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RESEARCH ARTICLE

CUTANEOUS ANGIOSARCOMA AFTER TREATED BREAST CANCER: A CASE OF STEWART- TREVES SYNDROME

***Azegrar, M., Amaadour, L., Oualla, K., Benbrahim, Z., Arifi, S. and Mellas, N.**

Medical Oncology Department of Hassan II University Hospital of Fez, Morocco

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ABSTRACT

Cutaneous angiosarcoma (AS) is an aggressive malignant tumor. Chronic lymphedema is one of the major risk factor. Colled Stewart- Treves, It typically presents in women who develop lymphedema in the upper extremity secondary to axillary lymph node dissection for breast cancer surgery. We report a case of a 77-year-old women who underwent 12 year ago: a total mastectomy with an axillary lymph node dissection and an adjuvant chemotherapy and radiotherapy for left breast carcinoma, she presented 3 months ago edema on the left upper limb, Skin biopsy confirmed the diagnosis of angiosarcoma. A treatment by chemotherapy was proposed for the patient. Radical surgery is the best treatment to date for this rare disease. Conservative surgery with adjuvant radiotherapy is also possible. Systemic chemotherapy is reserved for locally advanced unresectable and metastatic forms. We advocate long term follow-up for every post mastectomy lymphedema to diagnosis this fatal disease when curable.

INTRODUCTION

Stewart-Treves syndrome (STS) is a rare, deadly cutaneous angiosarcoma (AS). Chronic lymphedema is one of the major risk factor. It is a rare syndrome (0.5 and 10%) who is manifested by angiomatous lesions of the arm in women who underwent mastectomy for breast cancer.

Observation: A 77 year- old women, underwent 12 year ago: a total mastectomy with an axillary lymph node dissection and adjuvant chemotherapy and radiotherapy for left breast carcinoma. She presented 3 months later an edema on the left upper limb with appearance of ipsilateral painful angiomatous nodules on the left arm and forearm. Histology objectified a pleomorphic spindle-shaped cell with vascular differentiation infiltrated into the entire dermis and forming vascular slits. Immunostainig was positive for CD34 and negative for HHv8. The Ki67 was positive in 50%. This findigs led to a diagnosis of angiosarcoma on the STS, No metastasis were found elsewhere and the patient was a candidate for chemotherapy by paclitaxel weekly with a good evolution after 5 cycles.

DISCUSSION

STS was first described in 1948 by Stewart and Treves (Breidenbach *et al.,* 2000). The prevalence is estimated at approximately 0.45% in patients who survive longer than five years after a mastectomy (Ho *et al.,* 2013) The incidence has been decreasing:

****Corresponding author:*** **Azegrar,**
Medical Oncology Department of Hassan II University Hospital of Fez, Morocco

Conservative surgery to minimise loss of breast tissue, coupled with axillary irradiation treatment, has led to a reduction in chronic lymphoedema to as low as 4% (Lee *et al.,* 1988). Lymphedema as a result of axillary lymph node dissection is considered a primary risk for developing AS (Ho *et al.,* 2013). Radiation therapy secondarily tends to induce the development of AS (maybe by fibrosis and proliferation of lymphatic vessels). Also, it is suggested that the axillary lymph node dissection predisposes to AS. Our Patient had these three risk factors. The disease affects descending arm, forearm, elbow or anterior thorax. The first sign of SST maybe a bluish bruise papule or nodule but lesions rapidly progress to ulcers or bourgennants tumors as the case of our patient. Histological examination with an optical microscope (Manner *et al.,* 2010) commonly found within a stromal edema, a nodular tumor proliferation, consisting blanks pace vascular cavities, consisting of many carriers endothelial cell atypia. The vascular and conjunctiva nature of these tumors is confirmed by immunohistochemistry, with positive staining for laminin, antibodies against factor VIII, type IV collagen and CD31 (Ordóñez *et al.,* 1984). Negativity of epithelial differentiation markers (EMA, cytokeratin) finally eliminates the main differential diagnosis, consisting of cutaneous metastases of breast cancer. High level gene amplification of MYC is also a distinguishing feature of cutaneous secondary angiosarcomas, which develops after chronic lymphoedema or irradiation. This is in contrast to primary angiosarcomas, which tend to be present deep in soft tissue and do not have high MYC gene amplification (Pizzi *et al.,* 1983). The treatment of secondary angiosarcoma is similar to angiosarcomas of another origin; it usually requires medical and surgical treatment, Patients who are treated with amputation rather than radiotherapy have a better prognosis.

Iconography

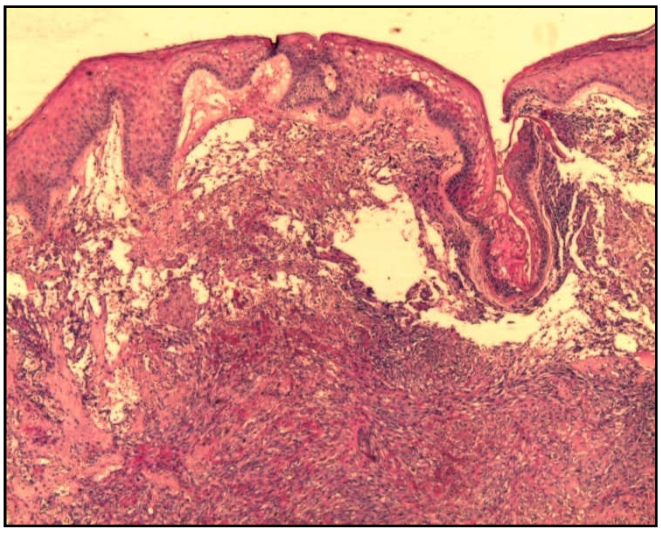

Image 1. HE GX50 staining: Dermal proliferation made of irregular fissures on the surface and massive in depth

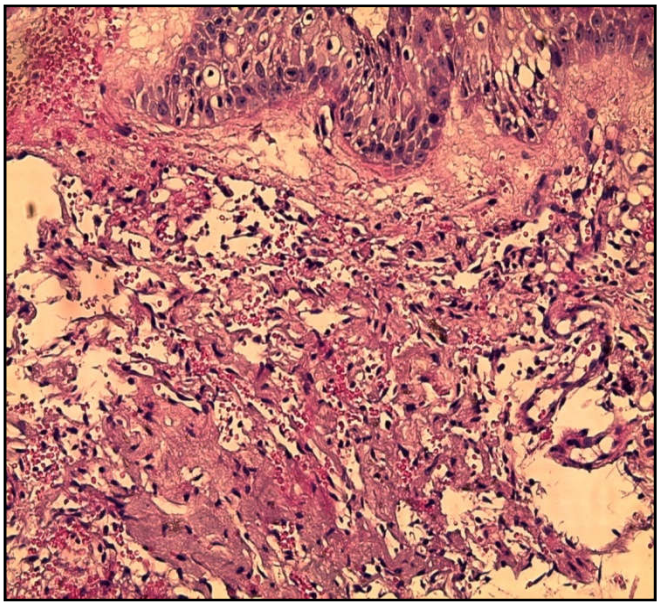

Image 2. Coloration HE GX200: Fentes vasse irrégulières en surface

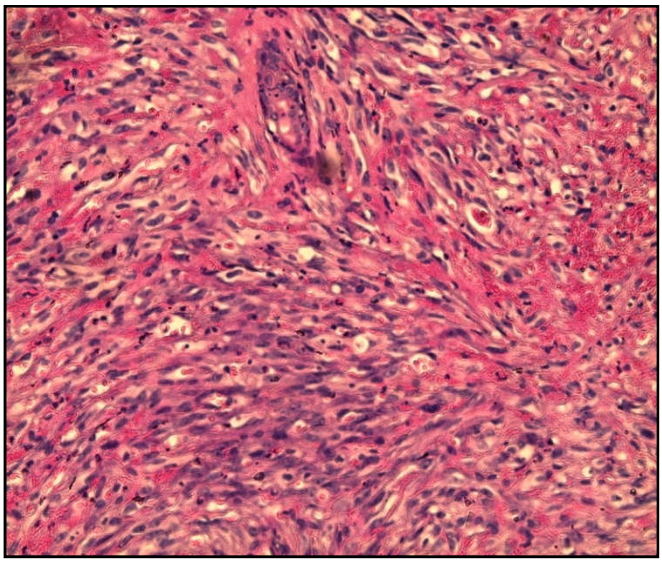

Image 3. HE GX200 staining: Massive fusocellular proliferation + extravasation of red blood cells

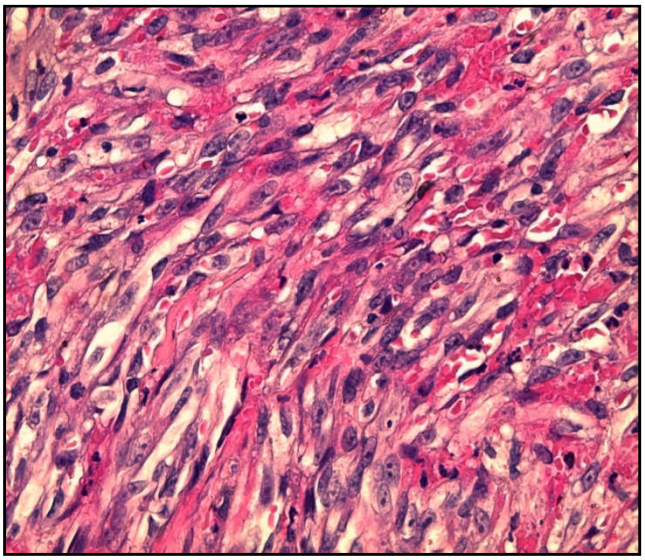

Image 4. HE GX400 staining: Massive fusocellular proliferation + extravasation of RBCs

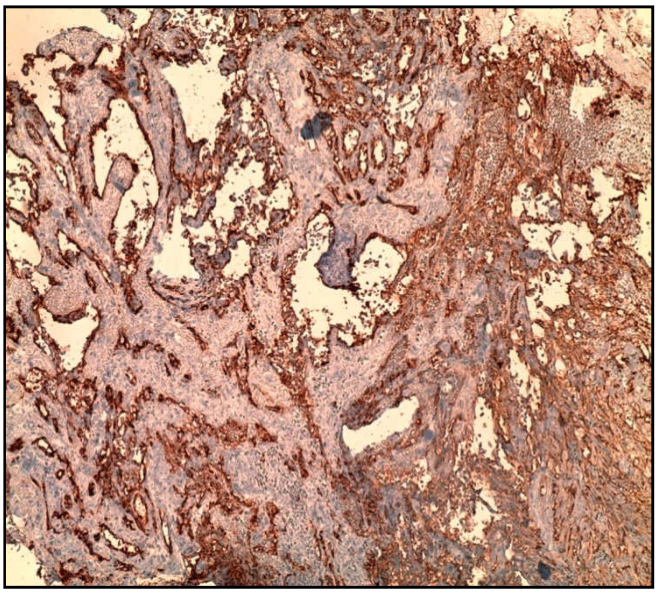

Image 5. IHC GX100: Tumor cells expressing anti-CD34 AC

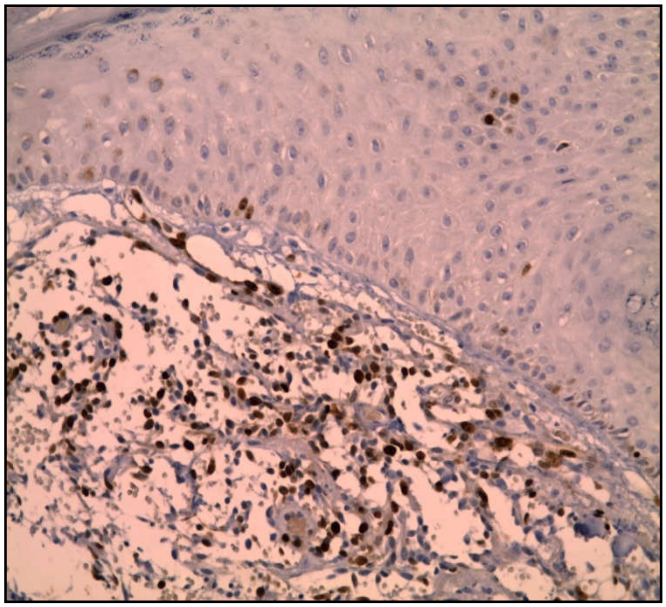

Image 6. 430 IHC GX200: Index de prolifération Ki67 élevé

Despite this, their overall survival remains very poor with a survival range of 5–8 months (Sharma and Schwartz, 2012). Chemotherapy agents used in this disease includes 5-fluorouracil, methotrexate, bleomycin, and/or a combination of actinomycin D, vincristine, doxorubicin, cyclophosphamide, and/or dacarbazine. Even with these treatment therapies, local recurrence and metastasis is high (Sharma *et al.,* 2012; Shon *et al.,* 2011) Treatment with immunotherapy has been demonstrated to be favorable as a palliative treatment for the management of pleural effusions caused by metastatic disease (8.9). The prognosis remains poor with a survival rate after 5 years estimated at 10% (Stewart *et al.,* 1948).

Conclusion

STS is a rare aggressive tumor. Early detection and treatment is important. Therefore, breast surgeons and radiologists should be aware of skin changes during the follow-up of patients who have undergone breast surgery to improve her early detection.

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