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MALIGNANT FIBROUS HISTIOCYTOMA WITH PROGRESSIVE SYSTEMIC SCLEROSIS

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Forty years old female, suffering from progressive systemic sclerosis of six years duration presented with a proliferative growth over the right fore-arm showing sclerodermatous changes. Started as a small nodule gradually increasing in size over the course of six months to the size of 10 cms x 10 cms. The tender proliferative bluish red growth having multiple bleeding points, the base fixed to the underlying bone with no evidence of regional lymphadenopathy.

Key Words : Scleroderma, progressive systemic sclerosis, Histiocytoma, Sarcoma

Case Report

A 40-year-old female presented with thickening of the skin all over the body. Investigated and diagnosed as progressive systemic sclerosis six years back, now presented with a small nodule over the right forearm over the sclerodermatous skin which increased in size in six months duration to a proliferative growth of 10 cms x 10 cms (Fig 1).

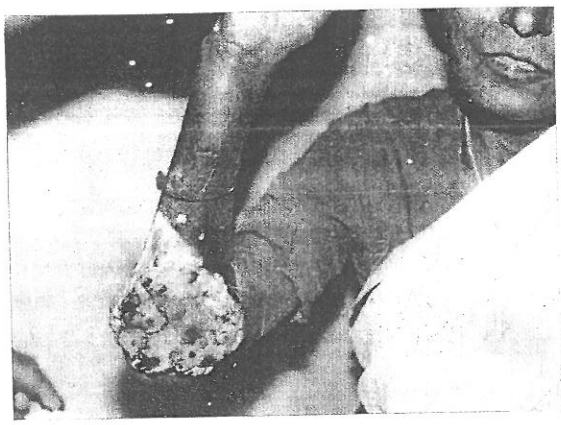

Fig 1 Clinical picture showing tumour over right forearm on sclerodermatous skin with scleroderma over acral region.

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The proliferative growth was tender, bluish red in colour with multiple bleeding points. Base was indurated and fixed to the underlying tissue and bone. There were no significant regional lymphadenopathy.

Routine hematological and biochemical investigations for connective tissue disorders were within normal limits except Hypochromic Microcytic Anaemia. X-ray of right fore-arm- Medullary translucency and periosteal thickening of both bones of fore arm suggestive of malignancy and calcification of the soft tissue in the tumour region (Fig 2). X-ray of right hand - Sclerosis of terminal phalanges with evidence of calcium deposits. X-ray chest was normal. ECG showed inferior wall ischaemia. Endoscopy showed Hiatus Hernia. Skin biopsy right fore-arm shows- Histopathological features of classical scleroderma.

Histology of the growth shows an ulcerated skin epithelium underlying which is seen a tumour mass composed of spindle shaped fibroblasts arranged in stroi form pattern along with histiocytic cells with evidence of nuclear atypia in both types of cells with bizarre multinucleated tumour cells, islands of inflammatory cells also present with areas of hyalinised fibrous tissue, picture suggestive of "Malignant Fibrous Histiocytoma".

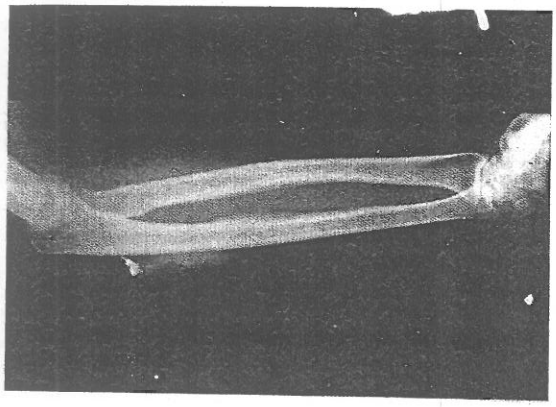

Fig 2 X-ray of the right forearm showing periosteal thickening and translucency of the medullary region and calcification in the tumor area.

Discussion

Malignant fibrous histiocytoma is a pleomorphic high grade sarcoma. It is subdivided clinically into superficial and deep varieties. The superficial variety normally confines to the subcutaneous tissue as a nodular lesion and may rarely invade the skin and causes ulceration in addition to the underlying structure involvement. The deep variety confines itself to the muscles or to the deeper structures of thigh and buttock area and it is twice as common as the superficial variety. Metastasis to the regional lymph nodes and lungs are frequent and fatality is mainly due to later involvement. Among the several varietites described histopathologically the most commonest variety is the classical storiform pleomorphic pattern.[1] In literature various association of malignancies with systemic scleroderma like cancer of lungs, thyroid, ovary, brain, cervix, breast, stomach, oesophagus and lymphomas and leukemias have been reported.[2] Even though the incidence of sarcomas in association with systemic sclerosis is rare, it has been reported. Fibrosarcoma of the tibia in a patient with scleroderma has been reported.[3] Lung cancers are significantly more frequent than other tumours. This case is published in view of its clinical rarity and academic interest.

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