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DERMATOFIBROSARCOMA PROTUBERANS OF THE ANTERIOR ABDOMINAL WALL – A RARE CASE REPORT.

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ABSTRACT

Dermatofibrosarcoma protuberans is a rare, slow-growing fibrohistiocytic neoplasm that commonly favors young to middle-aged adults. It is most commonly seen on the trunk and frequently recurs locally after an incomplete excision, but distant metastasis is rare. It is most commonly seen in the third to fifth decades but has been reported in all the age groups. We report a case of dermatofibrosarcoma protuberans on the abdominal wall.

KEYWORDS : Dermatofibrosarcoma protuberans(DFSP), Fibrohistiocytic lesion,Recurrence.

INTRODUCTION

Dermatofibrosarcoma protuberans is a locally aggressive mesenchymal neoplasm1. The lesions of this tumour present as indurated plaques above the skin surface 1,2. It was first recognized by Taylor3 in 1890 and described by Darrier4 in 1924, but the term Dermatofibrosarcoma was coined in by Hoffman5 in 1925. It has been reported to involve many body surfaces, mainly the trunk (42-72%) followed by extremities (16-30%) and less commonly in the head and neck (10-16%)6,7 . Although it constitutes less than 0.1 % of all malignant neoplasms, it represents the most common skin sarcoma (nearly 1% of all soft tissue sarcomas) more than 1% of head and neck malignant tumours and 7 % of all head and neck sarcomas 7,8,.

CASE REPORT

A 31 year-old woman presented to the surgery OPD with a swelling on the anterior abdomen which was increasing in size since past 5 years. Clinical examination revealed a 7x6 cm, firm, dark brown mass on the anterior abdomen which was tender on palpation. The mass was attached to the skin and was free from the underlying structures. There was no history of abdominal distension, pain abdomen or altered bowel habits. Past, personal and family history was not significant. There were no other complaints and the search for lymphadenopathy was negative elsewhere. CT examination revealed a large, nodular, well circumscribed, soft tissue mass, centered in the anterior abdominal wall. Clinically it was diagnosed as desmoid tumour and all the investigations were within normal limits. Wide local excision was carried out and the mass was sent for histopathological examination.

Gross findings :

Revealed a skin covered globular mass measuring 9x9x5 cms. On cut section it was capsulated and grey white. There were no areas of hemorrhage and necrosis (Figures 1 & 2).

Microscopy :

Histopathological examination, of the completely excised specimen, revealed a tumour composed of elongated cells with spindle shaped nuclei arranged in storiform pattern with minimal mitotic activity. It was diagnosed as "Dermatofibrosarcoma Protuberans", on histopathological examination(Figures 3 & 4).

DISCUSSION

Dermatofibrosarcoma protuberans is a rare low grade malignant tumour of the skin and soft tissue with high propensity for recurrence and local invasion9. It is predominantly seen in middle aged adults with a few cases reported in children10.

In some studies, a slight male pre- dominance (55-57%)11 has been reported, although in others no gender prediliction was established12. No evidence of hereditary or familial predisposition exists. Dermatofibrosarcoma is considered to be a pathologist's diagnosis more than that of a surgeons. The relatively infrequent ocurrence of DFSP lessens its clinical awareness and diagnosis is often made at histology13. So recognition to this rare tumour is important because of excellent prognosis after adequate surgical excision13,14. Soft tissue sarcomas are mesenchymal neoplasms comprising 1% of adult malignant tumours. Less than 5% of sarcomas appear as primary abdominal wall tumours15. Desmoid tumours constitute major proportion of abdominal tumours, while DFSP constitutes the least. Owing to the rarity the natural course of abdominal wall tumours is poorly understood with only a few small retrospective series representing these lesions exist15. Even though 60 years have elapsed since the recognition of DFSP as an entity the histogenesis is still unclear and is thought to be of histiocytic or neurogenic origin16. The etiopathogenesis of DFSP reveals a tranlocation between chromosomes 17 and 22 involving COL1A1 (collagen type 1a1 gene) and PDGFb (platelet derived growth factors) genes respectively17. Initial presentation is of a single red to blue, firm, blanchable, cutaneous nodule and in the later stages it appears as a multilobular mass protruding from the skin. Immunohistochemistry of DFSP has been largely unrewarding in that antisera specific α- 1 anti chymotrypsin, lysozyme and S 100 protein have been uniformly negative within the tumour cells.

Immunohistochemical studies reveal positivity for CD34 that differentiates DFSP tumour cells from normal stromal cells and dermatofibroma[18,19]. DFSP generally stains positive for CD34 and negative for S-100 protein and factor XIIIa[16]. Differential diagnosis of DFSP include dermatofibroma, atypical fibroxanthoma, malignant fibrous histiocytoma, myxoid liposarcoma and fibrous hamartoma of infancy. The standard treatment is surgical excision with wide, pathologically negative margins. Tyrosine kinase inhibitor imatinib has shown to induce regression in advanced DFSP where completely negative surgical margins are difficult to obtain.

CONCLUSION

DFSP is a rare indolent low grade sarcoma with local infiltrative tendency. Abdominal wall DFSP is a seldom seen entity. Histopathology plays a pivotal role in the diagnosis which has prognostic significance. Wide local excision with pathologically negative margins play a significant role in preventing local recurrence and distant metastasis. In young individuals DFSP must be differentiated from fibrous hamartoma of infancy, a lesion that shares similar growth patterns and cytological features.

Fig 1- Gross appearance of the specimen.

Fig 2- Cut section of the specimen.

Fig 3- Low power view.

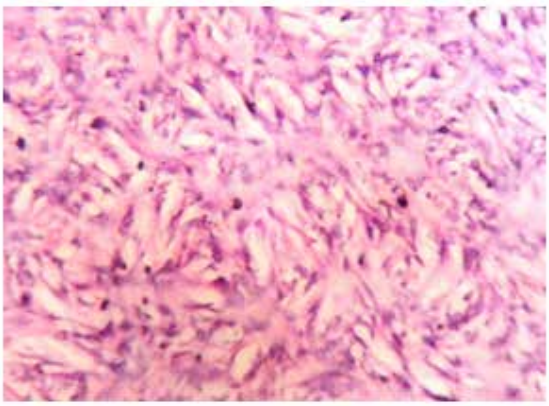

Fig 4- High power view

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