

STEWART-TREVES SYNDROME IN A MALE PATIENT

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ABSTRACT

Lymphangiosarcoma is an uncommon vascular tumor that usually develops in chronic lymphedema. The etiology of lymphangiosarcoma remains unknown. It develops mainly after mastectomy, in association with chronic lymphedema (Stewart-Treves syndrome), or after radiotherapy. The early diagnosis of this aggressive tumor is important, as it has a high risk of local recurrence and metastasis. Stewart-Treves syndrome occurs in 0.5% of patients who survive mastectomy for more than 5 years. The mean age at appearance of the angiosarcoma is 62 years, and the mean interval between mastectomy and the appearance of the tumour is 10.5 years. Only two cases have been reported in men following mastectomy. We present a 70-year old male patient with lymphangiosarcoma which developed three and a half years after the diagnosis of breast carcinoma. The patient underwent mastectomy of the right mammary gland with ipsilateral axillary lymph node dissection and had postoperative chemo- and radiotherapy.

Keywords: lymphangiosarcoma, Stewart-Treves syndrome, chronic lymphoedema, mastectomy

A 70-year old Caucasian male patient underwent mastectomy and axillary lymph node dissection on the right side for breast carcinoma. Postoperatively, the patient received both radio- and chemotherapy. He developed chronic lymphedema of the ipsilateral upper limb few months after the treatment. Three and a half years later the patient presented with a massive oedema of the right upper limb and multiple easy-bleeding cutaneous nodules. On clinical examination, the patient's right arm was swollen and had multiple disseminated bright red papules

and violaceous, solid, ulcerating nodules (Fig. 1 and Fig. 2). On the anterior right chest wall a red indurated large plaque was present with multiple ulcerous-necrotic nodules, covered with haemorrhagic crusts (Fig. 3). The right axilla was erosive with an abundant exudate (Fig. 4). On the right shoulder there were numerous bluish nodules (Fig. 5). On the right side at the back there was a large purpuric macule with brown haemorrhagic crust on top (Fig. 6).

A biopsy was performed. The histology demonstrated lymphangiosarcoma. There was hyperkeratosis and pseudo epithelial hyperplasia. In the entire papillary and the lower layer of the reticular dermis there were multiple anisomorphic angioblast with nuclei in different size situated in clefts of neovascularisation (Fig. 7, Fig. 8).

Immunohistochemical studies revealed antibodies against CD34 and antibodies against Vimentine (Fig. 9, Fig. 10).

We considered a wide differential diagnosis which included: Angioedema, Angioendotheliomatosis, Angiolymphoid hyperplasia cum eosinophilia,

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Received: February 4, 2014

Accepted: February 11, 2014



Fig. 1



Fig. 2

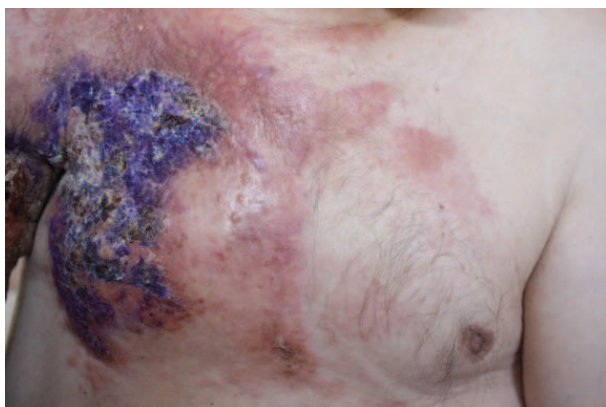


Fig. 3



Fig. 4

Kaposi sarcoma, Lymphangiectasia, Lymphangioma, Lymphocytoma cutis, Melanoma malignum, Metastasis cutis. The clinical appearance of the tumour, the histology results and the immunohistochemical data helped us to establish the diagnosis Steward-Treves syndrome.

The patient underwent systemic and local antibiotic therapy. Considering the size of the tumour and the patient's general condition, surgery wasn't an option and he was referred to the oncology unit for palliative poly-chemotherapy.

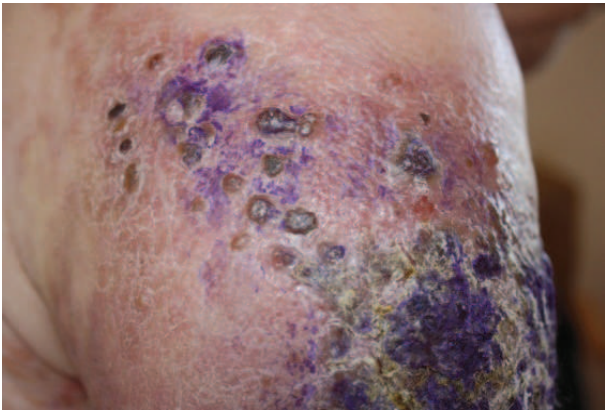


Fig. 5



Fig. 6

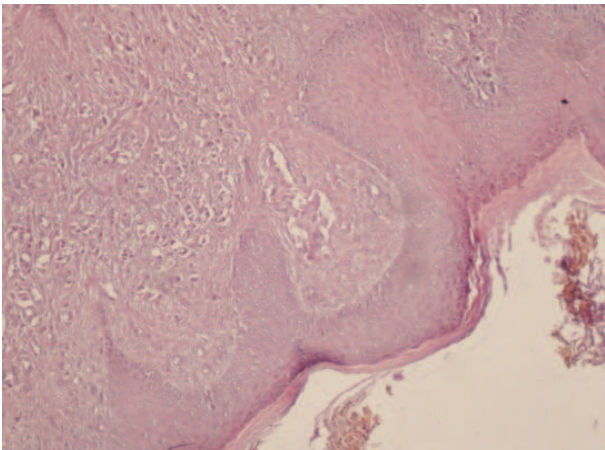


Fig. 7

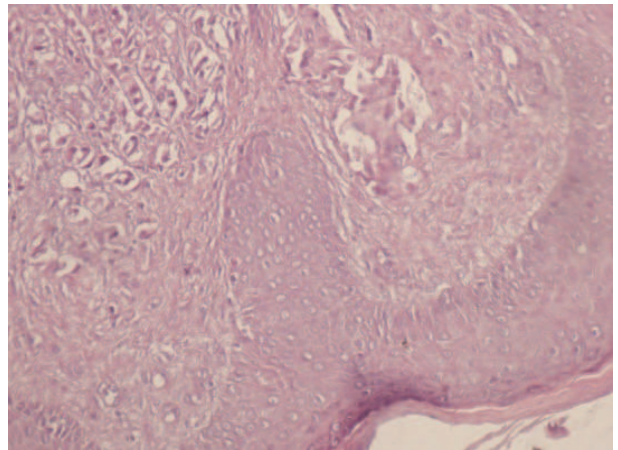


Fig. 8

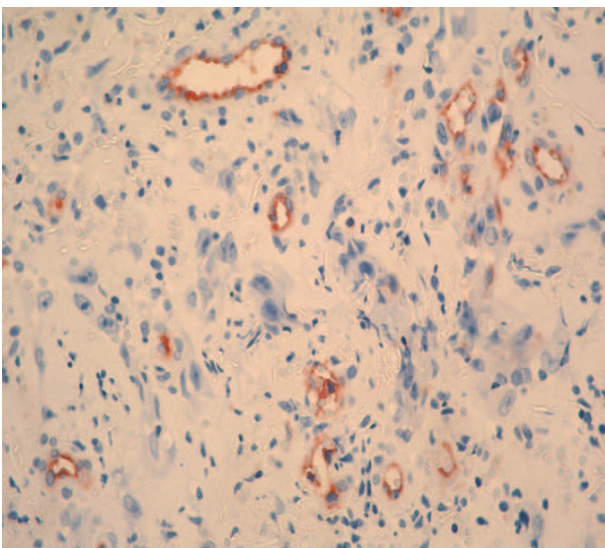


Fig. 9

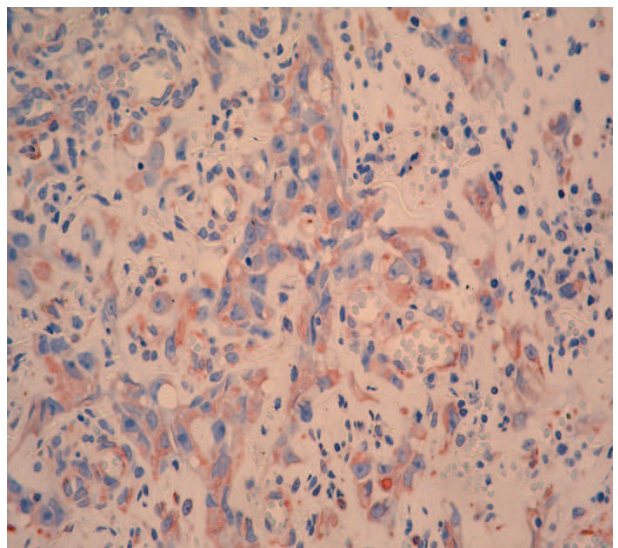


Fig. 10

DISCUSSION

Steward-Treves syndrome is a rare deadly cutaneous lymphangiosarcoma that develops in longstanding chronic lymphoedema (1). By definition is a malignant vascular tumour, arising from lymphatic endothelium in association usually with lymphoedema provoked by radical mastectomy due to therapy for breast cancer. In the meantime lymphangiosarcoma has been reported to occur in patients with lymphoedema of other origin such as congenital, idiopathic, traumatic, filarial, or postsurgical causes of lymphoedema (2). The syndrome was first described in 1906 by Lowenstein, who reported a case of angiosarcoma in a chronic posttraumatic lymphoedema (3). Later, in 1948 Steward and Treves described six cases of lymphangiosarcoma in female patients with elephantiasis chirurgica after mastectomy (4). Only two cases have been reported in men following mastectomy (5).

Lymphangiosarcoma is about 5% from all angiosarcomas (6). Stewart-Treves syndrome occurs in 0.03% of patients who survive mastectomy for more than 5 years (7). The mean age of appearance of the lesions is between 65 - 70 years, and the mean interval between mastectomy and the appearance of the tumour is 10.5 years (8).

The pathogenic mechanism by which lymphoedema induces the development of angiosarcoma is complex. Due to the high incidence of patients with postmastectomy angiosarcoma Stewart and Treves suggested that the main causative factor is the existing of a "systemic carcinogenic factor". Schreiber et al. offered the idea of local immunodeficiency that might lead to oncogenesis (9). Sternby et al. suggested that radiation therapy played an important role in the pathogenesis of the disease (10). Styring et al. defined the role of radiotherapy as an indirect cause of lymphangiosarcomas (11).

McConnell and al. presented the following classification of the stages in the development of lymphangiosarcomas (12):

1. Stage of longstanding chronic lymphoedema.
2. Stage of premalignant angiomas.
3. Stage of malignant angiosarcoma.

Imaging studies used in staging include magnetic resonance imaging, computed tomography

(CT), and radiography. CT is preferred for ruling out metastatic disease to the lung. F-fluorodeoxyglucose positron emission tomography/CT has been found to be helpful in demonstrating the subcutaneous spread of Stewart-Treves syndrome (14).

The prognosis, even with wide surgical excision and subsequent radiotherapy, is poor (13).

Treatment of Stewart-Treves syndrome should consist of radical, wide surgical excision (or limb amputation if obtaining wide negative margins by excision is doubtful) and subsequent radiotherapy. Chemotherapy and radiation therapy have not improved survivorship significantly (16). Nevertheless, even aggressive therapy has resulted in a mean survival rate of 24 months and a 5-year survival rate of only 10%. Untreated patients usually live for 5 to 8 months after diagnosis (17).

CONCLUSION

We present a clinical case of Stewart-Treves Syndrome in a man as a rare but important complication of chronic lymphoedema as a consequence of mastectomy and axillary lymph node dissection. Early recognition of suspicious dermal lesions on a lymphoedematous extremity, immediate biopsy and adequate treatment are important to improve the prognosis of this aggressive disease.

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