Tropical Stewart-Treves syndrome.

A review of Lymphangiosarcoma occurring in patients affected by chronic lymphatic filariasis

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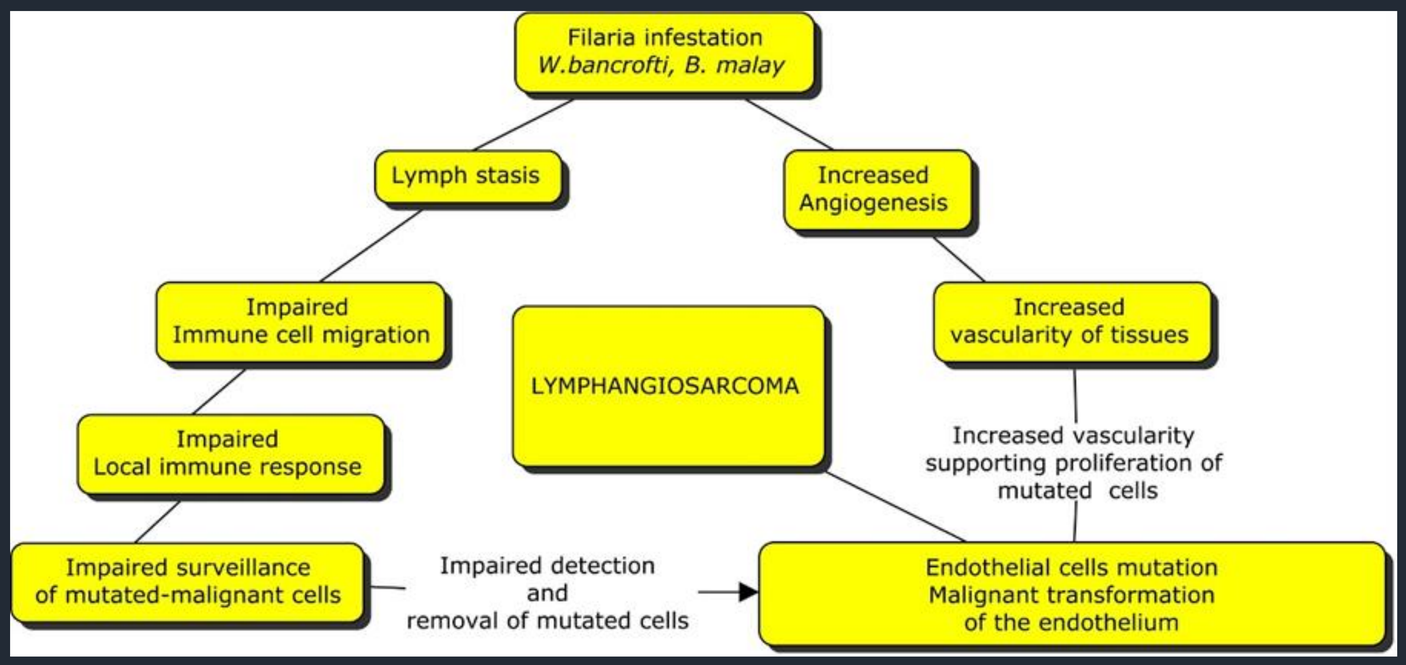
Background

Lymphatic filariasis is a parasitic disease caused by filarial nematodes (including W.bancrofti, B. malayi and B. timori) , transmitted to humans by mosquito vectors (Anopheles, Aedes, Culex, and Mansonia). One of the clinical manifestations of lymphatic filariasis is chronic limb lymphoedema. Lymphangiosarcoma has been described as a potential complication of chronic lymphedema of various aetiology, including lymphatic filariasis.

Aim and Methods

The aim of the study was to describe the occurrence of Stewart-Trevor syndrome in patients with a history of chronic lymphatic filariasis. A review of the literature was performed by consulting PubMed, EMBASE, and Scopus databases using the keywords “filariasis” AND “lymphangiosarcoma” OR “Stewart-Treves syndrome”. Case reports were considered for inclusion. All available cases reporting the association were included, with no exclusion criteria. Descriptive statistics (mean, standard deviation; median 25th–75th percentile) were used to summarize the continuous variables while frequency and proportions for the categorical variables using the computer software JASP, version 0.14.1.



Results

We retrieved nine cases with the mean age of patients being 45±15.6 years (ranging from 19 - 67 years). The majority (n=six cases) of patients were men. Lymphangiosarcoma was diagnosed in all patients after a long history of chronic lymphatic filariasis, of at least 10 years (mean 19.8±7.4 years).

Lesions (the majority ulcerative) were present from 2 to 24 months before lymphangiosarcoma diagnosis, and were associated with lymphadenopathy in five cases. Pain, anaemia and infection were occasionally reported. Differential diagnosis included Kaposi’s sarcoma and lymphangiomatosis; in one case a benign lesion was diagnosed before malignant transformation.

Patients were treated in the majority of cases with major amputation (eight cases) which was above the knee in five cases, below the knee in two cases, in the attempt to achieve eradication of the local disease.

Exitus was reported in two patients, in one for unknown causes and in the other because of the appearance of lung metastasis 3 months after diagnosis.

In patients still alive at report, lung and peritoneal metastasis were reported after 8 years of follow up in one case; four cases were free of metastasis at follow up, but in one case disability secondary to amputation was reported; in one case recurrence of lymphangiosarcoma on the amputation stump occurred.

Conclusions

Lymphangiosarcoma should be considered among differential diagnosis in patients affected by chronic lymphatic filariasis presenting with skin lesions. Lymphatic filariasis patients should be monitored for possible development of lymphangiosarcoma with prompt biopsy of suspicious lesions.

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