

Short communication

Dermatofibrosarcoma protuberans

Sir, a 35-year-old lady presented with multiple elevated lesions on the lower abdomen, present over the preceding 5 years. To start with, there were a few pea-sized lesions which gradually increased in number and size to attain the present status. Cutaneous examination revealed multiple papules, plaques and nodules (ranging in size from 5mm to 7cm) over the left lower quadrant of the abdomen, superimposed on an indurated plaque. Few of the lesions were erythematous and mobile. However, some of the lesions were adherent to the underlying skin (**Figure 1**). Histopathological examination showed a hyperplastic and hyperpigmented epidermis. Besides, there was a horizontally oriented neoplasm with fibrocytic differentiation extending from upper reticular dermis to subcutis (**Figure 2**). The fibrocytes were monomorphous, elongated and spindle shaped, arranged in a storiform pattern. In addition, there were mast cells, moderate mucin and a few lymphocytes scattered throughout the fibrocytic proliferation. The cells were notable for mild to moderate nuclear atypia (**Figure 3** and **4**).



Figure 1 Multiple papules and plaques overlying a firm nodule on lower abdomen.

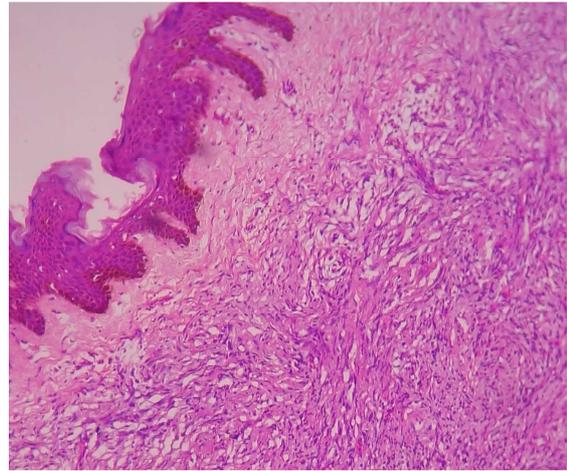


Figure 2 Hyperplastic and hyperpigmented epidermis overlying a horizontally oriented neoplasm with fibrocytic differentiation. (H&E x 100).

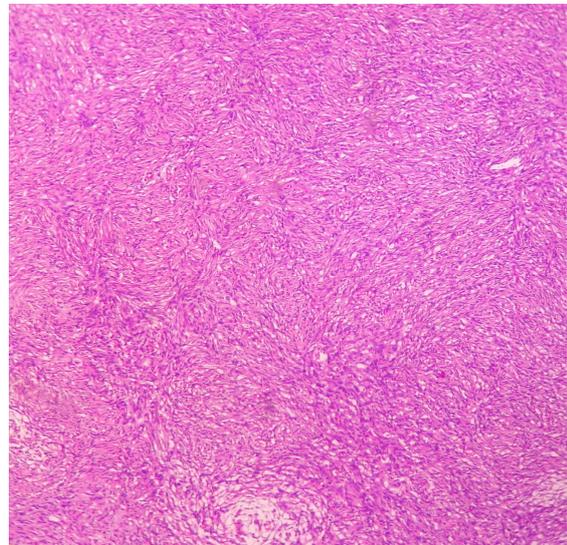


Figure 3 The fibrocytes are monomorphous, elongated and spindle shaped, and are arranged in a storiform pattern. (H&E x 100).

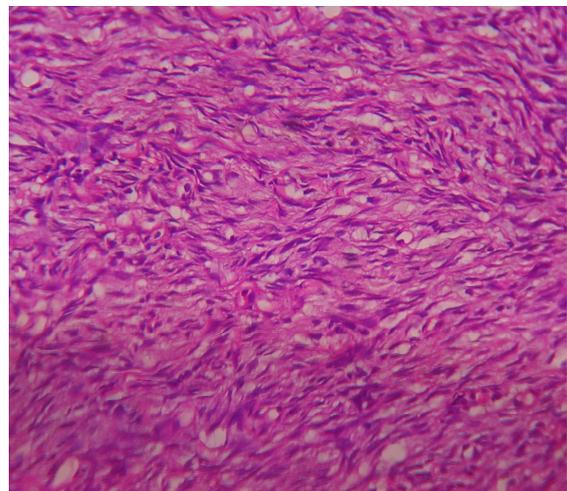


Figure 4 Higher magnification (H&E x 400).

Based on clinicopathological correlation, a diagnosis of dermatofibrosarcoma protuberans was made and she was referred to surgeon for deep excision with wide margins. The patient is under periodic follow-up to detect any recurrence at the earliest.

Dermatofibrosarcoma protuberans (DFSP) is an uncommon, slow growing, locally aggressive dermal tumor.¹Clinically, it is characterised by solitary or multiple violaceous to reddish blue papulonodules and plaques, usually over the trunk and proximal extremities.²Rarely, the lesions may be found on head and neck, pubic region and genitalia.^{3,4}Variants include confluent nodules forming a sclerotic plaque-like lesion, keloid-like sclerotic plaque, tumor-like and atrophic plaque. Histopathology is diagnostic. On immunohistochemistry, CD34 was positive. Histological variants include fibrosarcomatous type, pigmented type and myxoid type. There is a marked tendency for local recurrences, however, distant metastases is rare. Surgical excision with wide margins (2-3 cms) is the treatment of choice. For non-resectable cases, imatinib may be given.⁵

References

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Palmoplantar keratoderma progradiens et transgradiens associated with periodontitis – A case of Papillon-Lefevre syndrome

Sir, Papillon-Lefèvre syndrome (PLS) is characterized by palmoplantar keratoderma and a generalized aggressive periodontitis affecting both the primary and the permanent dentition.¹ It was first described by Papillon and Lefèvre in 1924.² It is considered to be a very rare syndrome having autosomal recessive inheritance and a prevalence of about 1-4 cases per million population. The history of parental consanguinity has been seen in 20% to 40% of the cases.³ Increased susceptibility to infections like recurrent otitis media occurs in 20-25% of the patients.¹ In this article we report an 18-year-old female with palmoplantar keratoderma associated with severe periodontitis.