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A CASE OF DERMATOFIBROSARCOMA PROTUBERANS WITH ATYPICAL PRESENTATION

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ABSTRACT

DFSP is a locally invasive, slow-growing tumor of the subcutaneous tissue that rarely metastasizes but recurs frequently after surgical excision. This case is reported for the rare verrucous presentation of dermatofibrosarcoma protuberans occurring in a mentally retarded female with association of fibroid uterus

KEY WORDS: Dermatofibrosarcoma protuberans**,** Soft Tissue Tumour, Verrucous, Fibroid Utreus

INTRODUCTION

Dermatofibrosarcoma Protuberans (DFSP) is a locally invasive and slow-growing tumor of the subcutaneous tissue. It rarely metastasizes. It was first described in 1924 as a progressive and recurrent dermatofibroma [1]. DFSP has an annual incidence of only 0.8 cases per million and presents typically at mid-adult life with a slight male predominance [2]. The trunk and proximal extremities are the most frequent locations of the disease, but it can occur at any other site.

CASE REPORT

A 37 year old mentally retarded female with stunted growth was brought to our outpatient department by her mother with complaints of swelling over left thigh of two years duration. The lesion started as an erythematous papule two years back and has grown to a plaque of present size with multiple nodules within a period of six months. There is history of spontaneous bleeding from the nodules.

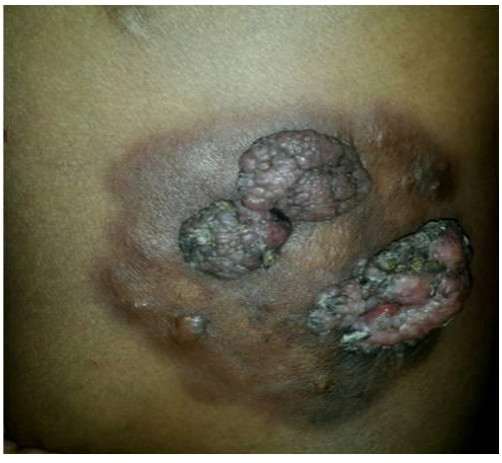
Examination revealed well defined erythematous, firm ,indurated plaque of size 7 x 6 cm over lateral aspect of left thigh(figure 1). Over this plaque were three nodules of size varying from 4 x 3 cm to 2 x 5 cm with verrucous surface.

The clinical differential diagnosis entertained were dermatofibrosarcoma protuberans, squamous cell carcinoma and neurofibroma

All routine investigations were normal. Mantoux was negative. Ultrasonogram of the left thigh showed ill-defined, irregular soft issue thickening in subcutaneous plane infiltrating adjacent muscle. Routine ultrasound abdomen revealed fibroid uterus.

Skin biopsy taken from the nodule showed densely packed monomorphic spindle shaped cells in dermis arranged in a storiform pattern(figure 2). The cells infiltrated the subcutis also. Immunohistochemistry revealed strong and diffuse staining of the spindle cells for CD34(figure 3). These features were consistent with the diagnosis of dermatofibrosarcoma protuberans. Patient was then referred to surgical oncologist and wide excision of the tumour with grafting was done.

Figure 1



DISCUSSION

Dermatofibrosarcoma protuberans is a locally aggressive sarcoma of intermediate malignancy that affects young to middle age adults. The initial presentation of dermatofibrosarcoma protuberans

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Figure 2

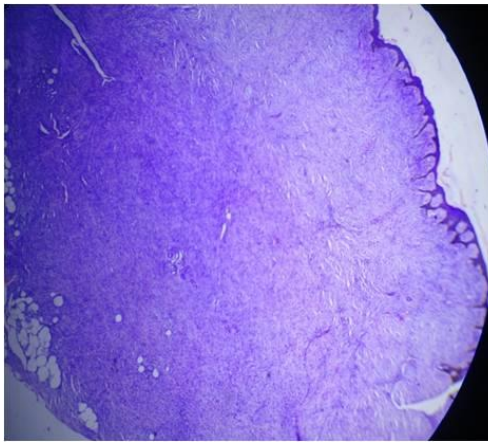
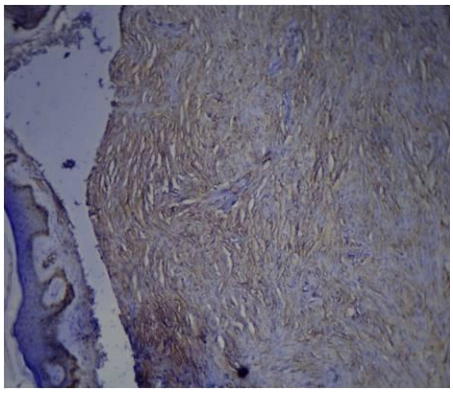


Figure 3



is described as a single raised red-bluish firm cutaneous nodular plaque with surrounding discolouration[3]. Usually the surface of the nodule is smooth, but in our case the surface is verrucous. The expression of CD34 is almost a consistent finding and it is extremely useful in differentiation of dermatofibrosarcoma protuberans from benign fibrous histiocytoma[3], dermatofibroma and other soft tissue tumours[4].

This case is reported for the rare verrucous presentation of dermatofibrosarcoma protuberans occurring in a mentally retarded female with association of fibroid uterus.

REFERENCES

1. Darier J: Dermatofibromes progressifs et recidivants ou fibrosarcomes de la peu. Ann Dermatol Syphiligr (Paris) 1924, 5:542–562

2. McLoughlin PM, Girach M, Wood GA: Dermatofibrosarcoma protuberans of the scalp. Br J Oral Maxillofac Surg 1992, 30:401–403

3. Wacker J, Khan – Durani B, HartSchah W modified Moh's micrographic surgery in the therapy of dermatofibrosarcoma protruberans analysis of 22 patientsAnn Surg Oncol 2004; 438-444.

4. Abenoza P, Lillemoe T : CD 34 And Factor X111a in the differential diagnosis of dermatofibroma and dermatofibrosarcoma Protruberans Am J Dermatopathol 1993; 429-434.