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Stewart-Treves Syndrome: A Rare and Fatal Complication of Modified Radical Mastectomy.

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ABSTRACT
INTRODUCTION: Stewart-Treves Syndrome is a deadly and rare disease. It is defined as the association between chronic lymphedema and angiosarcoma. It was originally described in women who developed lymphedema as a result of axillary lymph node dissection associated with mastectomy for breast cancer.
MATERIALS AND METHODS: Here, we present the case of a 55 year old hypertensive obese female with multiple hemorrhagic and elevated purple black nodular lesions on her left upper arm. Patient had a significant lymphedema in her left upper limb for the last 18-20 years. She also had a history of left sided modified radical mastectomy 20 years ago. A punch biopsy was taken from the lesion.
RESULTS: The Histopathological report confirmed it to be a case of High Grade Angiosarcoma. Patient was referred to the Oncology/Radiotherapy department of our hospital for consideration of Chemotherapy/ Radiotherapy.
DISCUSSION: The treatment ofStewart-Treves syndrome is complex; it usually requires medical and surgical treatment.The overall prognosis of this disease is very poor; it has a high rate of local recurrence and metastatic disease.
KEYWORDS: Stewart-Treves Syndrome, Angiosarcoma, Lymphedema, Mastectomy

I. INTRODUCTION

The Stewart-Treves syndrome is a rare and deadly entity, which is defined as angiosarcoma arising in the setting of chronic lymphedema. It typically presents in women who develop lymphedema in the upper extremity secondary to axillary lymph node dissection for breast cancer surgery. It is extremely uncommon in the lower extremities as a result of idiopathic chronic lymphedema.

II. CASE REPORT

A 55 year old hypertensive obese female has presented with multiple hemorrhagic and elevated purple black nodular lesions on her left upper arm. Patient had a significant lymphedema in her left upper limb for the last 18-20 years. She also had a history of left sided modified radical mastectomy20 years ago for infiltrating ducal carcinoma of breast. Patient received Chemotherapy, Radiotherapy and Hormonal therapy. A punch biopsy was taken from the lesion.

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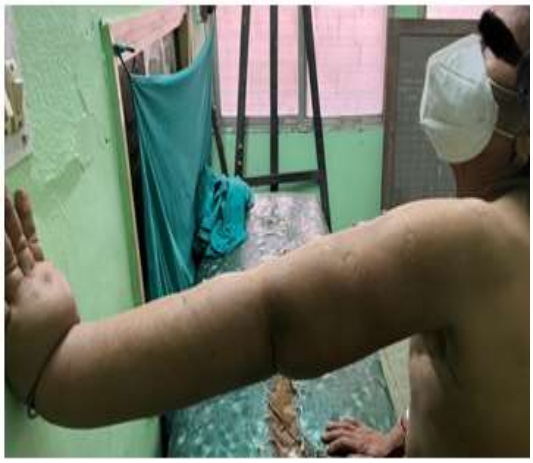

III. RESULTS

The Histopathological report confirmed it to be a case of High Grade Angiosarcoma. Patient was referred to the Oncology/Radiotherapy department of our hospital for consideration of Chemotherapy/ Radiotherapy.

IV. DISCUSSION

The incidence of this disease is very low. Lymphedema affects approximately 14% of patients who undergo complete axillary lymph node dissection.

Patients usually have a significant history for breast cancer and radical mastectomy, usually 5-15 years before sarcoma presentation. The mean onset between radical mastectomy and lymphangiosarcoma is 11 years.

Lymphedematous changes are required for the development of this disease. Usually, this lymphedema occurs without a history of complications, such as thrombosis or infection. Lymphangiosarcomas are much more frequent in the upper extremity, 90% of cases present like this. Moreover, lymphangiosarcomas are present in the ipsilateral side in which the radical mastectomy was performed and the lymphedema developed. The lymphedema in the upper extremities gradually extends from arm to forearm and dorsal aspects of the hand and fingers. Local discomfort develops when the skin distends, but this occurs usually in advanced stages of the syndrome. Recurrent erysipelas may develop in areas of long-standing chronic lymphedema. Patients may also note atrophic skin that has become pachydermatous with wrinkle lines. Telangiectasias and hyperkeratosis may also occur. The initial lesion includes either a palpable subcutaneous mass or a poorly healing eschar with recurrent bleeding. In more advanced stages, multiple reddish blue macules or nodules develop and may become polypoid. There may also be small satellite areas surrounding the nodules that may become confluent. As the lesions progress, the overlying atrophic skin becomes ulcerated, leading to infection and bleeding. Necrosis of the skin can also happen. If left untreated, extensive cutaneous nodules appear and metastatic disease develops. The most common site of metastasis are the lungs, followed by liver, bone, soft-tissue structures and lymph nodes via the hematogenous route.

V. CONCLUSION

The treatment of Stewart-Treves Syndromeis complex; it usually requires medical and surgical treatment.Even when a 2-3-cm wide margin of resection is obtained at the primary surgery, a high local recurrence rate is observed. Long-term survivorship after chemotherapy/radiation is reported in case reports, but the overall results are poor.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. Treatment with immunotherapy has been demonstrated to be favorable as a palliative treatment for the management of pleural effusions caused by metastatic disease.

Preventative measures should be encouraged given the disappointing treatment results. These include treatment of the chronic lymphedema, weight loss, pressure garments, physiotherapy, compressive devices, microsurgery, and laser therapy.

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