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Angiosarcoma in chronic lymphedema a rare case of Stewart-Treves syndrome

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Abstract :
The Stewart-Treves Syndrome is defined as an angiosarcoma (very aggressive malignant tumor originating from endothelial cells) appearing in a specific clinical setting. This tumor develops in patients suffering from chronic lymphedema. The diagnosis relies on medical history, clinical examination and histological assesment (biopsy or resection). This syndrome represents a rare clinical entity. It mostly arises from lymphedema induced by radical mastectomy in breast cancer patients. Unfortunately, the prognosis is poor. A large surgical resection is the treatment of choice, if the patient is a candidate for surgical resection with a curative intent. Radiotherapy is sometimes used as a palliative local treatment. Chemotherapy is only used in more advanced cases, not curable by surgery alone. We report a case of angiosarcoma in long standing filarial lymphedematous lower limb in a female patient.

Keyword : Angiosarcoma, chronic lymphedema, Stewart-Treves Syndrome, filarial leg

INTRODUCTION:

Angiosarcoma arising from a chronic lymphedematous limb is a rare presentation. Although we come across many cases of filarial leg in our country, we rarely encounter a malignancy like angiosarcoma arising from it. Adding to that, this condition has a very poor prognosis because of its highly malignant nature. A radical surgical resection followed by chemotherapy forms the mainstayof treatment.

CASE HISTORY:

A 52 year old female patient with filarial lymphedematous right lower limb for the past 12 years presented with an ulcerative lesion over the medial aspect of right thigh of 3 months duration which had been rapidly progressing. On admission, the ulcer was of size 12 x 10 x 2 cms and was covered with foul smelling slough and had a necrotic base. The edges of the ulcer were everted. The margins were very friable and it bled on touch. It was a severe tender ulcer. Right inguinal lymph nodes were enlarged.

Edge biopsy was done and sent for histopathological examination. It showed admixture of malignant spindle cells and inter connecting vascular spaces lined by pleomorphic endothelial cells suggestive of angiosarcoma. Immunohistochemistry results showed it to be positive for CD34 and negative for HMB45 and cytokeratin, thus confirming the diagnosis of angiosarcoma. Inguinal lymph nodal biopsy also showed angiosarcomatous deposits. CT chest was done to rule out any lung metastasis. Prognosis was explained to patient. Disarticulation at the hip level was done and patient was discharged after one week.

Fig .1. Lymphedematous (Rt) lower limb Fig.2. Fig.2
Ucer with slough Fig.3. Ulcer after slough excision.

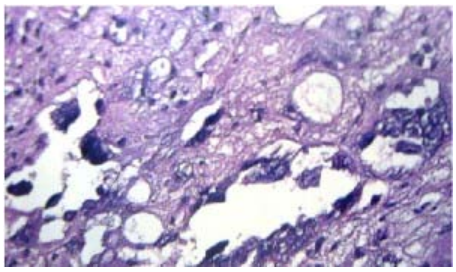

Fig.4. HPE showing malignant spindle cells and inter connecting vascular channels lined by pleomorphic endothelial cells.

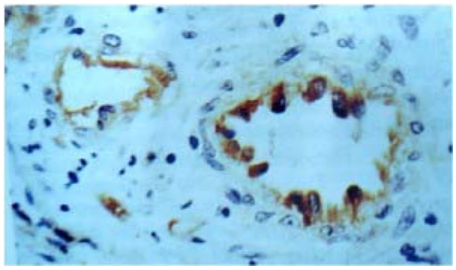

Fig.5. Immunohistochemistry -CD 34 showing cytoplasmic positivity of pleomorphic endothelial lining cells.

DISCUSSION

Stewart Treves syndrome (lymphangiosarcoma) is a rare, aggressive cutaneous angiosarcoma often associated with long standing lymphedema . It mostly arises from lymphedema induced by radical mastectomy in breast cancer patients. It also develops from Milroy lymphedema and in idiopathic, congenital, traumatic or filarial lymphedema .Actually *lymphangiosarcoma is a misnomer . This malignancy seems to arise from the endothelial cells of blood vessels instead of lymphatic vessels. A more appropriate name is hemangiosarcoma. As far its clinical features, it first appears as a bruised area or as*

a purplish discoloration in an extremity. It progresses to an ulcer with crusting and finally to an extensive necrosis involving skin and subcutaneous tissue. There may be satellite lesions from original locale and it metastasizes widely. The various differential diagnosis are ecchymosis, pyogenic granuloma, malignant melanoma, malignant lymphoma, kaposi's sarcoma, lymphangiectasia, lymphangioma, metastatic carcinoma of skin etc. Histologically it presents as admixture of malignant spindle cells and inter connecting vascular spaces lined by pleomorphic endothelial cells. Metastasis from primary tumor can be ruled out by electron microscopy and immunohistochemistry . Angiosarcoma cells express

positive endothelial markers such as CD34, Vimentin , Factor VIII antigen, *Ulex europaeus-I. MRI is recommended to evaluate the local extent of angiosarcomas. However, its true value is in question because of poor results in delineating the margin of the tumor. It metastasizes to lungs, chest walls, liver and bone and has got a high recurrence rate.Chest CT scanning should be performed to rule out metastatic disease to the lungs before the patient undergoes extensive surgery. USG abdomen, CTabdomen and bone scan should be done for metastatic work up.*

The main treatment option is limb amputation. Recent studies have shown that chemotherapy and radiotherapy before amputation increases the survival rate. Chemotherapy with intraarterial *Mitoxantrone and Paclitaxel has shown to be effective. Liposomal doxorubicin has also been tried. The prognosis is very poor as malignancy spreads rapidly. .*

CONCLUSION:

Angiosarcoma arising from a chronic lymphedematous limb is a rare entity which we come across. In India we encounter many cases of filarial lymphedematous limbs but the incidence of angiosarcoma Is very rare. Adding to that ,it is a highly malignant tumor with a very bad prognosis. Early diagnosis and intervention is needed to reduce the morbidity and mortality.

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