Quiz

Finger like growth and multiple nodules over right upper back

CASE HISTORY

A 31-year-old male agriculturist, presented to skin OPD with asymptomatic multiple nodules over right upper back of 4 years duration. The lesion started as a single skin colored nodule over right upper back which progressed to form multiple nodules within a span of 1 year. These nodules were excised in a local hospital. After 6 months, these nodules reappeared in the same site. There is no history of trauma or any systemic illness before the onset of lesions. Examination revealed large area of atrophied skin over right upper back with multiple skin-colored nodules of varying sizes from 0.5 cm to 5 cm and a finger like growth of 6 cm [Figure 1]. The posterior part of the lesion showed areas of depigmentation while anterior part showing hyperpigmentation. An ulcer was seen at the tip of the finger like growth. The surface of a large nodule showed telangiectasia. A skin biopsy from a small nodule showed typical histopathological features [Figures 2 and 3].

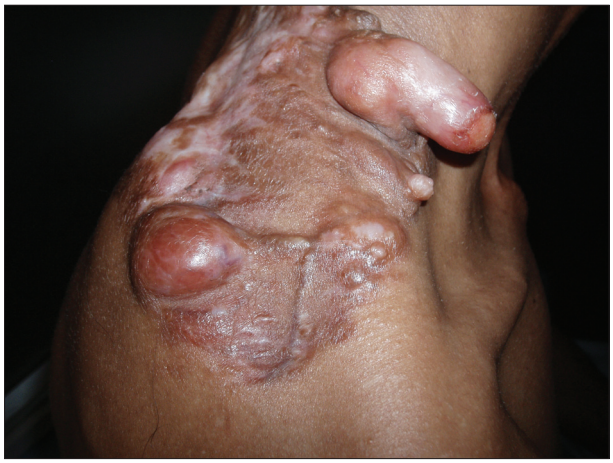

Figure 1: Finger-like growth and multiple nodules seen over upper back

WHAT IS YOUR DIAGNOSIS?

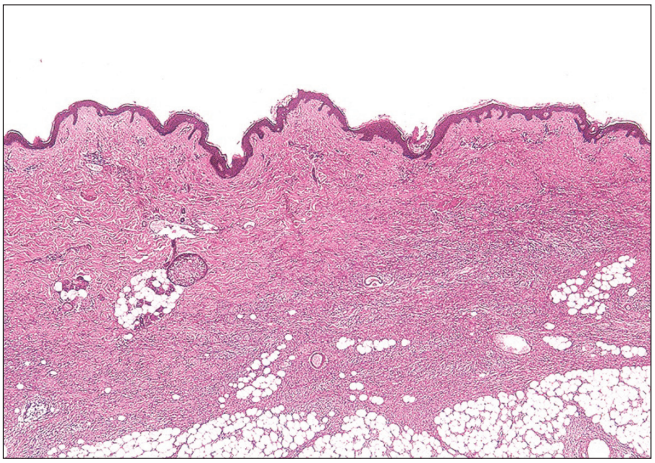

Figure 2: Epidermis showing orthokeratosis, mild acanthosis, and effacement of rete ridges. There is diffuse infiltration of spindle cells throughout dermis extending in to subcutaneous fat (H and E, ×40).

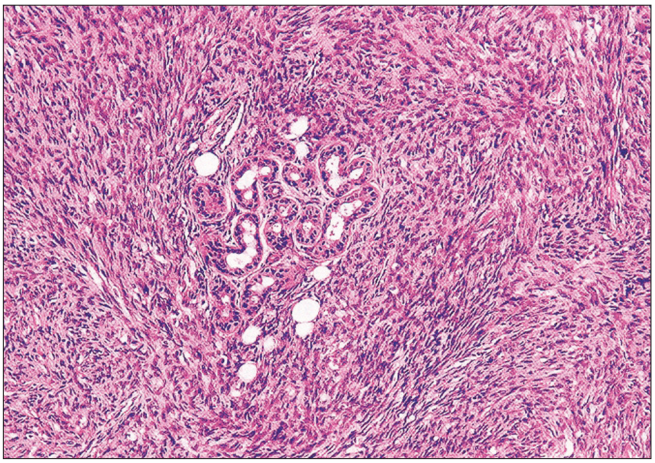

Figure 3: Storiform pattern of arrangement of spindle cells in dermis (H and E, ×200)

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DIAGNOSIS

Dermatofibrosarcoma protuberans

Histopathology of the nodule revealed typical storiform proliferation of monotonous, slender spindle cells with dark nuclei in the dermis and infiltration of the subcutaneous fat in a "honeycomb" pattern. These above histopathological features [Figures 2 and 3] and clinical appearance [Figure 1] were suggestive of dermatofibrosarcoma protuberans (DFSP).

DISCUSSION

DFSP is a soft tissue tumour of fibrous tissue, which is a slowly growing, locally aggressive tumor, characterized by marked tendency for local recurrences. Distant metastasis is rare. It typically presents in third and fourth decades with slight male predominance.[1] DFSP can also manifest at birth and childhood and diagnosis is difficult in this age group, as lesions resemble vascular birthmark.[2] Common sites of involvement are trunk and proximal extremities, but the occurrence at sites of previous trauma (including sites of vaccination, scars, and the site of an arteriovenous fistula) has been documented.[3] Tumor slowly grows and develops as a multinodular cutaneous mass, several centimeters in diameter. Rapid growth of the tumor is reported in a case.[4] Overlying skin of the tumor appears to be of reddish-blue discoloration. Polypoid tumors can be rarely seen.[5] Trunk (especially abdominal wall and chest) and lower limbs (particularly the thighs) are common sites of involvement. Involvement of hands and feet is rare.[6]

Tumor has a characteristic local recurrence and varies in different series from 20 to 50% of cases. Local recurrences can be reduced by wide excision and by Mohs surgery.[7]

Histopathology of DFSP is diagnostic. The tumor is located in dermis and there is diffuse irregular infiltration of the subcutaneous fat in a typical lace-like pattern. Cells are arranged in a storiform or "rush mat" pattern characterized by numerous whorls of cells. Hyperplasia of epidermis is rarely seen and degree of hyperplasia is inversely related to the distance of the tumor from the epidermis. Histological variants of dermatofibrosarcoma like fibrosarcomatous type, pigmented type, myxoid type, and dermatofibrosarcoma with myoid nodules can be seen. The sarcomatous changes show a fascicular "herring-bone" pattern with higher mitotic rate. In the pigmented variety, also called Bednan tumor, there are pigmented dendritic cells present in varying amounts. In the myxoid pattern, the ground substance may efface the typical storiform pattern and tumor cells appear in a stellate pattern. The myoid nodules are characterized by smooth muscle proliferation around blood vessels within the tumor.

Immunohistochemical staining shows positivty for CD 34 and negativity for other markers including Factor XIIIa, S-100, desmin, and actin.[8] Stromylesin is a useful marker in differentiating dermatofibroma from dermatofibrosarcoma as it is positive only in the former and negative in the latter.

Treatment of dermatofibrosarcoma protuberans is wide surgical excision with a margin of 2-3 cm. The recurrence rate varies from 20% to 50%. The recurrence rate can be reduced to 2% to 7% by Mohs micrographic surgery.

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