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GIANT DERMATOFIBROMA – A RARE CASE REPORT

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ABSTRACT
Dermatofibroma, also known as benign fibrous histiocytoma is a soft tissue tumour that usually occurs in mid adult life and shows a slight female predominance. Giant dermatofibroma is a very rare clinical variant with only 22 cases reported in literature till date. It is characterized by an unusually large size dermal tumour, benign biological behaviour despite its large size and same histopathological characteristics as conventional dermatofibroma.

INTRODUCTION

We report a case of giant dermatofibroma in a 7 year old boy, who presented with a tender slow growing swelling on the shoulder, of four months duration.

CASE REPORT

A 7 year old presented to the OPD with complaints of a large, tender swelling over the right shoulder for past four months. The patient gave a history of surgical excision of the swelling six months back, only to recur within a span of two months. He had no preceding history of trauma or local irritation. Dermatological examination revealed a single, well circumscribed, bluish black, hemispherical nodule measuring about 7 x7 cm (Fig 1).

On palpation, it was tender, not mobile and non-pulsatile. There were no other swellings elsewhere. Routine systemic examination was normal. The tumour was excised with a wide margin and sent for histopathological examination, which revealed thinned out epidermis with effacement of rete pegs, subepidermal free zone (Grenz zone) and a tumour mass in the dermis consisting of spindle shaped cells arranged in a whorl like pattern. (Fig 2 a, b, c)

DISCUSSION AND CONCLUSION

Dermatofibroma [1-6] (Syn: Benign fibrous histiocytoma, Histiocytoma cutis, Subepidermal nodular fibrosis, Sclerosing angioma) is a common benign dermal tumour which appears as yellowish brown, slightly scaly, firm papule or nodule on the limbs. On squeezing the lesion, 'dimple sign' is seen, which indicates tethering of the lesion to the overlying epidermis. The exact aetiology is not known. But, the previous theory that they are dermal response to injury has been challenged and now cytogenetic studies favour neoplastic proliferation.

Various clinic-pathological variants of dermatofibroma have been described namely cellular, aneurismal, atypical (Pseudosarcomatous or dermatofibroma with monster cells), epitheloid and atrophic. Rarely, Giant type, eruptive (associated with immunosuppresion), ulcerated, erosive or lichenoid types can also occur .Histological findings in dermatofibroma consists of proliferation of spindle cells arranged in storiform pattern, epidermal hyperplasia, fibroblast proliferation with collagen production and infiltration of histiocytes. The features widely accepted as defining a giant dermatofibroma were first described by Requena et al [7] in their 1994 series of eight cases of dermatofibroma: (a) size >5 cm; (b) pedunculated; (c) benign biological

Figure 1. Showing a single, well circumscribed, bluish black, hemispherical nodule measuring about 7 x7 cm over right shoulder.

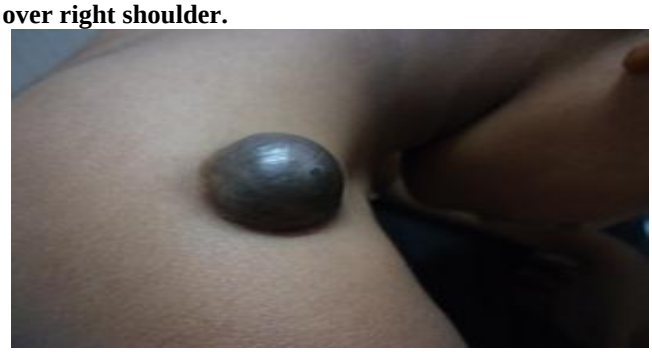


Figure 2a. Shows a thinned out epidermis with effacement of rete pegs, subepidermal free zone (Grenz zone) and a tumour mass in the dermis (Magnification X 10)

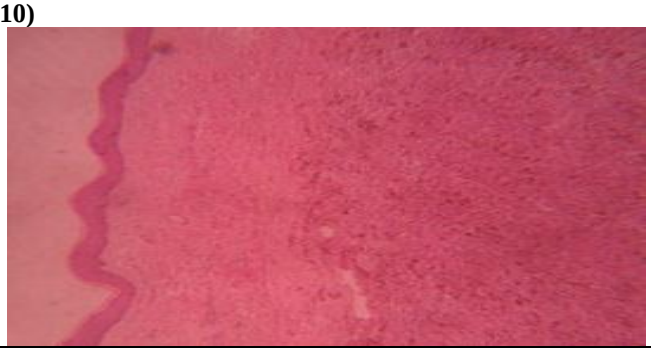


Figure 2b. Shows tumour mass in the dermis consisting of spindle shaped cells arranged in a whorl like pattern (Magnification X10)

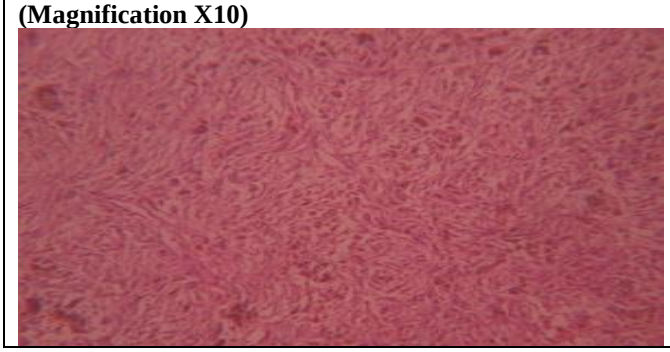
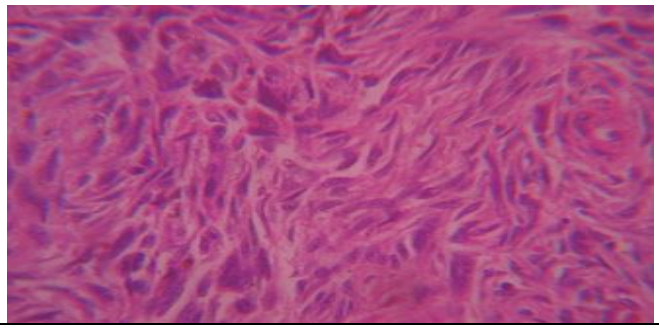


Figure 2c. Shows spindle shaped histiocytes in whorl like arrangement (Magnification X40)



behaviour despite its size; and (d) the same histopathological characteristics as conventional dermatofibroma. In the same series, none of the lesions excised recurred at an average of 35 months follow-up, suggesting that surgical management alone is satisfactory in managing giant dermatofibroma.

The differential diagnosis of dermatofibroma must include dermatofibrosarcoma protuberans and desmoid tumour. Immunohistochemical staining for CD34 is usually negative in benign lesions (positive in 85% of dermatofibrosarcoma protuberans) and may be the only distinguishing characteristic. However, it should be noted that cellular dermatofibroma may focally stain positive for CD34 though this is predominantly seen at the periphery of the tumour. Staining for factor XIIIa is positive in dermatofibroma and tends to be negative in dermatofibrosarcoma protuberans [8,9].

This case is reported because of its rarity and to differentiate it from other soft tissue tumours such as dermatofibrosarcoma protuberans and desmoid tumour.

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