BREAST VASCULITIS PRESENTING AS A TUMOR-LIKE LESION. 
A CASE REPORT

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Breast vasculitis presenting as a tumor-like lesion is rare. The differential diagnosis in these cases can be extremely difficult but is very important for treatment and follow-up. We report the case of an 80-year-old woman who was admitted to our service with a lesion resembling an inflammatory carcinoma of the breast. We discuss the pathological and clinical characteristics, the presentation, assessment and treatment of this case and its resolution. Few reports in the literature have addressed the possible occurrence of systemic vasculitis mimicking cancerous lesions. The most common location of such lesions was shown to be the breast in one review. Although rare, such manifestations can be effectively recognized and treated.

Key words: breast vasculitis, granulomatous angiopanniculitis, tumor-like lesion.

Introduction

Vasculitis (Wegener’s granulomatosis, giant cell arteritis, polyarteritis nodosa, Behcet’s syndrome and various other types of vasculitis) can present both as systemic diseases and as isolated lesions without systemic symptoms or signs. Systemic vasculitis presenting as a tumor-like lesion is very rare. Correct diagnosis and appropriate treatment are sometimes difficult, but they are necessary in order to avoid potentially serious necrotic complications. Few reports on these lesions are available in the literature. Kariv published a review analysis of 79 cases and four new cases presenting as tumor-like lesions¹. One of these new four cases was a polyarteritis nodosa presenting as a breast mass in a 48-year-old woman with low-grade fever, weight loss and myalgia of one month’s duration, associated leukocytosis, thrombocytosis, anemia and high alkaline phosphatase. In his review the most common type of vasculitis with a tumor-like presentation was Wegener’s granulomatosis (28 cases; 35%) and the most common location of such lesions was the breast (22%). Recently, Nooijen described a non-necrotizing granulomatous angiopanniculitis in a 63-year-old woman mimicking a breast cancer². A correct diagnosis could be made by means of fine needle aspiration cytology and histological examination of the specimen after lumpectomy.

We observed a bilateral breast mass in an 80-year-old woman mimicking carcinomatous mastitis with a highly peculiar presentation.

Case report

An 80-year-old woman was admitted as an inpatient to our Service in the General Surgery Department in June 2002 for mastalgia with severe nodularity, erythema and tenderness of the periareolar skin (8 cm diameter) (Figure 1). The symptoms had a sudden onset: five

Figure 1 - Mastalgia with severe nodularity, erythema and tenderness of the periareolar skin (8 cm diameter). a: left breast; b: right breast.
days previous to consultation she noted progressive spontaneous tenderness, edema and erythematous nodules in both breasts. The family history was negative for neoplastic and autoimmune diseases, the father had had type II NIDDM. The past medical history was negative for cardiovascular, respiratory, gastrointestinal, neoplastic and autoimmune diseases. One episode of anemia was reported that had occurred in the perimenopausal period. This had been treated with oral iron supplementation when the patient had become menopausal.

On admission, routine blood counts and biochemistry were normal. Thyroid stimulating hormone, follicle stimulating hormone, luteinizing hormone, 17β-estradiol, progesterone, and prolactin levels were also normal. X-ray of the chest was unremarkable. On physical and gynecological examination there were no other abnormal findings. A mammography showed edematous breasts apparently affected by bilateral inflammatory carcinoma (Figure 2). Breast ultrasound showed marked parenchymal hyperechogenicity without any pattern, suggestive of malignancy. Power color Doppler analysis showed no increase in the vascular signal.

A liver ultrasound was normal. Fine needle aspiration cytology was inadequate (graded C1).

A surgical biopsy was performed by periareolar incision; the biopsy included cutaneous, subcutaneous and glandular layers. This showed leukocytoclastic vasculitis without any atypical features (Figure 3). “Fibrinoid” (eosinophilic) necrosis of the blood vessel walls accompanied by fragmented neutrophil nuclei or “nuclear dust” was shown. In the same specimen a mixed inflammatory cell infiltrate was seen, characteristic of a more advanced phase.

On the basis of the pathological results, blood levels of antinuclear antibody, extractable nuclear antigen, anti-DNA autoantibodies and complement factor C4 and C3 were measured. All were increased.

Prompt and appropriate pharmacological treatment

Figure 2 - Mammography showing an edematous breast mimicking an inflammatory bilateral carcinoma. a: right craniocaudal; b: left craniocaudal.

Figure 3 - Leukocytoclastic vasculitis. a: Focal presence of endothelial cells with the appearance of fibrinoid eosinophilic necrosis (postcapillary venule) (H/E). b: The lesion appears to be in an evolutive phase with a mixed inflammatory cell infiltrate composed of neutrophils and lymphocytes (H/E).
including oral anti-inflammatory drugs and steroids was given. Resolution of the symptoms occurred after a month of continuous treatment. The patient is currently disease free and her immune status is back to normal.

Discussion

Systemic vasculites are very peculiar diseases presenting with a vast array of local and systemic symptoms and signs usually related to inflammatory processes such as anemia, fever and fatigue. Unusual presentations are tumor-like lesions. Such lesions could initially misdirect the diagnosis and delay appropriate medical treatment. Trueb described a periarteritis nodosa presentation by an intact lobular architecture and the presence of emboli. Breast vasculitis, on the other hand, is characterized by lymphatic and small vessel involvement by tumor cell emboli. The usual histological type in inflammatory breast disease presenting as a breast lesion and among Kariv’s four cases of systemic vasculitis one was a breast mass.

In his review, Kariv refers to 79 reported cases, 18 of which located in the breast. In four patients the breast mass was bilateral and in six cases a biopsy was necessary to reach a diagnosis. In seven cases a surgical approach was used consisting of an extensive mastectomy or lumpectomy with axillary lymph node dissection.

The literature confirms that the most common location of tumor-like vasculitic lesions is the breast. The differential diagnosis includes the following diseases: bacterial infections of the breast, inflammatory breast cancer, lymphoma, leukemia, sarcoma and tuberculosis. Bacterial infections are usually seen only in the lactating breast and infections are associated with fever and leukocytosis. The most striking physical findings in inflammatory carcinomas related to the skin are usually unilateral and the axillary lymph nodes are often involved. The usual histological type in inflammatory breast carcinoma is poorly differentiated with subdermal lymphatic and small vessel involvement by tumor cell emboli. Breast vasculitis, on the other hand, is characterized by an intact lobular architecture and the presence of necrotizing vasculitis involving small and medium-size arteries. This is accompanied by fibrinoid necrosis of the vessel walls, thrombotic luminal occlusion and a mixed inflammatory infiltrate extending into the surrounding adipose tissue. Lymphomas, leukemias and tuberculosis are usually easy to rule out in the differential diagnosis because of the associated systemic symptoms. Wegener’s granulomatosis is usually unilateral in 40 to 50-year-old women and may present as a 2 to 6 cm lump with positive axillary lymph nodes and cutaneous lesions. Mammography shows a spiculated lesion with ill-defined margins mimicking cancer; these lesions have characteristic histopathological features. It is therefore necessary, in our opinion, to include vasculitis in the differential diagnosis of a tumor-like lesion and consequently to deliver prompt and appropriate treatment, avoiding unnecessary surgery. When an elderly woman presents with a unilateral or bilateral breast mass associated with systemic symptoms and parameters of inflammation, vasculitis should be suspected. Vasculitis is characterized by specific histological hallmarks; the diagnosis is easily made after a biopsy.

Conclusions

Few reports are available in the literature of systemic and localized vasculitis mimicking breast disease. The most common type of vasculitis with a tumor-like appearance is granulomatous angioinflammation of the breast. This case study suggests that systemic vasculitis may present as a tumor-like lesion in older patients. Localized vasculitis presenting as a cancerous lesion of the breast is rare; however, inflammatory systemic indices are good indicators and can be effectively used as diagnostic tools. Prompt and appropriate medical treatment should be given in order to avoid unnecessary surgical biopsy.

References