

Primary Mammary Angiosarcoma: A Case Report

Andaloussi SM^{*}, Benhessou M¹, Ennachit M¹ and Kerroumi ME¹

¹Department of Mohamed 6 Centre for the Treatment of Cancer, Ibn Rochd University Hospital Center, Casablanca, Morocco

*Corresponding author:

Meriem Serraj Andaloussi,
Department of Mohamed 6 Centre for the
Treatment of Cancer, Ibn Rochd University
Hospital Center, Casablanca, Morocco,
E-mail: serrajmeriam@gmail.com

Received: 27 Apr 2021
Accepted: 12 May 2021
Published: 17 May 2021

Copyright:

©2021 Andaloussi SM. This is an open access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and build upon your work non-commercially.

Keywords:

Angiosarcoma Breast; Radiology; Mastectomy; Radiotherapy

Citation:

Andaloussi SM, Primary Mammary Angiosarcoma: A Case Report. *Ame J Surg Clin Case Rep.* 2021; 3(3): 1-5

1. Abstract

Angio Sarcomas (AS) are rare tumors, accounting for about 1% of soft tissue sarcomas.

Mammary AS can be separated into two categories: primary AS, occurring in the breast parenchyma, and secondary AS, which usually develop in the skin, or sometimes in the chest wall or breast parenchyma following treatment. by surgery and radiotherapy for breast cancer. Both forms behave badly and have a poor prognosis. We report a case of angiosarcoma of the breast diagnosed by histology and the treatment consists of a mastectomy supplemented by chemotherapy and radiotherapy.

It is generally manifested by a polymorphic, often confusing clinical picture, which is a source of diagnostic delay.

RESUME

2. Introduction

Angiosarcoma of the breast is a very rare conjunctival tumor. It is a primary malignant proliferation of endothelial cells in the vascular tissue of the gland. Primary mammary SA is rare (0.05% of primary breast cancers), but it is the second most common malignant mesenchymal tumor. more common in the breast, after malignant phyllodes tumors [1]. It is generally manifested by a polymorphic, often confusing clinical picture, which is a source of diagnostic delay. It is characterized by a strong malignancy, the evolution of which is towards rapid recurrence and the appearance of visceral metastases. Its histological diagnosis is based on a rigorous analysis of the excision pieces [2].

Our goal, by reporting a case of angiosarcoma of the breast, is to highlight the rarity of this aggressive tumor, its difficulty of diagnosis and its therapeutic management.

3. Observation

Mrs TF, 32 years old, married, nulligeste, nulliparous, operated for a right ovarian cyst in 2009 (without documents) who consults for a nodule in the left breast, discovered on autopalpation, evolving for a year 3 months, gradually increasing in size and having received neo-adjuvant chemotherapy based on three courses and then sent to our structure for additional PEC. Clinical examination of the breasts found a 6 cm cup straddling the upper left quadrants without nipple retraction, nipple discharge or inflammatory signs (FIGURE 1). The right breast is without detectable abnormality. The ganglionic areas are free. According to the reference letter, at the initial examination before neoadjuvant chemotherapy, a mass a mass overlapping the upper quadrans of the left breast of 9cm was noted, with a pulsatile, angiomatic purplish appearance of the opposite skin.

The radiological examinations before chemotherapy were as follows: The mammography showed the presence at the level of the superointernal quadrant of the left breast a rounded nodule, whose edges appear perfectly clear, measuring 7 mm long axis, without micro calcifications or thickening of the skin opposite (FIGURE 2). On ultrasound, it is a hyperechoic placard, sitting at the union of the upper quadrants, comprising a homogeneous hypoechoic oval nodule with clear edges, with a long axis parallel to the skin planes, with a structure of 12 mm long axis (FIGURE 3). with bilateral axillary lymphadenopathy with a dedifferentiated appearance. The examination is classified BIRADS4. The magnetic resonance imaging showed a nodule of oval shape and irregular outlines in places straddling the Superior Quadrants(SQ), it is in homogeneous T1 hypo intense and T2 hyper signal. It enhances intensely, early and in homogeneously without rapid washout, with a

highly suspicious appearance, associated with architectural disorganization classified BIRADS4 of the ACR (FIGURE 4).

Ultrasound-guided micro biopsies show slightly atypical vascular proliferation with a fusocellular component compatible with angiosarcoma (intermediate grade) with a Ki 67: 40%. An extension assessment carried out is negative.

A left mastectomy was performed on April 21, 2017. The histological study of the part revealed the presence of a tumor of hemorrhagic and necrotic vascular appearance measuring 5X4cm, suggesting an intermediate grade angiosarcoma. complete resection (FIGURE 5 ,6). The post-chemotherapy response is estimated at 20%.



Figure 1: Clinical examination of the breasts found a 6 cm cup straddling the upper left quadrants without

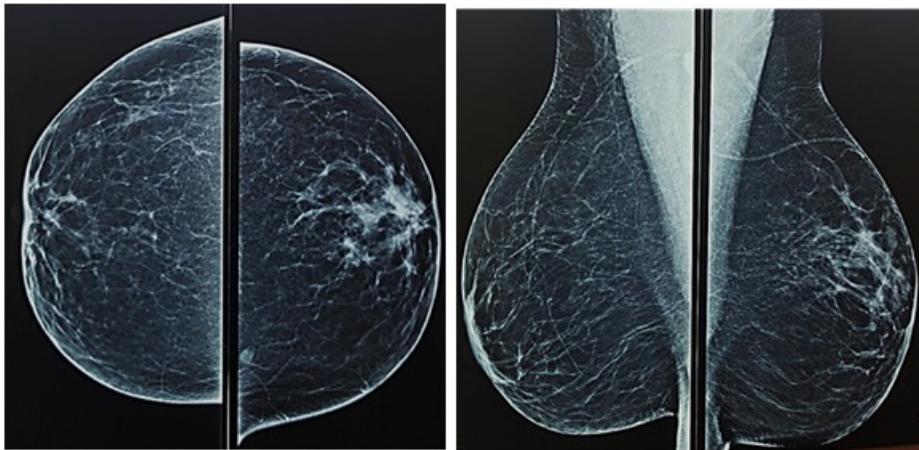


Figure 2: Presence at the level of the superointernal quadrant of the left breast a rounded nodule, whose edges appear perfectly clear, measuring 7 mm long axis



Figure 3: On ultrasound, it is a hyperechoic placard, sitting at the union of the upper quadrants, comprising a homogeneous hypoechoic oval nodule with clear edges, with a long axis parallel to the skin planes, with a structure of 12 mm long axis

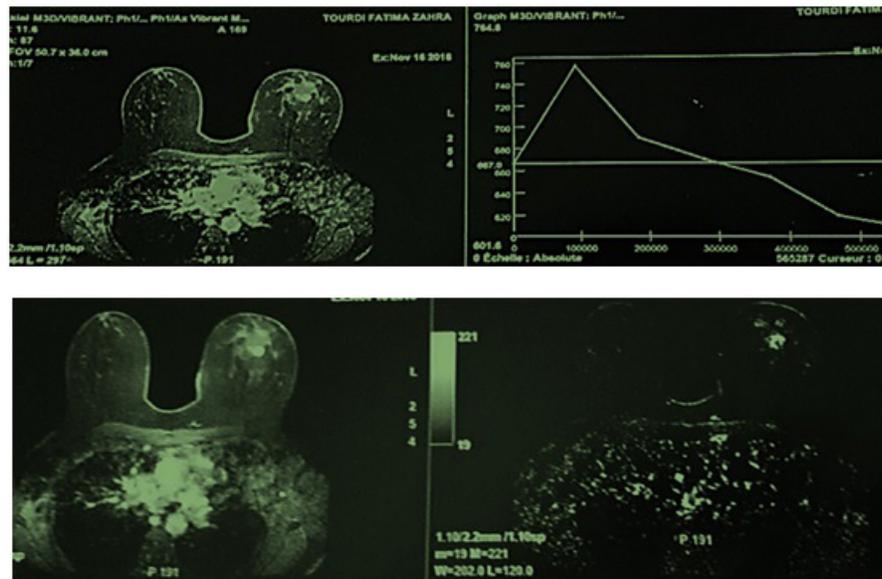
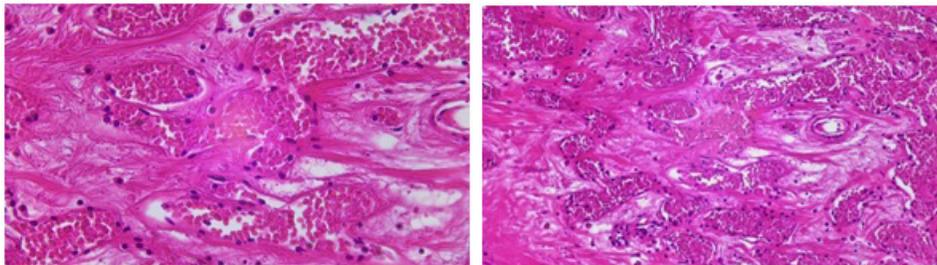


Figure 4 :The MRI showed a nodule of oval shape and irregular outlines in places straddling the SQ, it is in homogeneous T1 hypointense and T2 hypersignal. It enhances intensely, early and inhomogeneously without rapid washout classed BIRADS 4 of ACR



Figures 5,6: Histological features of breast angiosarcoma

The treatment is completed by adjuvant chemotherapy: three courses of (adriablastine: 50 mg / m², cyclophosphamide: 500 mg / m²) then radiotherapy at a dose of 50 Gy on the wall and the scar. Two years later, the patient was re hospitalized for dyspnea and chest pain, and the chest x-ray showed pulmonary metastases from the release of balloons. After this episode, the patient's general condition deteriorates rapidly.

She died two months later in a table of cachexia.

4. Discussion

These are malignant tumors made of endothelial cells that line the lumen of the blood vessels. Angiosarcomas are preferentially located in the skin and subcutaneous tissues of the head, limbs and in the liver. The breast represents 9% of all locations. Although the mammary gland is one of the most frequent sites of these tumors, angiosarcomas of the breast remain rare; they represent 0.04 to 1% of all malignant breast tumors [3].

It is a tumor which affects the young woman during period of genital activity as the case of our patient, especially between 30 and 40 years. In our observation, it was a 32-year-old woman. But, all age groups are concerned, from 13 to 85 years old. Man is exceptionally affected [4].

They typically present as a unilateral, rapidly growing breast mass

associated with mastodynia in 30% of cases [5]. Involvement of the left breast is much more frequent for Norris and Taylor, while for Gabriele et al., It is the right breast which is most affected [6].

The tumor size reported in the literature was often large and this has been linked by several authors to the neglect and the rapidity of the evolution of these tumors. In fact, the mean size of mammary sarcomas varied between 4.8 and 12.5 cm and can reach 41 cm in some series [5, 7]. Two signs are very suggestive of angiosarcoma and which are present in our case: the pulsatile nature of the tumor and the purplish, angiomatous or reddish appearance of the skin, opposite the tumor [8]. This characteristic was the case with our patient.

In addition to the clinical examination, the imaging data are non-specific or even misleading. Angiosarcoma of the breast presents as a rounded parenchymal opacity with clear contours and sometimes polylobed, dense, homogeneous, generally large in size. Usually, angiosarcoma of the breast is not accompanied by calcifications. This is explained by the rapid development of the lesion, leaving no time for calcareous salt deposits to form [8, 9].

Ultrasound finds a heterogeneous lesion of tissue echo structure with fluid areas indicating the presence of necrotic or hemorrhagic phenomena, which may lead wrongly to the diagnosis of mamma-

ry hematoma. Color Doppler shows the presence of intense vascularization characterized by numerous vascular spots [9].

The contribution of the scanner is important, it shows a mass which is enhanced in a very important way after injection of the product of contrast and a partial homogenization for the late stages. On Magnetic Resonance Imaging (MRI), angiosarcoma presents as a mass with hypo intense in T1 and hyper signal in T2 with tabular areas in the periphery with more intense hyper signal in T2, suggesting the presence of blood vessels [4,10].

Faced with the lack of specificity of imaging methods, only histology can confirm the diagnosis [9]. However, he faced diagnostic difficulties, especially on biopsies. He must be carried over the entire tumor. Under certain conditions, the extemporaneous examination constitutes one of the pillars of the diagnostic workup, especially when there is a discrepancy between clinical practice, radiology and cytology. Its interest is to confirm or rule out the malignancy and to guide the extent of the surgical procedure. False positive diagnoses are exceptional and false negatives result in a second surgery [5]. Macroscopically, the tumor varies in size between 1 and 20 cm (average 5 cm). It is poorly defined, spongy when cut and hemorrhagic. Histologically, tumor proliferation infiltrates the lobular stroma and surrounding tissues. It is made up of irregular anastomotic vascular cavities, lined with one or more layers of endothelial cells. Mammary angiosarcomas are graded [4]. three grades are defined, which were correlated with prognosis by Donnel et al. and Mérino et al. [11, 12]. Type I corresponds to a low-grade tumor without a massive area of necrosis, hemorrhage or papillary formation. Type II corresponds to an intermediate grade tumor without necrosis or hemorrhage; the foci of massive proliferation should be dispersed within the tumor and less than 20% of the tumor surface. Type III corresponds to a high grade tumor and has the least favorable prognosis.

Differential diagnoses of well-differentiated forms of mammary AS include post-radiation Atypical Vascular Lesions (AVL), PASH (Pseudoangiomatous Stromal Hyperplasia), angioliipoma, benign vascular lesions (hemangiomas) and papillary endothelial hyperplasia (Masson's tumor). The early forms of Kaposi's sarcoma are also part of the differential diagnosis of well-differentiated angiosarcoma, as are angiomas and solitary fibrous tumors, which are very rare in the breast. Poorly differentiated forms of angiosarcoma can be mistaken for carcinoma, melanoma or other types of sarcoma [1]. The diagnostic difficulty arises for the pathologist at both ends of the spectrum. Grade I angiosarcoma can be mistaken for a hemangioma. A grade III angiosarcoma poses the problem of differential diagnosis with poorly differentiated tumors. Immunohistochemistry is of great help by confirming the vascular nature of proliferation thanks to the CD31, CD34 and factor VIII antibodies, and the negativity of cytokeratin. However, these vascular markers may be lacking in poorly differentiated forms [4, 13].

Surgery is the cornerstone of treatment for primary mammary sarcomas [14]. However, its modalities are controversial [15]. In fact, simple mastectomy is considered the “gold standard” for primary mammary sarcomas [5].

Adjuvant radiotherapy is often associated with surgery. There is no proof of efficacy of adjuvant chemotherapy in localized forms after surgery and radiotherapy. For metastatic forms, chemotherapy treatment is generally offered. Taxanes have interesting anti-angiogenic activity [1, 16].

The majority of authors report an unfavorable prognosis for angiosarcomas [5].

The mean recurrence-free survival is less than 3 years and total survival less than 6 years [1]. Metastases are frequent; The dissemination is done strictly by blood; and consequently, metastases are localized first in the liver and the lung then in the whole organism with predilection for the subcutaneous tissue, the bone, the brain, without forgetting the contralateral breast [12]. In the literature, the rate of loco regional relapses varied from 12 to 73% with an average of 20% [5].

Prognostic factors for angiosarcoma include age (worse prognosis in elderly patients), size, tumor extension and the status of the excision margins [16].

5. Conclusion

Angiosarcoma of the breast is a rare vascular tumor with a formidable prognosis. His diagnosis must be

evoked in front of a swelling of the breast with rapid growth and which results in an often benign mammographic aspect. Histological diagnosis is often difficult, and it is wrong in 37% of cases. Only early surgery can hope for longer survival. New molecules, in particular targeted therapies, are therefore eagerly awaited, mainly in metastatic forms.

References

1. Russ EL. “Difficultés diagnostiques sur biopsies mammaires. Cas no 4: carcinome papillaire infiltrant.” *Annales de Pathologie*. 2017; 37.
2. Thériault M, Lalonde L, David J, Gaboury L, Trop I. Angiosarcome primitif du sein. *Image Femme* 2007;17: 189—95.
3. Berroho K, Iraqi M, Achiq A, Chraibi C, Alami MH, Fillali A, et al. “Angiosarcome du sein: à propos de deux cas.” *Journal Africain du Cancer/African Journal of Cancer*. 2011; 3: 199-203.
4. Boufettal H, Noun M, Samouh N, Hermas S, Zamiaty S, Benayad S, et al. “Angiosarcome mammaire: à propos d’un cas.” *Annales de pathologie*. 2013; 33: 217-220.
5. Lahat G, Lev D, Gerstenhaber F, Madewell J, Le-Petross H, Pollock RE. Sarcomas of the breast. *Expert Rev Anticancer Ther*. 2012; 12: 1045–51.
6. Gabriele R, Borghese M, Corigliano N, Barbaro M, Conte M. [Phylolodes tumor of the breast. Personal contribution of 21 cases]. *G Chir*. 2000; 21: 453–6.

7. Al-Benna S, Poggemann K, Steinau HU, Steinstraesser L. Diagnosis and management of primary breast sarcoma. *Breast Cancer Res Treat.* 2010; 122: 619–26.
8. Elhadj OEA, Nasri M, Thabet S, Goucha A, Rahal K, Elmay A, et al. “Sarcomes mammaires primitifs: à propos de 30 cas traités à l’institut Salah-Azaiez de Tunis.” *Cancer/Radiothérapie.* 2017; 21: 45-50.
9. Rachidi, I., et al. “Angiosarcome mammaire: A propos d’un cas/ Mammary angiosarcoma: About one case.” *International Journal of Innovation and Applied Studies* 20.2 (2017): 589.
10. Elwady N, Ousehal A, Alibou F, Tazi Z, Boukind H, Kadiri R. Angiosarcome du sein. Aspects radiologiques. À propos d’un cas. *J Radiol* 2000;81:50—3.
11. Champeaux-Orange E, Bonneau C, Raharimanana B, Favre A, Ibrahim M, Breteau N. Angiosarcome mammaire primitive: à propos de deux cas: primary breast angiosarcoma: Two cases report. *Cancer/Radiothérapie.* 2009; 13:209–12.
12. Bennani A, Chbani L, Lamchahab M, Wahbi M, Amarti A, Melhouf MA, et al. Primary angiosarcoma of the breast: a case report. *Diagnostic pathology.* 2013; 8.
13. Chen KT, Kirkegaard DD, Bocian JJ. Angiosarcome primitive du sein chez l’homme. *Ann Pathol* 2005;25(3):235—9.
14. Toesca A, Spitaleri G, De Pas T, Botteri E, Gentilini O, Bottiglieri L, et al. Sarcoma of the breast: outcome and reconstructive options. *Clin Breast Cancer.* 2012; 12 :438–44.
15. Wang F, Jia Y, Tong Z. Comparison of the clinical prognostic and features between the primary breast sarcomas and malignant phyllodes tumor. *Jpn J ClinOncol.* 2015; 45: 146–52.
16. Weiss S, Goldblum J, Folpe AL. *Enzinger and Weiss’s soft tissue tumors.* Elsevier Health Sciences. 2007; 1304.