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DERMATOFIBROSARCOMA PROTUBERANS

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A 30-year-old man presented with multiple noduloulcerative lesions on right upper limb for 4 years. He had haemorrhagic pleural effusion bilaterally. Histology of skin lesions showed spindle - shaped cells arranged in cartwheel pattern. He died in spite of anticancer chemotherapy.

Key Word: Dermatofibrosarcoma protuberans

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a cutaneous fibrous tumour that characteristically exhibits marked tendency to recur.

The tumour begins as a firm fibrous plaque, presents with little change for a number of years but eventually enters a phase of rapid growth with formation of multiple nodules which may ulcerate. DFSP, though locally aggressive, metastasises extremely rarely. Arrangement of tumour cells in cartwheel or whirligig pattern is believed to be histologically characteristic. The treatment for this tumour is adequate surgical excision and it carries excellent prognosis. Metastases, though rare, are invariably fatal. We report a case of DFSP with cutaneous and pulmonary metastasis.

Case report

A 30 - year - old farmer presented with complaints of multiple noduloulcerative lesions over right upper extremity for 4 years. He noticed a small swelling on the dorsum of right hand which gradually enchroached on the web space between middle finger and forefinger. After 1 year the lesion ulcerated discharging yellowish fluid. It was treated with excision and radiation on same site as well as along the right forearm and arm medially. These lesions were excised along with amputation of middle finger and forefinger of right hand but lesions again appeared after 1 year. For last 2 months he developed progressive dyspnoea. He gave history of injury to right hand about 10 years back.

When he presented to us he was afebrile, pale, cachectic, and dyspnoeic.

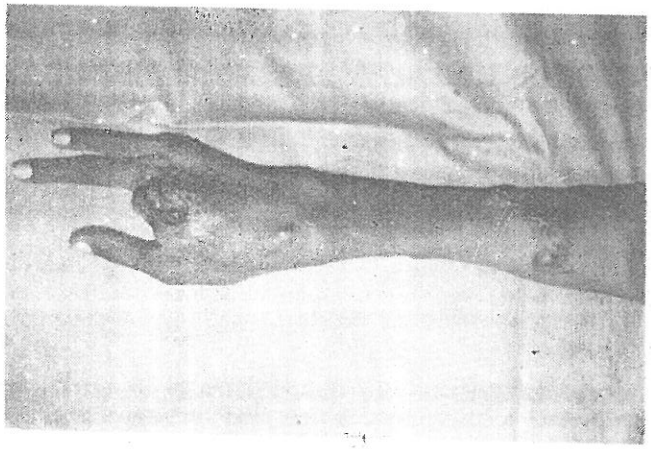

Fig. 1. Noduloulcerative lesions on right hand and forearm

Oval ulcerative lesion of 3cm x 6cm with undermined edges and indurated base was presented on dorsum of right hand (Fig. 1).

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Multiple, firm, nontender, skin-coloured nodules of size 3 cm to 6 cm were present on dorsum of right hand, forearm, and arm. They were attached to skin but not to the deeper structures and a few of them were ulcerated with necrotic floor, undermined edges, and yellowish discharge. On the arm nodules had coalesed to form indurated plaque but otherwise there was no fibrous band in between the nodules. The right axillary nodes were not enlarged. Examination of the respiratory system revealed decreased movements, dull note, and reduced air entry bilaterally at bases.

His haemoglobin was 7.0 gm%. Biochemical parameters were within normal limits. Serum VDRL test and ELISA for HIV were negative. Pus culture produced coagulase-positive staphylococci sensitive to erythromycin and ampicillin, while culture for *Mycobacterium tuberculosis* and fungi revealed no growth. X-ray chest showed bilateral pleural effusion and diagnostic pleural tap showed haemorrhagic fluid but

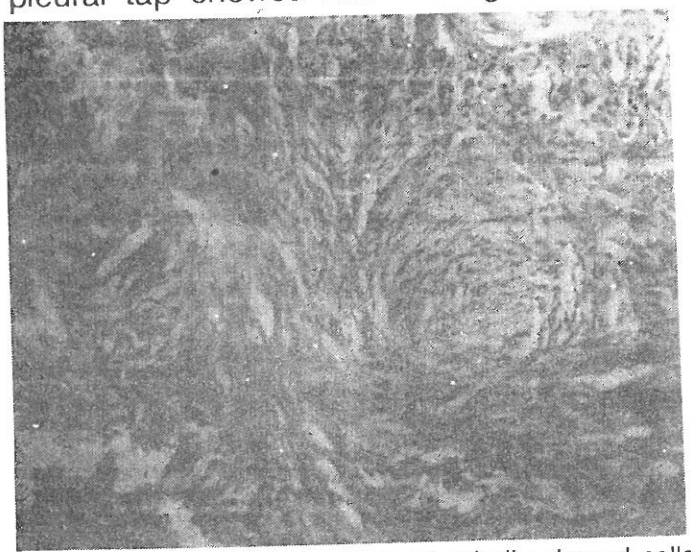

Fig. 2. Typical whorled pattern of spindle shaped cells (H and E x 100)

pap smear was inconclusive. Skin biopsy was obtained from multiple sites and on H and E staining showed thinned epidermis below which spindle-shaped cells were present in the dermis arranged in whorls and sheets at places showing cartwheel pattern (Fig. 2). The tumour was highly cellular and showed large hyperchromatic and pleomorphic nuclei and moderate amount of eosinophilic cytoplasm. Occasional mitotic figures were seen. Few polymorphs and histiocytes with haemosiderin were present. The biopsy was diagnostic of DFSP.

The patient was treated with ampicillin 500mg qid for 10 days for secondary infection. He was given 2 blood transfusions and therapeutic pleural tapping was done to relieve dyspnoea. But pleural effusion used to reform the very next day. The patient was put on anticancer chemotherapy in the form of inj. dacarbazine 250 mg per sq. metre per day intravenously for 5 days and inj. doxorubicin hydrochloride 60 mg per sq. metre as a single rapid intravenous infusion, both to be repeated after 21 days. In spite of this the patient deteriorated and died. Autopsy could not be performed.

Comments

DFSP is a slow growing tumour originating in connective tissue of dermis. Clinically it presents as firm, fibrous mass attached to skin with size varying from 0.5 cm to a large multinodular plaque of 20 cm diameter. The commonest site is thorax. In Taylor and Helwig's series[1] the highest incidence was between 20 to 40 years with male preponderance and most common location was shoulder followed by head, neck, and limbs. Trauma was mentioned as one of the aetiological factor which was also present in our case.

Considerable variations in size

accounts for multiple differential diagnoses like keloid, fibroma, desmoid, fibrosarcoma, neurofibrosarcoma, liposarcoma, sclerosing angioma, Kaposi's sarcoma and melanoma. In the present case diagnosis could not be reached clinically due to multicentricity of lesions. DFSP on histology reveals relatively uninvolved zone immediately below atrophic epidermis Interwoven fibrocellular fascicles comprising of uniform spindle-shaped cells and collagen are present in subcutaneous tissue. At the point of intersection of fascicles an acellular collagenous focus appears from which fascicles radiate giving the typical cartwheel appearance. Sometimes storiform, stillate, or whorled pattern may also be seen. Taylor and Helwig[1] stated that cartwheel configuration of tumour is highly characteristic. The present case showed typical cartwheel pattern in biopsy sections from multiple sites. McPeak et al [2] observed that high mitotic rate is ominous sign and usually associated with metastasis. The present case showed occasional mitotic figures.

DFSP is notorious for recurrences after excision. The local invasion can occur in subcutaneous fat, fascia, muscle, and bone. This tumour metastasises rarely.[3] McPeak et al[2] detected 5 cases of metastases in their series of 86 cases which were invariably fatal. Lima[3] reported 4 cases of pulmonary metastasis in his series of 300. The present case had multiple skin metastases which showed typical histology with pulmonary metastasis. Repeated filling of lungs with haemorrhagic effusion and rapid deterioration of patient indicated pulmonary metastasis.

The treatment of DFSP is surgical excision with adequate margin of at least 3 cm of normal appearing tissue. The origin of this tumour is controversial. Hushimoto et al[4] on electron microscopy demonstrated features compatible with neural cells but Lautier et al[5] after immunohistochemical study of tumour suggested fibroblastic character.

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