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Original Article

Stewart-Treves Syndrome: Case Report

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Abstract

I In 1948 Stewart and Treves1 described a syndrome related to the association between lymphangiosarcoma and chronic lymphedema due to radical mastectomy and radiotherapy. Currently, literature data reveals around 400 published cases[2]. However, this pathology is becoming each time rarer due to the growing indication of conservative breast surgery and sentinel lymphonode research, thus reducing the need of axillary lymph node dissection with subsequent lymphedema. Described will be the case of a woman that developed angiosarcoma in shoulder 17 years after mastectomy with adjuvant radiotherapy.Stewart-Treves syndrome is related to the rise of angiosarcoma in patients with chronic lymphedema. It is currently treated as a rare disease. We describe the case of a woman who developed angiosarcoma in shoulder 17 years after mastectomy with adjuvant radiotherapy.

Keywords: Lymphedema; Sarcoma; Lymphangiosarcoma; Radiotherapy; Mastectomy. Syndrome.

Introduction

Described for the first time in 1948 by Stewart and Treves,[1] this syndrome was related to the association between lymphangiosarcoma and chronic lymphedema due to radical mastectomy and radiotherapy. It tends to be treated as a rare disease, with approximately 400 cases published in the literature.[2] The tendency of this pathology is becoming each time rarer due to the growing indication of conservative breast surgery with sentinel lymphonode research, which has been reducing the need of axillary lymph node dissection and consequently, its main complication; lymphedema. For reasons of rarity and difficulty of diagnosis, we will describe a current case.

Case Report

A 70-year-old woman with history of left breast cancer diagnosed in 1990 (infiltrative duct carcinoma), submitted to neoadjuvant radiotherapy in thoracic wall, supraclavicular fossa and axillary region (total dose of 5,000 cGy) followed by radical mastectomy (Halsted) conforming to the standard treatment of the time. Patient developed clinically important lymphedema in addition to repeated erysipelas. In January of 2008, patient presented nodular, fast-growing and painless purplish lesion on the left shoulder.

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Physical exam of left shoulder encountered lesion, fixed deep plane and measuring 8.0cm x 6.0cm. Submitted to incisional biopsy with histopathologic angiosarcoma diagnosis; confirmed by immunohistochemical analysis with positivity for the markers CD31, CD34 and D2-40. Nuclear magnetic resonance (NMR) of the shoulder and left arm evidenced tumoration in shoulder with extension to the deep plane and soft tissue; Computerized tomography (CT) of thorax and ultrasound (US) of total abdomen without evidence of metastases.

Proposed as curative treatment, was resection of the lesion with three-dimensional wide margins. During intraoperative, frank invasion of the brachial plexus was evidenced and interscapulothoracic amputation was then opted for, the fashioning of a posterior flap ample for the primary closing of the resection area as the patient had been submitted to a radical mastectomy with resection of the pectoralis major, which would make the fashioning of an adequate anterior flap unfeasible. The patient had good evolution with discharge on the third postoperative day. Final histopathologic report of angiosarcoma with deep infiltration of muscular planes and free margins is shown in Figure 2. Adjuvant treatment was not indicated.

Discussion

Classically, lymphangiosarcomas are tumors of high-degree malignancy and poor prognosis.[3] Clinically, they manifest as ecchymosis or telangiectasia with multifocal tendency and rapid growth, which could coalesce and produce plates or cutaneous nodules. They can present satellite lesions, ulceration,

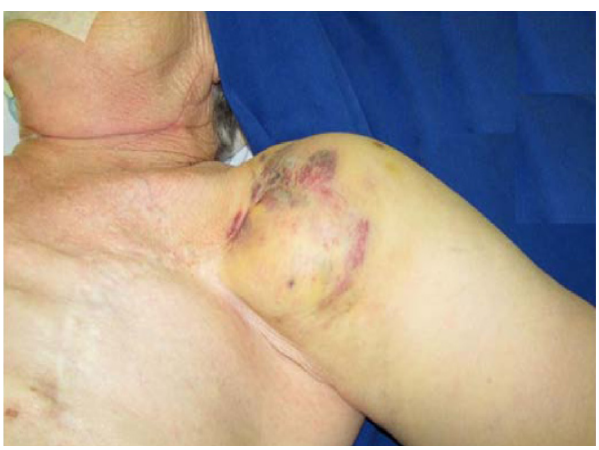

Figure 1 - Lesion of the left shoulder about 8.0cm x 6.0 cm, with presence of hematoma in various phases of evolution

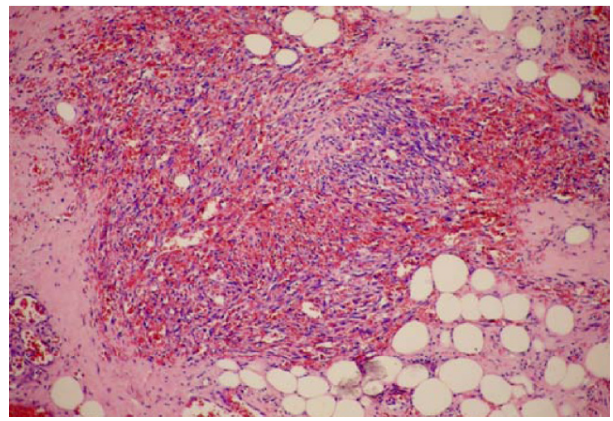

Figure 2 - Histological aspect of the lesion. Vascular lesion of small-vessel inter-anastomosic with fusicellular cells presenting intense atypias infiltrating adipose tissue

pain and neurological or vascular symptoms depending on the relationship with the vascular-nerve bundle of the member affected.[4] They tend to present local diffusion through subcutaneous for the adjacent structures and early systemic hematogenic diffusion mainly for the lung.[3]

The most common location is the arm, followed by the forearm, elbow and thoracic wall.[3]

The average time interval between mastectomy and lymphangiosarcoma development is 11 years. The etiopathogenesis is related to the development of chronic lymphedema that generates a hyperstimulation to the formation of new lymphatic vessels with loss of control of the normal regulation which predisposes the emergence of cellular mutations of the endothelial of such vessels leading to the development of angiosarcoma. [2]

The diagnosis is made based on the history and physical exam associated to the biopsy of the lesion. The biopsy can be made through longitudinal incision in the affected member.[3]

Staging should be made through CT or NMR of the affected member and thorax CT, as these tumors metastasize more frequently via hematogenic than lymphatic, the lung being the main affected organ. [2-3]

The standard treatment with curative intention consists of resection of the lesion with three-dimensional wide margins with the purpose of limb preservation.[3] In the cases that resection with preservation of the limb is not possible (frank invasion of the vascular-nerve bundle) the curative surgery consists of amputation or classic disarticulation of the limb. A preservation strategy of the limb would be through Tickhoff-Limberg surgery, which consists of

the resection of the articulation of the shoulder with preservation of the vascular-nerve bundle and of the limb. In the described case, such procedure was not indicated due to the involvement of the vascular bunch of the axilla. Classic work of the literature comparing three-dimensional resection with amputation did not show increase in the survival, being an average of 19 months, and that in 2 years of attendance, half of the patients had died, with 5 year survival of 10% .[5] The early diagnosis associated to oncologic resections is the best strategy to offer to the patients a greater survival with a better quality of life.

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