Breast Angiosarcoma.: a Case and Literature Review

S.Belaazri1, FZ.Lamine2, T.Berrada2, N.Zeraidi2, A.Baidada2, A.Kharbach1
1Service Obstetrics Gynecology and Endocrinology M3 Maternity Souissi, Chu Ibn Sina, Rabat, Morocco
2 Service Obstetrics Gynecology and endoscopy M1 Maternity Souissi, Chu Ibn Sina, Rabat, Morocco

Corresponding Author: Sofiane Belaazri
sbelazri23@hotmail.com

ABSTRACT

The angiosarcoma of the breast is a tumor of endothelial origin. It is a rapidly growing tumor, with a diagnosis often difficult. Simple mastectomy is the reference treatment. The adjuvant treatment has not proven any evidence. The prognosis is disappointing. We report a case of a patient aged 53 years, operated for a breast angiosarcoma.

Keywords: breast cancer, angiosarcoma, mastectomy

1. INTRODUCTION

Angiosarcoma of the breast is a rare malignant tumor of endothelial origin(1). Its prevalence in malignant mammary tumors was 0.04% and in mammary sarcoma 8%(2). It is characterized by high malignancy and clinical and radiological presentation polymorphic source of error and diagnostic delay. We present a case of breast angiosarcoma in a 54-year-old woman. The aim of our work is to clarify the clinical features, radiological, therapeutic and prognosis of this tumor.

2. CASE

Patient aged 53, diabetic for 14 years on oral anti-diabetic, hypertensive for four years, menarche at age twelve, menopausal for a year, nulliparous. The onset of symptoms goes back six months before hospitalization, by self-examination of a nodule in the right breast, without other accompanying signs. The all moving in a conservation context of the general state. Clinical examination revealed the presence of a nodule 6 centimeters at the upper outer quadrant of the right breast, movable relative to the two shots, painless, no inflammatory signs in sight, the contralateral breast is unremarkable. The lymph nodes are free. A mammogram was performed, she revealed at the upper outer quadrant of the right breast: an inhomogeneous opacity with a small addition of water opacity tone facing (fig 1) and the presence of a discrete skin thickening with nipple retraction; presence at the upper outer quadrant of the left breast of a small opacity water tone blur limit by location. Breast ultrasound revealed the presence at the upper outer quadrant of the right breast: a tumor process of tissue heterogeneous echogenicity with irregular boundary speculated place by vascular Doppler measuring approximately 27 x 16mm with a presence in skin thickening and an interesting edema the entire upper outer quadrant (fig 2). The left breast was a small nodular lesion homogeneous hypoechoic well-limited oval measuring 10 x 6 mm (fig 3). The right breast classified BI-RADS 4; left breast classified BI-RADS 3. A biopsy of the right breast revealed the presence of a well-differentiated angiosarcoma. The patient received a single mastectomy without lymph node dissection, and three meeting post-operative radiotherapy with anapath has a safety margin of 2cm. The postoperative course was simple. The patient showed no recurrence or distant metastasis in a year of surveillance, then it was forgotten.

3. DISCUSSION

The angiosarcoma of the breast is also called hemangiosarcoma, hémangioendothéliosarcome, angioplasty or malignant hemangioma. These are malignant tumors made of endothelial cells that line the lumen of blood vessels. Angiosarcomas preferentially sit in skin tissue and subcutaneous of the head, limbs and the liver. The breast is among the most common sites. It represents 0.04% of malignant breast tumors and 8% of mammary sarcoma\(^{(1)}\). This is usually a tumor of the young woman, the maximum occurred between 30 and 45 years, with extremes of 14 to 82 years. Our patient age of 53 years.

According to some authors, several factors have been implicated in the genesis of angiosarcoma such as a history of trauma, previous breast surgery or radiation\(^{(3)}\). The hormone-dependent raised by the presence of estrogen receptors remains challenged\(^{(4)}\), no factor was found in our patient.

The clinical description of mammary angiosarcoma differs little from that of a conventional breast carcinoma. This is a painless mass, large up to 19cm, with rapid growth, the almost constant association with an abnormality of the skin over the form of a reddish, purplish or pulsatile sometimes associated with cutaneous ulcers\(^{(2)}\). Bilateral involvement is common. While node involvement is exceptional. Our patient had a nodule 6 centimeters without lymphadenopathy or other associated clinical signs.

The mammographic appearance is not very specific. The tumor appears as a rounded or lobed parenchymal opacity, very dense, homogeneous, of large and often blurred boundaries sometimes occupying the entire

---

mammary gland. Unlike carcinomas, no specular extension, and calcifications are exceptional. This is explained by the rapid development of the lesion, leaving no time to calcareous salt deposits from forming. Furthermore, shrinkage and thickening skin next to the tumor are rarely described in angiosarcoma of the breast\(^2\). Breast ultrasound finds a heterogeneous echogenicity lesion with liquidiennes beaches indicating the presence of necrotic-hemorrhagic area where the interests of the Doppler showing vascular spots\(^2\).

CT scans can show a mass that is enhanced dramatically after injection of the contrast agent and a partial homogenization for late time. In magnetic resonance imaging, angiosarcoma appears as a mass with a hypointense on T1 and T2W, with tubular periphery areas having a more intense signal on T2, suggesting the presence of blood vessels\(^2\).

The histological diagnosis of breast angiosarcoma is difficult; it may even be wrong. Thus, when the tumor is well-differentiated confusion with Benin angioma. Conversely, when the tumor is undifferentiated, by disregarding its vascular nature where the immunohistochemistry of interest using the endothelial cell markers\(^3\). The Donell et al. classification classifies angiosarcoma in grades three histprognostic\(^5\)(Table1):

**Table I: Classification histprognostic Donell et al. [5] Angiosarcoma of the breast**

<table>
<thead>
<tr>
<th>Aspect histologique</th>
<th>Grade 1 (bien différencié)</th>
<th>Grade 2 (moyennement différencié)</th>
<th>Grade 3 (peu et indifférencié)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lésion interessant le parenchyme mammaire</td>
<td>Présentes</td>
<td>Présentes</td>
<td>Présentes</td>
</tr>
<tr>
<td>Canaux vasculaires inter-anastomotiques</td>
<td>Présentes</td>
<td>Présentes</td>
<td>Présentes</td>
</tr>
<tr>
<td>Cellules endothéliales hyper-chromatiques</td>
<td>Présentes</td>
<td>Présentes</td>
<td>Présentes</td>
</tr>
<tr>
<td>Projections papillaires intraluminales en touffe</td>
<td>Absentes</td>
<td>Absentes ou minimes</td>
<td>Présentes</td>
</tr>
<tr>
<td>Foyers de cellules fusiformes</td>
<td>Absent</td>
<td>Absents ou minimes</td>
<td>Présents</td>
</tr>
<tr>
<td>Mitoses</td>
<td>Rares ou absentes</td>
<td>Présentes dans les surfaces papillaires</td>
<td>Nombreuses</td>
</tr>
<tr>
<td>Lacs sanguins</td>
<td>Absents</td>
<td>Absents</td>
<td>Présents</td>
</tr>
<tr>
<td>Présence de nécrose</td>
<td>Absence</td>
<td>Absence</td>
<td>Présence</td>
</tr>
</tbody>
</table>

Surgery is the unique treatment of angiosarcoma. Because of the infiltrating nature and multifocal, total mastectomy more or less extended is the indication of choice. A simple lumpectomy can be proposed for small tumors less than 3 cm, with a wide margin of safety than 1 cm. Some authors consider that a healthy margin of 1 cm is required; if it is not reached, a recovery is systematic mastectomy or enlargement of resection\(^6\). Axillary dissection is unnecessary, except in cases of a palpable node or advanced forms because lymph node metastases are rare.

Conservative treatment should be supplemented with external radiotherapy in the manner of conservative treatment for carcinoma: a 50 Gy in 25 sessions over the entire breast with an overlay on the tumor bed with external radiotherapy or brachytherapy in the case of residual tumor or of the high-grade tumor. Adjuvant irradiation of the chest wall after mastectomy is advocated by several authors. However, for others, it does not seem to prevent local recurrence\(^3\).

The role of chemotherapy in the treatment of breast sarcoma is not currently well established\(^7\).

Indeed, a meta-analysis supported the contribution of adjuvant chemotherapy with doxorubicin on improving the probability of survival without recurrence, however no significant effect on overall survival\(^8\). Following clinical trials, adjuvant chemotherapy is strongly recommended in breast sarcomas exceeding 5 cm and those of high histological grade and that by extrapolating the results observed in sarcomas of the extremities. Doxorubicin and/or ifosfamide are the most used agents in the treatment of primary breast sarcomas\(^8\).

A new treatment option may be offered by biological treatment tests using antibodies antifacteurs growth\(^9\). The operation torque of VEGF and its receptor VEGFR, main vascular growth factor, provides an ideal therapeutic target for inhibiting tumor angiogenesis, and that, using either anti-VEGF or soluble receptor monoclonal antibodies reducing the rate of circulating factor\(^10\).

Another biological target can be materialized by the fibroblast growth factor (FGF) and its receptor, the simultaneous action of VEGF and FGF may enhance
the antitumor activity\textsuperscript{(11)}. We must insist on the prognostic value of the degree of histological differentiation: the relapse-free survival rate at five years was 76% for grade I, 70% for grade II, 15% for grade III. The seniority of the symptoms is a good prognostic factor, while age is not a prognostic factor for the majority of authors. Genetically, there is great heterogeneity among tumors, but it seems that the aberrations of chromosomes 8q and 20p are most frequently found and are also worth exploring\textsuperscript{(12)}. Our patient has benefited from conservative treatment with radiotherapy.

The natural history of this tumor is to the rapid recurrence despite surgical excision. Spread is through blood. Metastases are especially pulmonary, liver, bone, brain, and the contralateral breast\textsuperscript{(1,2)}. Breast angiosarcoma is a poor prognostic tumor; median survival is two years.

4. CONCLUSION

The angiosarcoma of the breast is a rare tumor. Its diagnosis is difficult. The treatment of choice is the simple mastectomy. Radiotherapy and chemotherapy remain unproven. The prognosis is severe and depends the speed of the treatment.

REFERENCES