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Case Report

Rare Malignancy of the Breast: Primary Angiosarcoma, About a Case and Literature Review

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Abstract

Breast angiosarcomas are rare mesenchymal tumors of vascular endothelial origin, they are most often radiation-induced, more rarely primary; and characterized by a particular clinical and radiological presentation, local recurrence has a poor prognosis.

We will report the case of a young patient, who presented with primary angiosarcoma of the breast, treated with conservative surgery, then revision surgery for early local recurrence, followed radiotherapy, and in whom we encountered diagnostic problems on the radiological and pathological level. We will discuss through this observation the clinical, radiological and pathological peculiarities of primary angiosarcomas of the breast.

Keywords: Breast Angiosarcoma; Breast MRI; Surgical Treatment

Introduction

Mammary sarcomas are extremely rare malignancies, accounting for less than 1% of breast tumour cases and less than 5% of sarcomas [1]. Their incidence is estimated at less than 50 cases per 10 million women. There is no consensus on the exact definition of breast sarcoma. For some, phyllodes sarcomas are excluded because of their epithelial component, while for others, the distinction is not necessary given the similar prognosis and management [2].

Mammary angiosarcomas represent 0.04 to 1% of all malignant breast tumors [3] and occur either spontaneously in young women under 40 years of age, they are then called «primary», or after chest radiotherapy, in older patients [4].

The diagnosis of certainty is histological, requiring further immunohistochemical studies to confirm the vascular character of angiosarcomas that express endothelial markers, namely CD31 and CD43 [5].

We report the observation of primary angiosarcoma in a 32-year-old patient. Through the latter we will discuss the epidemiological, diagnostic, and therapeutic aspects of these rare tumors.

Observation

This is a 32-year-old patient, nulligeste, without particular history, who consults for a swelling of the left breast evolving for a year. The clinical examination finds a nodule at the junction of the internal quadrants, 06 cm in diameter, painless, mobile in relation to the deep plane and the skin, without orange peel or inflammatory signs. Mammography found an opacity of very low tone of blurred contour measuring 66.8mm of major axis, sitting at the level of the internal quadrants of the left breast; The ultrasound complement of the breasts finds a heterogeneous echoic structure of fuzzy contour of the internal quadrants. The mass is classified ACR 3, the axillary lymph node areas were free.

A microbiopsy of the nodule was performed, the histological study found a benign mesenchymal proliferation made of regular adipose tissue and blood capillaries and concluded angiolipoma. The patient was operated three months later she benefited from a lumpectomy, the histological study concluded once again to an angiolipoma with imprecise excision limits. Six months later the patient consults again because the mass has reappeared and has increased very quickly in volume. The clinical examination found a mass of 05 cm sitting in the internal quadrants. A breast MRI was made and objectified a left breast mass of the internal quadrants of 64mm, presenting in hypersignal stir in T2, hyposignal T1, gradually rising centripetally essentially at its lower pole, revealing fatty areas, without signs of infiltration of the pectoral muscle, this mass is vascular evoking angiolipoma or angioma, classified ACR 3 (Figure 1).

Figure 1: Bilateral breast MRI in axial sections, T2 (a) and T1 fat-sat weighted sequences after injection of gadolinium chelates at an early time before subtraction (b).

We decide to operate on the patient, and perform a zonectomy, the histopathological study of the objective operating room a breast parenchyma seat of a neoplastic cell proliferation of mesenchymal nature infiltrating containing many anastomosed blood vessels of different sizes, irregular and bordered by atypical cells arranged in several rows to form tufts and papillae. The cells are pleomorphic with marked cytonuclear atypia and a moderate mitotic index, in places the cells are fusiform. The histological study also finds the presence of blood extravasation with large focus of hemorrhagic suffusions, and an absence of necrosis zone (Figure 2).

Figure 2: Histological section of the zonectomy piece: (a) low magnification showing cell proliferation of vascular nature with foci of blood extravasation and fibrin deposits. (b) high magnification showing elongated, moderately atypical ovoid tumor cells with images of mitosis (arrows).

An immunohistochemical study was performed and shows tumor cell positivity to CD 31 and D 34 (Figure 3). The diagnosis of intermediate-grade angiosarcoma of the breast was therefore retained.

Figure 3: Immunohistochemistry images: (a) IHC X 40(CD31): CD31 vascular tumor cell positivity: breast glandular structure arrow. (b): IHC X 40(CD34): CD34 vascular tumor cell positivity.

The patient was lost to follow-up, and consults again after six months for a local recurrence, she was reoperated and this time we proceed to a mastectomy. The histopathological study finds an intermediate grade angiosarcoma of the breast with healthy surgical boundaries, located 1.5 mm from the deep plane.

The patient received adjuvant radiotherapy, 3D conformational type, at a dose of 50 Gray with conventional fractionation/ spreading, namely 02 Gray per session, 05 sessions per week. The target volume was the chest wall with 3D margins of 03 mm.

Discussion

Mammary sarcomas are rare malignancies, these tumours include grade I and II phyllodes sarcomas and other entities that form a group encompassing grade III phyllodes sarcomas and angiosarcomas. The relapse-free survival of patients at 3 years reaches 57% for grade I and II phyllodes tumors, 45% for grade III phyllodes sarcomas and 7% for angiosarcomas [6].

Angiosarcomas account for 0.04% of all mammary malignancies and 8-10% of mammary sarcomas. They occur in women aged between 30 and 50 years, But, all age groups are concerned, from 13 to 85 years [3].

It is therefore either a tumor of the young woman occurring spontaneously, without obvious triggering factor, or a tumor of the older woman (average age of 69 years) occurring after irradiation as part of a conservative treatment of breast cancer [7]. These radio-induced sarcomas occur 5 to 10 years (median at 6 years) after 50 Grays irradiation.

Clinically the mode of revelation is almost always the appearance of a breast nodule that rapidly increases in volume, as is the case of our patient. Indeed, the size of the tumor is often large with a size between 2 and 11 cm, with an average of 5.3 cm [8]. The tumor can also be revealed by the sudden occurrence of a hematoma. The lymph node involvement is exceptional with this type of lesion.

Two signs are suggestive of breast angiosarcoma: the pulsatile nature of the tumor found in some cases and the angiomatous purplish appearance of the skin in relation to the tumor. These characteristics have always been absent in our patient.

The mammographic aspect is not very specific and can even be misleading simulating a benign lesion, which was the case of our patient; The tumor presents as a rounded or polylobed opacity, without calcifications, not very dense, homogeneous, of large size, often well circumscribed, there is sometimes a focal asymmetry of density [9]. In young women with dense breasts, the tumor is often difficult to see on mammography. On ultrasound, it can appear as a tissue mass (hyperechogenic, hypoechoic or mixed), heterogeneous with fluid ranges related to necrotic and hemorrhagic phenomena [10]. It can also be a mixed ultrasound range (hyper- and hypoechogenic) without real mass. The color Doppler study shows intense hypervascularization [9]. Unlike carcinomas, there is no spicular prolongation and calcifications are often exceptional.

MRI finds a heterogeneous mass in hyposignal T1 and hypersignal T2, sometimes with areas in spontaneous T1 hypersignal corresponding to hemorrhagic areas or venous lakes [11]. MRI can also assess subcutaneous infiltration and visualize large drainage vessels [11]. This examination has a crucial role for the assessment of local extension (in particular the infiltration of the deep muscular planes). It should also be noted that cases of secondary angiosarcomas occurring after radiotherapy are characterized by focis within the skin thickened following irradiation.

Histology is the only test that makes it possible to make the diagnosis of certainty. The term angiosarcoma refers to all malignant tumours whose cells manifest the morphological and functional properties of the normal endothelium. Macroscopically, angiosarcomas are infiltrative, poorly limited tumors with hemorrhagic areas. Microscopically, their differentiation is highly variable.

Distinction with atypical post-radiation vascular lesions (in the context of a history of radiotherapy) or even with benign lesions (hemangioma or angiolipoma) can be difficult in some cases. In the case of our patient, two histological readings concluded that a benign tumor was benign, which does not correspond to the evolution of the disease.

Some authors report that the FNCLCC histological grade for soft tissue sarcomas (Figure 4) is usable for primary breast sarcomas [12]. However, this grade is not routinely applied for primary breast angiosarcomas but rather a histological classification, specific this time to breast angiosarcomas, which is that of Donnell., *et al*. [10].

Immunohistochemistry is essential to confirm the vascular nature of the lesion. Angiosarcomas typically express endothelial markers:

Figure 4: Grading by the FNCLCC.

CD31 (most sensitive and specific marker), CD34, factor VIII, Ulex europaeus agglutinin 1 and vascular endothelial growth factor (VEGF) [13].

The cornerstone of the treatment of breast angiosarcomas is surgery, which remains the only curative treatment, as with all sarcomas, it consists of a wide excision with R0 resection. This surgery may consist of conservative treatment or a radical mastectomy. A simple lumpectomy may be proposed for small tumors smaller than 3 cm.

For some, the margins should be greater than 1 cm, while for others, 2 to 4 cm are necessary, especially in the case of sarcoma after radiation therapy [14]. Salvage mastectomy remains possible in case of local recurrence after conservative treatment, especially of a small tumor less than 5 cm and low grade [15].

Carcinological limitations condition local recurrence. Axillary lymph node dissection is not recommended given the low probability of lymph node extension of these tumours [7,16].

There is no room for adjuvant chemotherapy therapy, however the interest of neoadjuvant chemotherapy (improved resectability) and adjuvant radiotherapy (non-radiation-induced angiosarcomas) has been suggested [13] and should be discussed in a multidisciplinary consultation meeting on a case-by-case basis. Clinical trials are difficult to implement given the rarity of these tumors, however new therapeutic avenues have been studied, using in particular targeted anti-angiogenic therapies [8] but without success. Their management is therefore modelled on that of soft tissue sarcomas, taking into account prognostic factors namely margin status, histological grade and tumor size.

Breast angiosarcomas are tumors with a very poor prognosis, most series report an average overall survival of 18 to 36 months [17]. Local recurrence is frequent, mainly related to the quality of the initial excision, which was the case of our patient who recurred after six months twice because the limits of excisions were infiltrated. These recurrences are quickly followed by the appearance of distant metastases by hematogenous route (in order of frequency: lung, skin, subcutaneous tissue, liver, bone, contralateral breast, central nervous system, spleen, omentum, adrenal, muscles, ovaries [18]).

Conclusion

Breast angiosarcoma is an extremely rare tumour with a very poor prognosis. It can be primary affecting young women or secondary typically affecting older women treated with radiotherapy. Clinically, these are most often large tumors with frequent skin abnormalities. Imaging is not specific and often misleading, however MRI has an important place for the locoregional extension assessment, and must be performed before any surgical procedure. Only a histological study of the operating room supplemented by an immunohistochemical study makes it possible to have a diagnosis of certainty. This diagnosis is rarely made before the surgical procedure because it is difficult on microbiopsy. Surgery is the only curative treatment and must imperatively be R0, the status of the margins being the essential prognostic factor, which conditions the occurrence of local recurrence. Only early and complete surgery can hope for longer survivals.

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