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Case Report

Angiosarcoma of the Breast: A Case Report

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Abstract

Breast angiosarcomas are rare malignant tumors with poor prognosis. There were two types of presentation: primary angiosarcoma and secondary angiosarcoma. Secondary angiosarcoma occurs after radiation therapy, as part of conserving treatment of breast cancer (also associated with chronic lymphedema). We report the case of a 37-year-old woman with primary angiosarcoma. The clinical and radiographic features of breast angiosarcomas are reviewed, and the various treatment options reported in the literature are discussed.

Introduction

Primary breast angiosarcomas are very rare tumors, accounting for 0.04-1% of all malignant tumors of the breast (1). They are highly aggressive, with frequent early metastases. Diagnosis is often difficult due to their polymorphous clinical presentation.

Observation

We report the case of a 37-year-old female patient, with no notable pathological history, in whom autopalpation of the left breast objectified a nodule progressively increasing in volume. The patient underwent mammography + ultrasonography, which revealed a heterogeneous, hyperechoic, encapsulated, avascular Doppler mass in the upper left quadrant, with long axis parallel to the skin, non-attenuating, measuring 45 x 22 mm, suggestive of an ACR2 hamartoma. A biopsy was then taken, and the pathological report showed that the cells expressed CD34, CD31 and CD117, but not cytokeratin, D2-40, chromogranin, synaptophysin or CD30, hence the diagnosis of intermediate-grade angiosarcoma.

Thoracic-abdominal-pelvic CT: well-limited tissue mass of the left breast, with irregular contours, density and heterogeneous enhancement after contrast, measuring approximately 90 x 55 mm in diameter, coming into contact with the skin covering invading the areolar plate.

A few bilateral sub centimetric oval axillary lymph node formations, the largest measuring 6 mm in minor axis. Conclusion: Tissue mass of the left breast invading the areolar plate with no evidence of distant extension with bilateral infracentimetric axillary lymph node formations.

Our patient underwent a left mastectomy, and the pathology report showed an intermediate grade angiosarcoma (grade II) measuring 7 cm histologically. There were no vascular emboli or peri-neural engrainments. Paget's disease was absent, and the resection margins were healthy by more than 10 mm. The tumor proliferation index assessed by Ki67 was estimated at 30%.

Post-operative ultrasound of right beast : several homogeneous hyperechoic lesions, involving the different quadrants of the breast, well limited, with regular contours, not taking color Doppler encoding, compatible with lipomas, the largest measuring 22 mm long in the superior-external quadrant, classified ACR2.

The patient underwent adjuvant chemotherapy with 4 courses of MAI, followed by radiotherapy at a dose of 50 Gy in 25 fractions of 2 Gy per fraction over 38 days, with good locoregional control.

Ten months after the end of radiotherapy, the patient presented with diffuse infiltration of breast fat, involving the various quadrants of the breast against lateral in relation to mastitis of undetermined origin, a subcutaneous lesion in the lower-external quadrant of the breast, with a focus of mastosis at the lower quadrant junction, focal ductal ectasia at the upper quadrant junction with finely echogenic content; a biopsy was performed, revealing a histological and immunohistochemical appearance compatible with angiosarcoma, given the positivity of anti-CD34 and anti-CD31 antibodies. A right mastectomy was performed, the pathological findings of which were in favour of an intermediate grade 2 angiosarcoma measuring 11 cm histologically, with no vascular emboli or peri-nervous sheathing, and the resection limits were healthy by more than 10 mm. Patient proposed for subsequent adjuvant chemotherapy.

Discussion

Primary angiosarcoma accounts for 0.04% of all breast tumors and 8% of breast sarcomas (2-3). It was Schmidt et al. in 1887 who first suggested this histological type. Borman et al. described the first case in 1907 (4,5). It mainly affects women between the ages of 30 and 40. According to the literature, the diagnosis is made during pregnancy in 11.5% of cases (2); however, the hormonal influence remains debatable. The prognosis for this tumor is unfavorable, with a median survival time of 24 months and a five-year recurrence-free survival rate of 33% (6). Prognostic factors include the type, grade and histological size of the tumor, as well as involvement of the surgical transection slices (7). It must be differentiated from breast angiosarcoma induced by radiotherapy. The incidence rate of radiation-induced angiosarcoma is 0.5% of patients surviving more than five years after initial radiotherapy, and the prevalence is six cases per 100,000 under conservative treatment (4). They occur in older women (mean age 60) and have a lower metastatic potential (7%) (8). The average interval between irradiation and disease onset varies from three to 20 years (9), and the radiation dose

that can induce angiosarcoma ranges from 40 to 80 Gy (10). Clinically, the lesion always measures more than 2 cm (as in our patient) and presents as a painless nodule, sometimes with a pulsatile character. Moreover, Zellek et al. found in their series that tumor size correlated with ten-year recurrence-free survival, and that the size of the tumor was greater than 10 cm, the worse the prognosis (5). The surrounding skin may take on a purplish, reddish, or angiomatous appearance. The palpable nature of the tumor suggests a malignant vascular tumor. In 21% of cases, the involvement is bilateral, as in our patient (2). Metastases occur in 30% of cases, most frequently in the bone, lung, liver, and contralateral breast (5). Lymph node involvement is very rare (0-5% of cases) (8). On imaging, mammography may show a round, poly-lobed, sparse, homogeneous parenchymal opacity of large size, with blurred boundaries and no spicular extension or calcification. Ultrasound described a heterogeneous lesion, and Doppler showed continuous venous flow in the intratumoral venous structures. CT scans assess the degree of differenftiation, and MRI reveals T1 hyposignal and T2 hypersignal (12). Histologically, three grades are defined, which have been correlated with prognosis by Donnel et al. and Mérino et al. (7). Type I corresponds to a low-grade tumor without massive areas of necrosis, hemorrhage, or papillary formation. Type II corresponds to an intermediate-grade tumor with no necrosis or hemorrhage; foci of massive proliferation should be scattered throughout the tumor, covering less than 20% of the tumor surface. Type III corresponds to a high-grade tumor, with the least favorable prognosis.

In our case, the diagnosis was not made in the first instance, and in the literature, an erroneous initial histological diagnosis is found in 37% of cases (2). The standard treatment is total mastectomy (13). The quality of surgical excision margins is fundamental and is the main factor in predicting local recurrence. Some authors consider that a healthy margin of one centimeter is necessary; if this is not achieved, revision is systematically performed by mastectomy or enlargement of the exeresis (11). Axillary dissection is unnecessary unless there is a palpable node, as lymph node metastases are rare (13). Complementary radiotherapy is necessary in the case of lumpectomy; this involves irradiation of the whole breast, with additional treatment in the tumor bed (14). After mastectomy, it does not appear to reduce the rate of local recurrence (15). As far as drugs are concerned, anthracyclines, ifosfamide and dacarbazine have been shown to be effective in soft-tissue sarcomas in general (6). Targeted therapy, using anti-growth factor antibodies, is a promising therapeutic option.

Conclusion

Angiosarcomas of the breast are very rare tumors, occurring mainly in young women. Despite improved knowledge, the prognosis remains poor, even in the localized stage. R0 surgery, supplemented by adjuvant radiotherapy, is the standard treatment for localized disease.

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