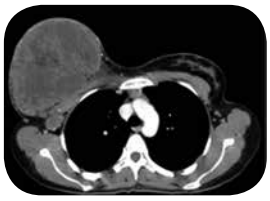

Angiosarcoma of the Breast: Report of Two Cases

Angiosarcoma de la mama: Presentación de dos casos

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Summary

Angiosarcomas are rare tumors that make up less than 1 % of breast cancers. These tumors may develop in a primary or secondary form, and 219 cases have been reported in the literature since the first case described by Schmidt in 1887. Here we present two cases of primary angiosarcoma of the breast and its main features in different imaging modalities.

Key words (MeSH)

Breast neoplasm
Magnetic resonance imaging
Biopsy

Resumen

Los angiosarcomas de la mama son tumores raros que conforman menos del 1 % de las neoplasias mamarias. Estos tumores pueden desarrollarse de forma primaria o secundaria y se han descrito 219 casos en la literatura mundial desde el primer caso informado por Schmidt en 1887. A continuación se presentan dos casos de angiosarcoma primario de la mama y sus principales características en los diferentes métodos diagnósticos.

Palabras clave (DeCS)

Neoplasias de la mama
Imagen por resonancia magnética
Biopsia

Case 1

Patient of 50 years with mass of 2 years of evolution in the right breast with progressive growth; Ultrasound and mammographic evaluation was performed. Mammography showed global glandular asymmetry, with increased right breast density associated with multiple nodular images, the largest of them in the right retroareolar region, BI-RADS category 0 (Figure 1). Subsequently, an ultrasound was performed with a finding of increased echogenicity of breast tissue, thickening of the skin and subcutaneous cellular tissue towards the medial quadrants of the right breast, with a heterogeneous mass insinuating towards deep planes, hypoechoic with echogenic bands in its interior, BI-RADS category 4C.

A biopsy was performed with a final pathology result of grade II angiosarcoma; A magnetic resonance imaging (MRI) was performed prior to surgical procedure, with multi-lobulated heterogeneous mass finding with nodular areas, some with cystic content towards retroareolar planes and medial quadrants, occupying much of the right breast (Figure 2). This mass presents an intense and heterogeneous enhancement with contrast medium, showing marked neovascularization (Figure 3). The lesion presents peaks of up to 239 % at 2.5 minutes with subsequent configuration of curves type III (Figure 4).

Case 2

25-year-old patient with induration of the right breast of one month of evolution associated with fever and general malaise, postpartum of two months and breastfeeding. Physical examination showed an increase in the size of the breast with induration and violaceous coloration of the skin. Ultrasound showed a diffuse increase in echogenicity without being able to determine a circumscribed mass (Figure 5). The mammographic evaluation showed a marked increase in the size of the right breast with diffuse increase in density that did not allow the visualization of focal lesions (Figure 6). Due to perperual history and the age of the patient, it was managed initially as a mastitis. A puncture was performed from which a high blood content was obtained and, for the characteristics of the tissue, a histological study was performed with result of high-grade breast angiosarcoma. Subsequently, a contrast-enhanced tomography of thorax and abdomen for staging was done. Initially, no lung or intraabdominal neoplastic involvement was found; however, axillary lymph node involvement was documented with invasion of the pectoralis major (Figure 7). The disease progressed and the patient died at 12 months of the diagnosis.

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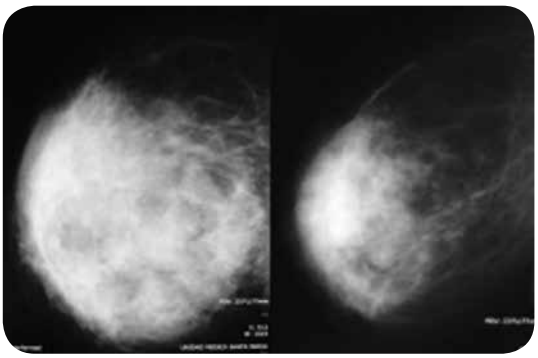

Figure 1. Cranio-lateral and oblique lateral projections of the right breast in which there is an increase in breast density and retroareolar nodules.

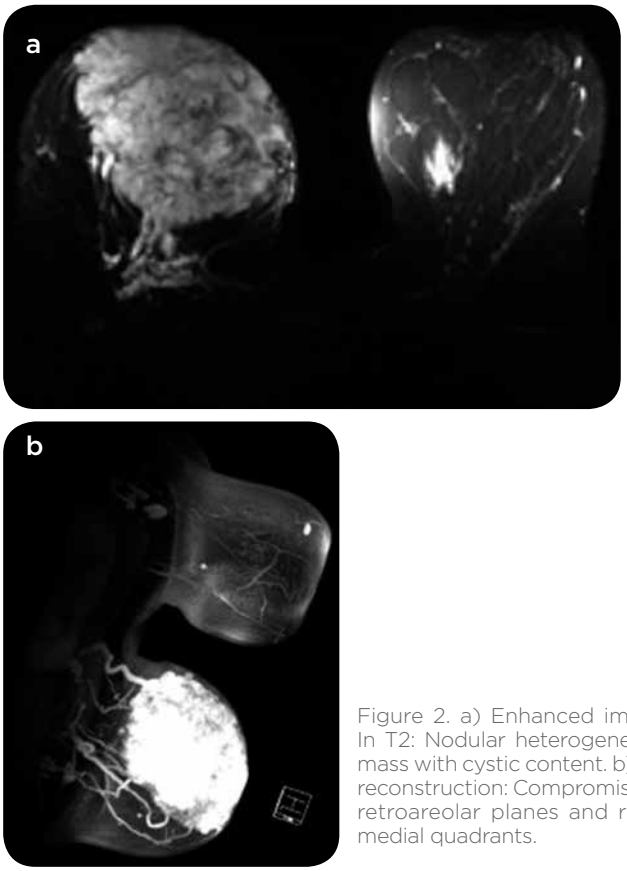

Figure 2. a) Enhanced image In T2: Nodular heterogeneous mass with cystic content. b) 3D reconstruction: Compromise of retroareolar planes and right medial quadrants.

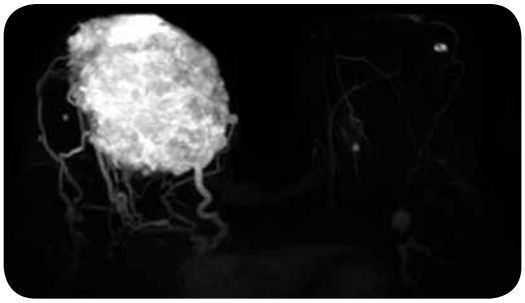

Figure 3. Dynamic T1 MIP: Shows intense and heterogeneous enhancement of the lesion with marked neovascularization.

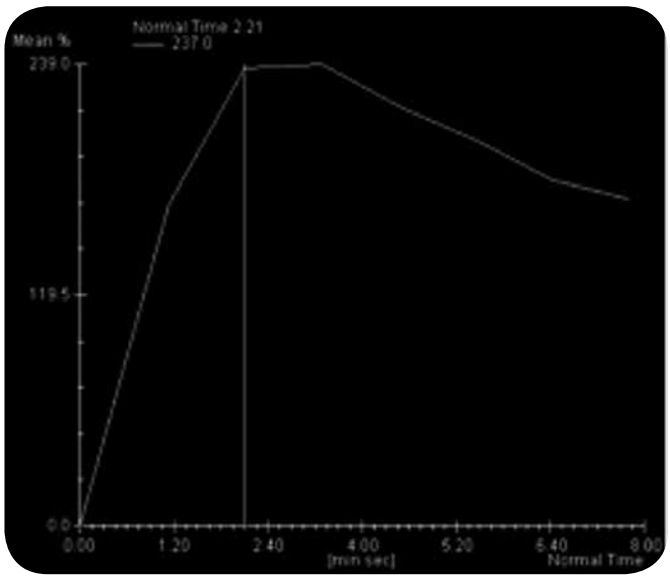

Figure 4. Type III curve of the lesion showing enhancement peaks that reach up to 239 % at 2.5 minutes with subsequent washing.

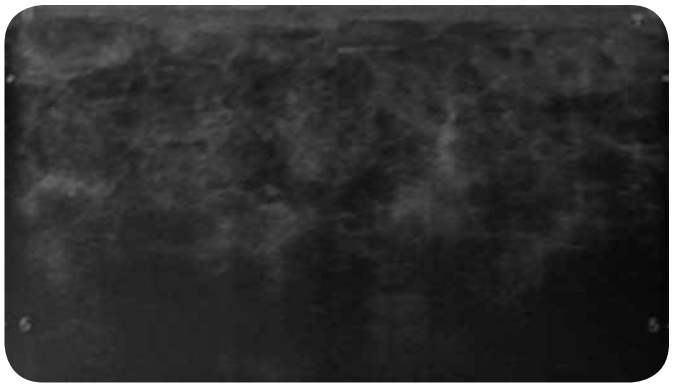

Figure 5. Diffuse increase in the echogenicity of the mammary parenchyma, Diffuse distortion of the glandular architecture without being able to delimit a clearly circumscribed mass.

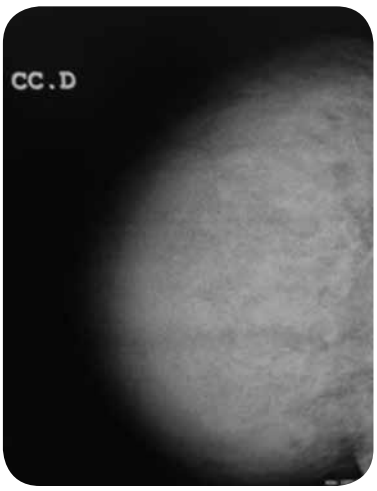

Figure 6. Mammography: craniocaudal projection of the right breast. Marked increase in breast size with diffuse increase in the density that makes it impossible to visualize focal lesions. No microcalcifications are identified.

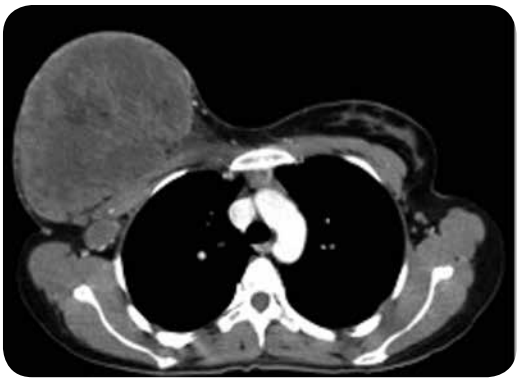

Figure 7. Contrast CT scan of the chest. It shows a large mass that occupies the whole of the right breast. The density is heterogeneous with peripheral contrast enhancement. The thickening of the skin, invasion of the pectoralis major and a large axillary adenopathy that has lost its morphology can be observed, which suggests secondary commitment.

Discussion

Angiosarcomas are malignant tumors that originate from endothelial cells that line the vascular channels, which are characterized by rapid proliferation and an extensive infiltrative component (1). Primary angiosarcomas of the breast occur sporadically in young women and usually appear as palpable masses. Secondary angiosarcomas occur more frequently after conservative management with radiotherapy, with an average latency period between 5-6 years.

The usual clinical presentation of angiosarcoma is that of a painless mass with violet coloration of the skin.

Primary angiosarcoma

Conform 0.04 % of all malignant tumors of the breast (2). Three degrees of angiosarcoma have been described (3). Those of low degree are composed of vascular channels that are anastomosed and invade adjacent breast tissue, those of intermediate grade present a more robust neoplastic vascular growth and an elevated mitotic rate, and those of high grade have clear sarcomatous areas, with necrosis, hemorrhage and infarction. There may be multiple degrees in the same tumor such that staging may not be possible with tru-cut biopsy, in which case complete excision would be required to properly determine the degree of the tumor (4,5).

Primary lesions occur in younger women (3rd and 4th decade); these lesions are rarely carcinomas, which appear later in life. Although angiosarcomas have been reported during pregnancy, there is no evidence that these tumors are dependent of hormones (1,6). Patients with primary angiosarcoma present with palpable mass which can be rapidly growing. Violent skin coloration occurs in up to one-third of patients and is attributed to the vascular nature of the tumor. Mammographically, the appearance is nonspecific, the most common finding being a poorly defined mass, with non-calcified or focal asymmetry. Fat, as a mammographic abnormality, obliges to include in the differential diagnosis hemangioma and angiolipoma.

Many women with primary angiosarcoma are very young and the density of the parenchyma can prevent visualization of the mass. Yang et al reported in their series that 19 % of patients had tumors that were not visible on mammography but were visible by ultrasound (US) and magnetic resonance imaging (MRI). In this series, in the echocardiographical evaluation, 38 % of the patients showed diffuse distortion of the glandular architecture, hyper or hypoechoic mixed regions without a circumscribed mass (2). Also, hyperechoic masses have been described with defined lesions and circumscribed lesions and in the assessment with color Doppler they are hypervascular. FDG PET can be used for staging, and in case presentations show focal and intense FDG accumulation in thoracic wall angiosarcomas, heart, pleura and liver (7).

In MRI, angiosarcoma is seen as a heterogeneous mass with low signal in T1 and high signal in T2. In high-grade lesions, irregular areas with high signal in T1, with hemorrhage or venous lakes can be observed. The enhancement of mass depends on the degree of the tumor. Low-grade angiosarcomas progressively enhance, while high-grade angiosarcomas enhance quickly and with wash, in some cases display large drainage vessels. MRI is useful for determining the extent of the tumor and surgical planning, and can also detect residual disease after excisional biopsy (2).

Secondary angiosarcoma

It is found in older women undergoing surgical management and with radiotherapy for breast cancer, with a mean age of presentation of 60 years. There are two types of secondary angiosarcomas: Secondary cutaneous angiosarcoma associated with lymphedema and postradiation angiosarcoma (7).

Cutaneous angiosarcoma associated with lymphedema

Described in 1948 by Stewart and Treves, it develops in lymphedematous extremities and the thoracic wall after mastectomy and axillary emptying. Increased conservative therapy and sentinel ganglion have decreased the incidence of angiosarcoma related to lymphedema.

Post-radiation angiosarcoma

It usually occurs after conservative therapy and radiotherapy, and rarely after mastectomy. It affects the breast's dermis within the radiation field, but can develop in the mammary parenchyma. It is of low incidence (0.09-0.16 %). There is multicentric compromise in up to one-third of patients and the mean size is of 7.5 cm, with a range of 0.4 to 20 cm.

The average time between radiotherapy and the development of angiosarcoma can be as early as 1-2 years or as late as 41 years after treatment. Patients are presented with violet colored plaques or nodules with areas of skin discoloration that may be confused with ecchymosis and delay diagnosis (1,8).

These tumors are typically of high grade. In the mammography changes are seen when there is conservative management and previous radiotherapy, whereby thickening of the skin is due to angiosarcoma may be mistakenly masked or interpreted as modifications of the skin generated by radiotherapy. In the subgroup of cases with

compromise of the parenchyma, it can be seen as ill-defined and asymmetric masses. By ultrasound, skin lesions can be difficult to differentiate from post-therapy thickening of skin and intraparenchymatous lesions can be seen as heterogeneous areas with alteration of the glandular architecture. MRI shows rapid enhancement with contrast medium and plateau curves or washout (2,6).

Treatment

Mastectomy is the usual management. For small low-grade primary lesions conservative surgery can be considered. Chemotherapy with docetaxel may decrease the rate of recurrences and radiotherapy, at fractional doses, may reduce high-growth tumor cells (3,6).

Prognostic

The prognosis depends on the tumor grade. A disease free survival rate is estimated to be 76 % 5 years after initiation of the treatment. Metastatic involvement occurs frequently in bone, lung and liver. Metastases have also been found towards the contralateral sinus. Secondary angiosarcomas have poor prognosis, although sometimes 5-year survival may be better than that of other forms of cutaneous angiosarcoma. The results are subject to complete resection or not of the surgical borders (3).

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