International Journal of Academic Health and Medical Research (IJAHMR)
ISSN: 2643-9824
Vol. 5 Issue 12, December - 2021, Pages:30-32

Historical Size of Breast Sarcoma: About 2 Cases and Review of the Literature.

I.Alilou; M.Alami; K. Saoud; N. Mamouni; S. Errarhay; C. Bouchikhi; A. Banani.

Corresponding author: Dr Alilou Ismail**,** dralilouismail@yahoo.com,

Service de gynécologie et obstétrique 1, CHU HASSAN II FES, MAROC.

Abstract*: Breast sarcomas are rare malignant tumors of the breast. There are many histological subtypes of variable prognosis, the most aggressive of which is angiosarcoma. The reason for consultation is often related to the discovery of a breast nodule. The positive diagnosis is based on anatomopathological study. It is a cancer with little lymphophilia, the treatment is based on surgery such as mastectomy and rarely conservative treatment without lymph node dissection. Radiotherapy is often indicated, while chemotherapy remains of limited indication. The prognosis depends on tumor size, histological subtype, grading and resection margins.*

Keywords: breast sarcoma, diagnosis, surgery, radiotherapy, prognosis**.**

1. Introduction:

Breast sarcomas are very rare tumors. They represent less than 1% of breast tumors [1] [2]. They are part of the non-epithelial tumors of the breast. The most frequent form is histiofibtocytoma [2] [4]. These tumors have a variable prognosis depending mainly on the histological subtype, grade and tumor size. Rarely spontaneous, they are often secondary to breast irradiation. Treatment is based on surgery such as mastectomy, axillary dissection is rarely indicated due to the low metastatic potential of the lymph nodes. It is often completed by breast irradiation. Chemotherapy remains questionable in forms with a high risk of relapse.

In our work we present two cases of breast sarcoma of exceptional size exceeding 40cm of large diameter, discovered in two patients with a delay of consultation. We detail our management of these two patients, with a review of the literature.

2. Medical observation:

Case 1: Mrs. A. K., 45 years old, type II diabetic on oral antidiabetics, with a history of an appendectomy 20 years ago, with no other associated defects or other particular history, in particular no notion of irradiation. She consulted us for the management of a right breast mass that had been evolving for 1 year and was progressively increasing in volume with surface ulcerations and deformation of the entire right breast. The clinical examination revealed an altered patient with an enormous mass of 40 cm taking the whole right breast with a polylobed aspect with very important collateral venous vascularization, and presence of several ulcerated zones bleeding easily on the surface. The mobilization of the mass is not possible in relation to the thorax. Breast ultrasound and mammography revealed a right breast classified as ACR5 of the BIRADS system without axillary adenopathies. A microbiopsy of the mass showed fuso-cellular proliferation in favor of either a sarcoma or a high-grade phyllodes tumor. The extension workup, consisting of a thoracic-abdominal-pelvic CT scan and a bone scan, did not reveal any secondary localizations.

Case 2: Mrs. H. F., 53 years old, hypertensive and on oral antihypertensive medication, with no other particular history, notably no notion of irradiation. She consulted for a progressive increase of a right breast mass discovered 1 year ago. The clinical examination revealed a huge mass taking the whole right breast which is deformed and fixed to the thorax measuring 42 cm hard with a rich collateral venous circulation. Echo-mammography performed as part of the locoregional extension workup did not find any axillary adenopathies. A microbiopsy of the mass was done in favor of a breast sarcoma and the general extension workup done by a thoraco-abdomino-pelvic CT scan and a bone scan found a heterogeneous aspect of the pectoralis major muscle without frank invasion and without secondary localizations.

Both patients underwent mastectomy with removal of the pectoralis major muscle. The final anatomopathological results found healthy margins. The patients were subsequently referred for adjuvant radiotherapy. The 6-month follow-up did not detect any locoregional or general recurrence.

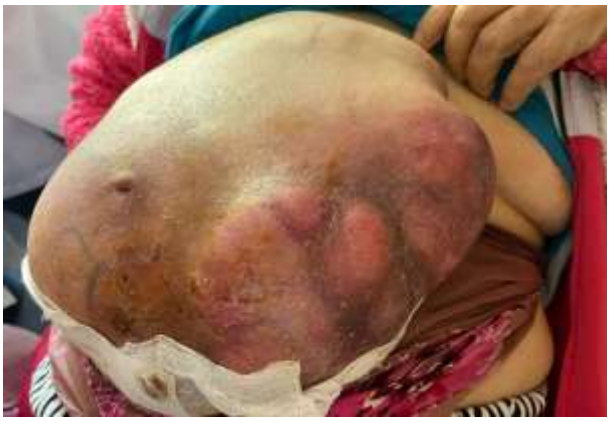

Figure 1: case 1: right breast mass with collateral venous circulation and ulceration

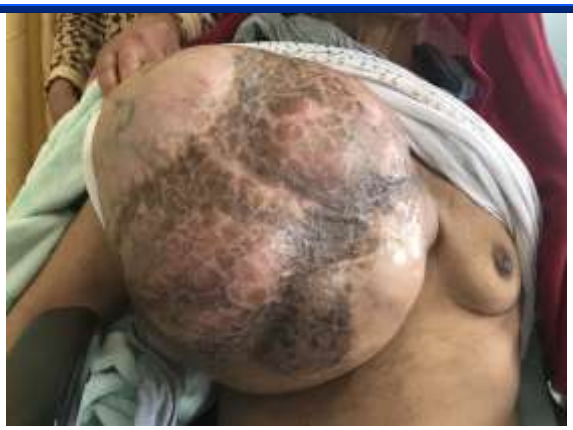

Figure 2 : case 2: mass taking the whole right breast with collateral venous circulation

3. **Discussion**:

Breast sarcoma is an almost exclusively female tumor. Its frequency is rare compared to other breast tumors and does not exceed 1% [1] [2]. The average age described in various studies is around 45 years [1][3][4]. The frequency of its location is similar between the two breasts, right and left. The most frequent reason for consultation is the discovery of a mass that increases rapidly in size associated with mastodynia [4][5]. The average size of discovery according to the series varies from 4.5 cm to 12.5 cm. In our cases[6][7], the enormous tumor size is related to neglect. This tumor size influences the 5-year survival when its size exceeds 5cm. It decreases from 83% for tumors smaller than 5cm to 42% for tumors larger than 5cm[9][11].
Skin changes such as ulceration or edema are sometimes found. Palpation of an axillary adenopathy is exceptional, as breast sarcoma is a very non lymphophilic tumor.

From an anatomical point of view, several subtypes are present with a prognosis that varies with the subtype. Mostly, we find unclassified sarcomas which correspond to the subtype found in our two patients. More rarely, angiosarcomas, fibrosarcomas, liposarcomas, liemyosarcomas and exceptionally stromal sarcomas, osteosarcomas, dermatofibrosarcomas protuberans (DFSP), alveolar sarcomas of the soft parts and rhabdomyosarcomas can be found. Particular attention is given to angiosarcoma, which is often radiation-induced, due to its poor prognosis and rapid pulmonary metastasis [5][8].
Tumor grading is an important prognostic factor to determine. It takes into consideration cyto-nuclear differentiation, number of mitosis and necrosis. Angiosarcomas are often grade 3.

The radiological workup done by a breast ultrasound associated with a mammogram does not find any specific sign. Spiculated calcifications with a mass with cystic content are very suspicious. On the other hand, breast MRI shows specific signs in the form of hypointense tumors on T1-weighted sequences, hyperintense on T2 with early and prolonged contrast after injection of gadolinium. In addition, for low-grade forms, the presence of hypointense, non-contrasting areas of fat on T2-weighted sequences with fat suppression seems to be characteristic[15].

Surgery is the cornerstone in the management of breast sarcoma. The basic procedure is a mastectomy without axillary waxing. This is the procedure performed for our two patients. Conservative treatment increases the risk of recurrence according to Berg et al. Healthy margins are a key factor influencing overall survival and recurrence-free survival. However, conservative treatment can be proposed for tumors less than 5 cm in size with healthy margins of more than 1 cm [9] [12] [13].
Radiation therapy is the second weapon in the management of breast sarcoma and provides better local control of the disease, but does not improve overall survival. It is indicated for tumors larger than 5 cm for high-grade tumors, in case of conservative treatment or if the healthy margins are less than 1 cm. The usual dose is 60 Gy. Our two patients underwent adjuvant radiotherapy for tumor size. The role of chemotherapy in breast sarcoma remains a subject of debate. Its place is not well established. It is indicated in metastatic forms, high-grade tumors, and tumors larger than 5 cm [10].

The prognosis of these tumors is variable and depends on tumor size, histological subtype, histological grade and margins of surgical removal [8] [11].
In the literature, the risk of local recurrence varies from 12 to 73%. The main metastatic sites are the lung and the bone. The overall survival at 5 years is very variable from one study to another and ranges from 91% to 66% [6].

4. **Conclusion**:

Mammary sarcoma remains a rare entity. The prognosis is variable. Therefore, the realization of large-scale studies is difficult. The prognosis is based on clinical and histological elements. Surgery is the gold standard in management. It is often associated with radiotherapy, which provides better local control without improving overall survival. The indication for chemotherapy remains controversial. Trials currently being done on the place of immunomodulators may have very interesting results in the future.

5. **Bibliography**:

[1] Lakhani S.R., Ellis I.O., Schnitt S.J., Tan P.H., Van De Vijver M.J.: WHO classification of tumours of the breast.4th ed.2012.IARC PressLyon

[2] Pollard S.G., Marks P.V., Temple L.N., Thompson H.H.: Breast sarcoma. A clinicopathological review of 25 cases. Cancer 1990; 66: pp. 941-944.

[3] Zelek L., Llombart-Cussac A., Terrier P., Pivot X., Guinebretiere J.M., Le Pechoux C., et. al.: Prognostic factors in primary breast sarcomas: a series of patients with long-term follow-up. J Clin Oncol 2003; 21: pp. 2583-2588.

[4] Adem C., Reynolds C., Ingle J.N., Nascimento A.G.: Primary breast sarcoma: clinicopathologic series from the Mayo Clinic and review of the literature. Br J Cancer. 2004; 91: pp. 237-241.

[5] Coindre J.M., Terrier P., Guillou L., Le Doussal V., Collin F., Ranchère D., et. al.: Predictive value of grade for metastasis development in the main histologic types of adult soft tissue sarcomas: a study of 1,240 patients from the French Federation of Cancer Centers Sarcoma Group. Cancer 2001; 91: pp. 1914-1926.

[6] Lahat G., Lev D., Gerstenhaber F., Madewell J., Le-Petross H., Pollock R.E.: Sarcomas of the breast. Expert Rev Anticancer Ther. 2012; 12: pp. 1045-1051.

[7] Fields R.C., Aft R.L., Gillanders W.E., Eberlein T.J., Margenthaler J.A.: Treatment and outcomes of patients with primary breast sarcoma. Am J Surg. 2008; 196: pp. 559-561.

[8] Trojani M., Contesso G., Coindre J.M., Rouesse J., Bui N.B., De Mascarel A., et. al.: Soft-tissue sarcomas of adults; study of pathological prognostic variables and definition of a histopathological grading system. Int J Cancer 1984; 33: pp. 37-42.

[9] Monroe A.T., Fiegenberg S., Mendenhall N.: Angiosarcoma after breast conserving therapy. Cancer 2003; 97: pp. 1832-1840.

[10] Penel N., Bui Nguyen B., Bay J.O., Cupissol D., Ray-Coquard I., Piperno-Neumann S., et. al.: Weekly paclitaxel in metastatic angiosarcoma. A FNCLCC French sarcoma group (GSF-GETO) phase II trial. (ASCO Annual Meeting Proceedings). J Clin Oncol 2007; 25: abs 10002

[11] Bousquet G., Confavreux C., Magne N., de Lara C.T., Poortmans P., Senkus E., et. al.: Outcome and prognostic factors in breast sarcoma: a multicenter study from the rare cancer network. Radiother Oncol. 2007; 85: pp. 355-361.

[12] Rosenberg S.A., Tepper J., Glatstein E., Costa J., Baker A., Brennan , et. al.: The treatment of soft-tissue sarcomas of the extremities: prospective randomized evaluations of (1) limb-sparing surgery plus radiation therapy compared with amputation and (2) the role of adjuvant chemotherapy. Ann Surg 1982; 196: pp. 305-315.

[13] Penel N., Bui Nguyen B., Bay J.O., Cupissol D., Ray-Coquard I., Piperno-Neumann S., et. al.: Weekly paclitaxel in metastatic angiosarcoma. A FNCLCC French sarcoma group (GSF-GETO) phase II trial. (ASCO Annual Meeting Proceedings). J Clin Oncol 2007; 25: abs 10002

[14] Johnstone P.A., Pierce L.J., Merino M.J., Yang J.C., Epstein A.H., DeLaney T.F.: Primary soft tissue sarcomas of the breast: locoregional control with postoperative radiotherapy. Int J Radiat Oncol Biol Phys 1993; 27: pp. 671-675.

[15] Kikawa Y., Konishi Y., Nakamoto Y., Harada T., Takeo M., Ogata M., et. al.: Angiosarcoma of the breast-specific findings of MRI. Breast Cancer 2006; 13: pp. 369-373.