

DERMATOPATHOLOGY UNIT

UPP | Department of Dermatology

UPMC Dermatopathology "Case of the Month" Presentations

UPP - Department of Dermatology, Dermatopathology Unit

5230 Centre Avenue (412) 623-2614

Pittsburgh, PA 15232 (412) 682-6450 FAX

Kristina Paley, MD, Muammar Arida, MD, Thaddeus W. Mully, MD, Matthew J. Zirwas, M.D.

MARCH 2006 CASE OF THE MONTH

CLINICAL FINDINGS

Clinical History

76 year-old white female with 20 year history of chronic left lower extremity lymphedema that developed subsequent to left inguinal lymph node dissection for bladder carcinoma presented for evaluation of bleeding tumors on left lower extremity. The patient first noted appearance of violaceous nodules on left lower extremity 4 months prior to presentation.

Her past medical history was also significant for histories of breast and vulvar carcinomas both of which occurred after the diagnosis of bladder carcinoma and were in remission.

Physical Exam

Physical examination of the left anterior tibia revealed a 10 x 6 cm fungating, necrotic tumor on back-ground of a large indurated purpuric plaque (Fig 1). Proximal to the tumor multiple indurated violaceous nodules and plaques were noted (Fig 2).

Histopathology

Examination of a biopsy specimen taken from the indurated purpuric plaque (anterior shin) adjacent to the necrotic area revealed a nodular proliferation of enlarged atypical cells (Fig 3) forming inter-anastomosing channels which are filled with numerous erythrocytes and are lined by plump cells with prominent nucleoli (Fig 4). Some of the cells contained small intracytoplasmic vacuoles (Fig 5). Numerous mitotic figures were seen and there was both individual and en masse cellular necrosis (Fig 6, 7). Immunohistochemical stains revealed

that the atypical cells are positive for vimentin, and for the vascular markers CD31, CD34, and Ulex europaeus (Fig 8, 9, 10). The cells were negative for pankeratin, keratins AE1-AE3, S-100 protein, melan-a, factor XIIIa, and leukocyte common antigen. Examination of another specimen taken from an area away from the main tumor revealed verrucous epidermal hyperplasia overlying greatly dilated lymphatic spaces and ectatic blood vessels (Fig 11). Pronounced papillary dermal edema and extravasation of lymphatic fluid were noted. There appeared to be an increase in blood vessels which are lined by plump enlarged endothelial cells, of few of which are in mitosis (Fig 12, 13).

Compared to findings in the specimen taken from the main tumor, the changes in this second specimen were much more subtle. The pattern of the vascular proliferation appeared reactive; however, subtle involvement by angiosarcoma such as by intravascular extension couldn't be ruled out .

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Figures & Images

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Fig1



Fig 2

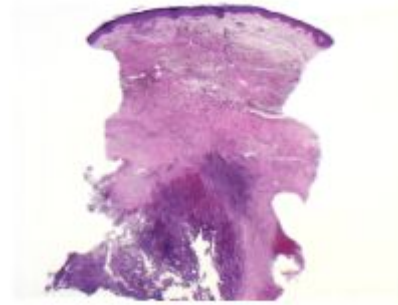


Fig 3

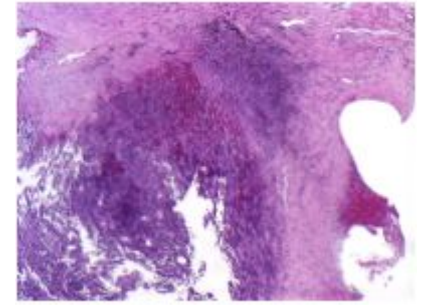
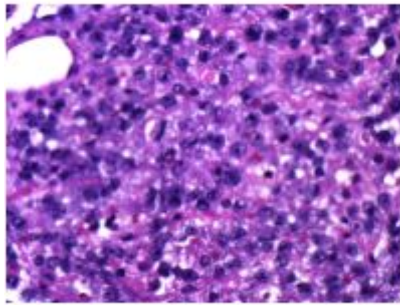


Fig 4



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Fig

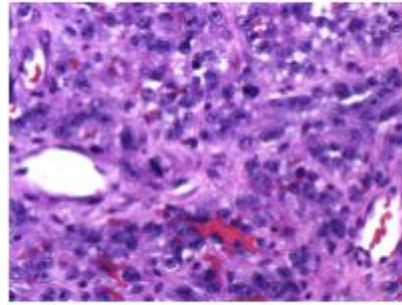


Fig 6

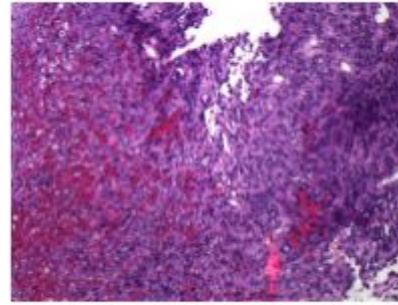


Fig 7

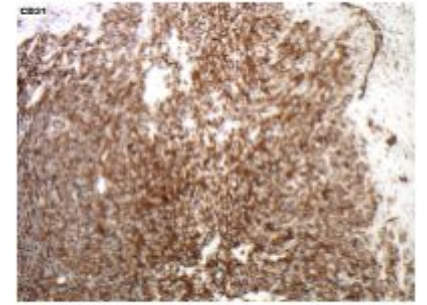
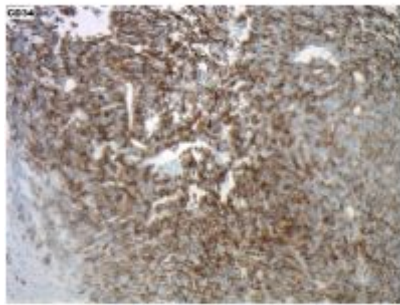


Fig 8



9

Fig

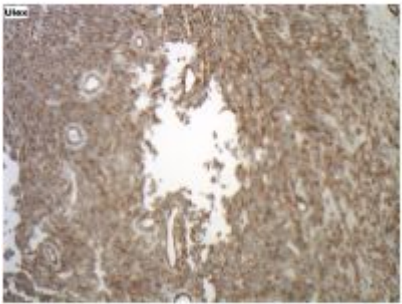


Fig 10

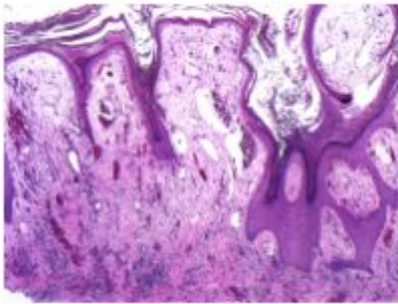


Fig 11

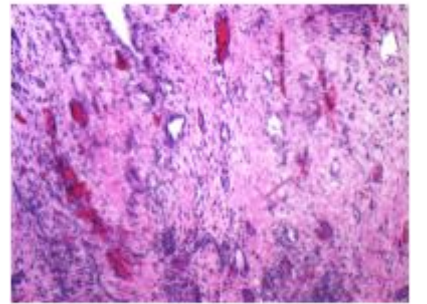


Fig 12

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DISCUSSION & DIAGNOSIS

Diagnosis

Stewart-Treves Syndrome (Lymphangiosarcoma arising in chronic lymphedema)

Discussion

Lymphangiosarcoma arising in chronically lymphedematous extremities was first described by Stewart and Treves in 1948 who noted this phenomenon in 6 females who have undergone radical mastectomy for breast carcinoma (1). Since that original report over 300 cases of lymphangiosarcoma arising at the sites of chronic lymphedema have been reported (2). This phenomenon is now frequently referred to as Stewart-Treves Syndrome.

The majority of cases, like the original report, have been noted in upper extremities in patients who have undergone mastectomies with axillary lymph node dissection for breast cancer. Lower extremity lymphangiosarcoma arising as a result of chronic lymphedema due to filariasis (3,4), obesity (5) idiopathic or primary causes (6,7) and preexisting malignancies (8) is less frequently reported. The discrepancy in incidence between upper and lower extremity lymphangiosarcoma is likely due to high prevalence of breast cancer in a female population. The average duration of lymphedema prior to development of lymphangiosarcoma is 9-12 years with total range of 5-25 years (1,2).

Lymphangiosarcoma is a very aggressive malignancy with a poor prognosis. If left untreated, expected survival is less than 4-6 months (2,9). Amputation of the affected extremity is the most effective treatment of lymphangiosarcoma (2,9). Radiotherapy and chemotherapy have also been reported to prolong survival in a limited number of cases (2,9). As there are no ideal treatments for lymphangiosarcoma, close follow up of patients with chronic lymphedema and early detection are essential factors that can potentially lower the morbidity and mortality from this malignancy.

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