

## CASE REPORTS

### DERMATOFIBROSARCOMA PROTUBERANS

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A 29-year-old male developed dermatofibrosarcoma protuberans over the thigh. It was large, multi-nodular and asymptomatic.

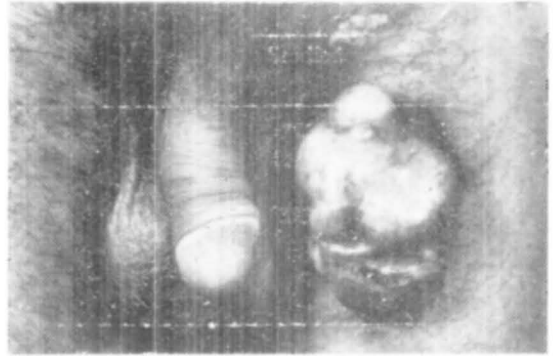
**Key words :** Dermatofibrosarcoma protuberans.

The term dermatofibrosarcoma protuberans was originally coined by Darier and Ferrand in 1924.<sup>1</sup> It is a rare, slowly-growing, locally invasive tumour arising in the dermis. Its origin is disputed. Some believe that it is histiocytic in origin,<sup>2</sup> while others think that it is fibroblastic.<sup>3</sup> Its incidence is estimated to be about 0.06% of all malignant tumours and 1.17% of all sarcomas.<sup>4</sup> It appears most commonly in the adults and males are affected more frequently than the females.<sup>4,5</sup> The commonest site of involvement is the thorax<sup>4</sup> but it has been reported on every cutaneous surface except the palms and soles.<sup>6</sup> Trauma has been considered as a predisposing factor in many cases.<sup>4,5</sup> It usually manifests as a multi-nodular, protuberant mass with ulceration in some cases. Despite its locally malignant nature, it metastasizes very rarely only and involves many organs including the lungs, brain, bones and lymph nodes.<sup>6-9</sup> Wide and deep excision is considered to be the treatment of choice.<sup>6</sup> The present case is reported because of the rarity of the condition and also because of the paucity of such cases reported from our country.

#### Case Report

A 29-year-old male had an asymptomatic growth over the left thigh of 7 years duration. It started as a small swelling, about 1 cm in size,

over the upper part of his left thigh anteriorly. There was no history of any trauma preceding the onset of the lesion. The swelling remained quiescent for 6 years, but during the last 1 year it started increasing in size rapidly and spontaneously to attain the present dimensions 7 × 6 × 3 cm. It was large, multi-nodular and pedunculated (Fig. 1). The swelling was reddish-



**Fig. 1.** A large multi-nodular mass situated over the left thigh.

brown in colour, firm in consistency and freely mobile over the underlying tissues. The skin over the swelling was stretched revealing the underlying blood vessels. The distal end of the swelling had superficial ulceration. There was a healed scar over the left inguinal region. No significant inguinal lymph adenopathy was detected. Clinically, there was no evidence of metastases anywhere. Histopathology of the biopsied tissue confirmed the diagnosis of

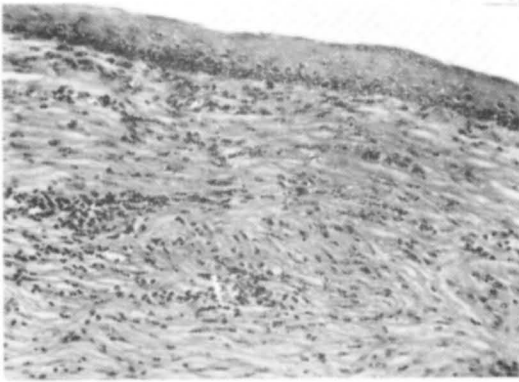
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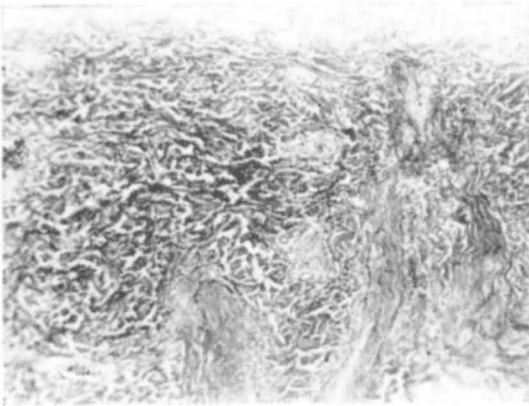
dermatofibrosarcoma protuberans (Figs. 2 & 3). Routine blood and urinalysis were normal. Liver function tests and chest X-ray were also normal. Under field-block, the tumour was excised including 1.4 cm of the normal skin surrounding the mass.

### Comments

This patient has shown characteristic features.<sup>10-11</sup> Because of the extensive local invasiveness of the tumour, inadequate excision predisposes to frequent local recurrences. Such recurrences



**Fig. 2.** Atrophy of the epidermis, cells with large spindle-shaped nuclei and non-specific inflammatory cells in the dermis.



**Fig. 3.** Characteristic storiform pattern resulting from the irregular arrangement of the fibroblasts with spindle-shaped nuclei in strands and whorls.

have been reported to occur in upto 60% of the cases.<sup>5,12,13</sup> In the series of McPeak et al,<sup>9</sup> 80% of the recurrences were observed in the first three years following excision of the lesions. It has been remarked that multiple operations always precede the appearance of metastases.<sup>6</sup> Therefore, it is essential to carefully follow-up patients of dermatofibrosarcoma protuberans for prolonged periods.

### References

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