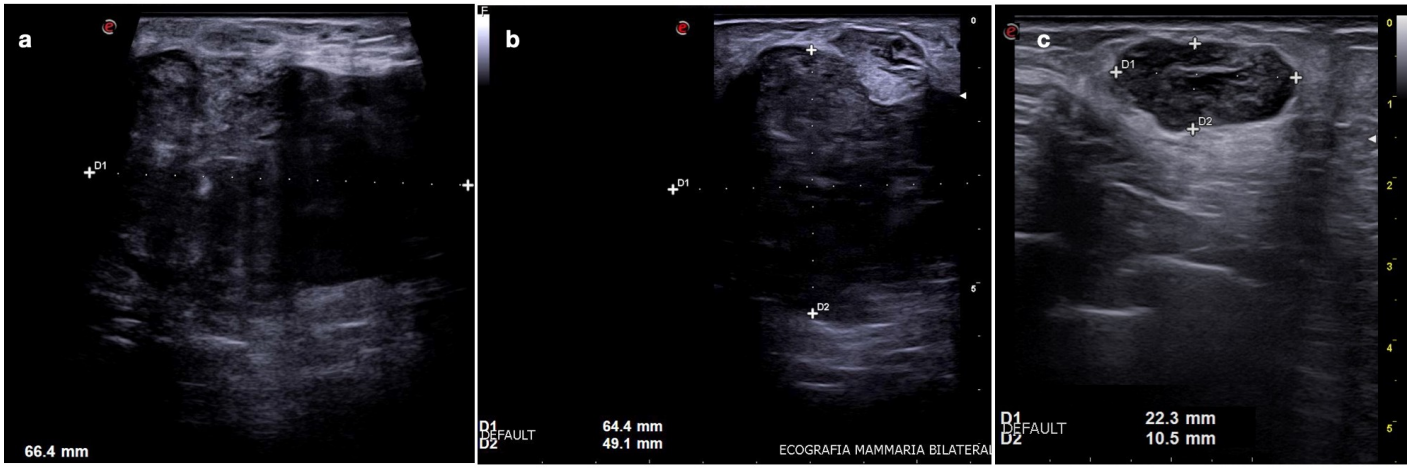

Figure 2. CT images showing the breast mass on eft side (a) and the other locality on right breast (b). The CT scans show also pleural effusion on left side and pulmonary atelectasy of left lower lobe.