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Fine needle aspiration cytology of dermatofibrosarcoma protuberans: A rare case report

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Abstract
Introduction: Dermatofibrosarcoma protuberans (DFSP) is an uncommon cutaneous spindle cell tumor with incidence 0.8 to 5 cases/million population/year.[1] found almost exclusively in the dermis, from which they often invade the subcutaneous tissue. It is a slow-growing, locally aggressive tumor of intermediate malignancy. It poses a diagnostic challenge when encountered on fine needle aspiration cytology (FNAC) owing to its rarity and characteristics that are indistinguishable from other spindle cell lesions. It has high tendency for local recurrence but low rate of distant metastasis.
Case report: 26 years old female presented in department of surgery as a mass in the epigastric region which was painless and progressive since 6 months. Ultrasonography was done which was suggestive of neoplastic etiology with possibility of dermatofibrosarcoma protuberans. Patient was advised FNAC.
Methods: FNAC was done without local anaesthesia and smears were made. Smears were fixed with methanol and stained by PAP's stain, H&E stain and unfixed slides were stained by MGG stain. Detailed cytomorphological features were studied.
Result: Smears showed spindle cell tumor suggestive of DFSP on cytology and it was confirmed by histopathology.
Conclusion: Distinguishing dermatofibrosarcoma protuberans from other spindle cell tumors and fibrohistiocytic lesions may pose significant challenges to the pathologist. However, in the appropriate clinical setting and applying strict diagnostic criteria, fine needle aspiration cytology is a reliable tool in establishing the diagnosis of this neoplasm.

Keywords: FNAC, dermatofibrosarcoma protuberans, spindle cell tumor

Introduction
Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing, intermediate grade neoplasm [1] having incidence 0.8 to 5 cases/million population/year [2]. It commonly favors young to middle aged adults. Common site of occurrence is trunk and extremities, but can occur at almost all sites. These are found almost exclusively in the dermis, from which they often invade the subcutaneous tissue [3]. It tends to recur in up to 50% of the cases but rarely metastasizes [4]. Fine needle aspiration cytology (FNAC) is being used as a diagnostic modality for initial diagnosis as well as recurrences and metastases of soft tissue lesions. FNAC of soft tissue can be done in outpatient department and the procedure is well-tolerated, rapid and cost effective and almost without any side effects. The overall diagnostic sensitivity and specificity of soft tissue tumors diagnosis on FNAC is more than 90% [5]. Studies on FNAC of DFSP are rare [6].

Case report
26 years old female presented in department of surgery as a mass in the epigastric region of 6 months duration. The swelling was painless and progressive since 6 months. Ultrasonography was done which was suggestive of neoplastic etiology with possibility of dermatofibrosarcoma protuberans. Patient was adviced FNAC.
Patient visited cytopathology section for FNAC. On examination, an ill defined, multinodular, immobile, nontender, firm to hard mass was seen over epigastric region measuring 1.5 x 1 cm^2. (Image 1) FNAC was done with 22-guage needle and 10cc syringe. Aspirate was hemorrhagic and smears were prepared. Wet fixed smears were stained with Papanicolaou stain, Hematoxylin and Eosin (H&E) stain. Air dried smears were stained with May-Grunwald-Giemsa (MGG) stain.

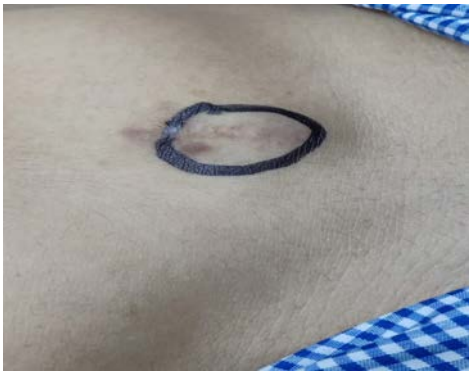


Image 1: Multinodular swelling over epigastric region.

Cytology smears showed low cellularity with spindle cells arranged in tightly cohesive bundles as well as storiform pattern. The individual cells were spindle shaped having scanty cytoplasm and show minimal nuclear atypia over haemorrhagic background. The overall cytomorphology was suggestive of spindle cell tumor, possibility of Dermatofibrosarcoma protuberans with clinical correlation (Image 2,3).Biopsy was adviced.

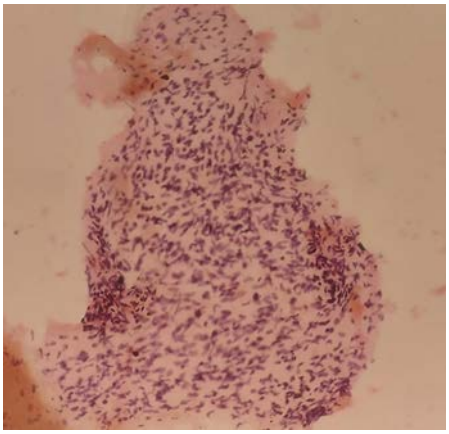


Image 2: spindle cells arranged in tightly cohesive bundles as well as storiform pattern. (H&E,MP)

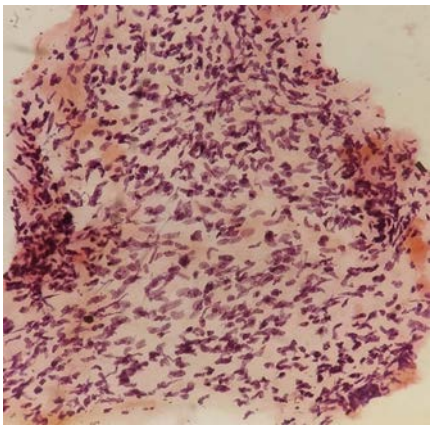


Image 3: Spindle shaped cells having scanty cytoplasm and show minimal nuclear atypia (H&E, HP)

The patient was operated in GCRI and specimen for biopsy was sent to histopathology section.Biopsy was suggestive of DFSP.(Image 4-7)

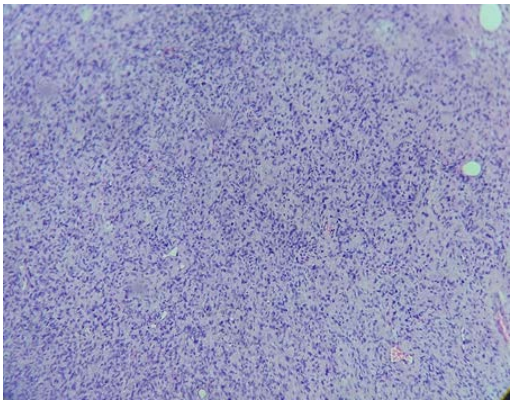


Image 4: Spindle tumor cells arranged in storiform pattern (H&E, LP)

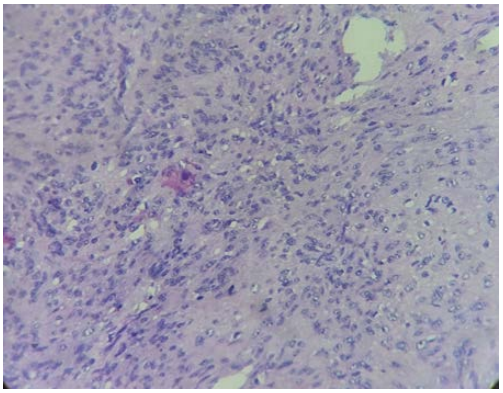
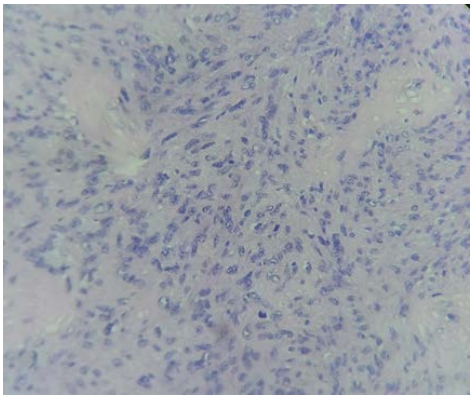


Image 5: Spindled tumour cells containing plump or elongated nuclei (H&E,LP)



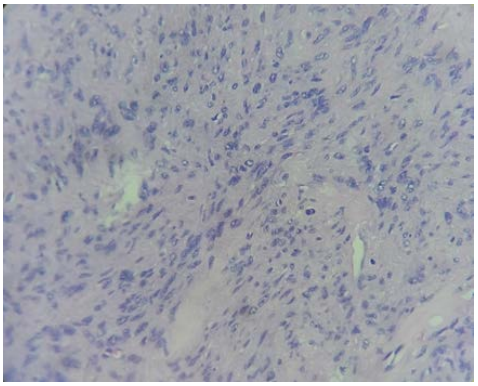


Image 5 & 6: Spindled tumor cells showing minimal atypia with 1-2 mitotic figures per high power field. (H&E, HP)

Discussion

DFSP was first described by Darriers and Ferrand as 'progressive and recurrent dermatofibroma' in 1924. Later it was named as DFSP by Hoffman in 1925 [7]. It is classified as Fibroblastic and myofibroblastic tumours with intermediate malignancy [8]. DFSP usually presents in young to middle-aged adults, with a slight male predominance [8].

DFSP is characterized by the presence of supernumerary ring chromosomes that contain the centromere of chromosome 22 and comprise interspersed sequences from chromosomes 17 and 22. Additional aberrations, such as trisomy 5 and trisomy 8, are also observed.

On FNAC Smears show fragments of variably cohesive spindle cells and dispersed single cells and stripped nuclei. There is moderate anisonucleosis, but the nuclear chromatin is granular and nucleoli are small. A streaming arrangement that vaguely suggests a storiform pattern may be seen [9].

On biopsy, DFSP is characterized by a diffuse infiltration of dermis and subcutis. It is composed of cytologically uniform spindled tumour cells containing plump or elongated wavy nuclei arranged in storiform, whorled, or cartwheel growth patterns. Cytological atypia is minimal and mitotic activity is low. The collagenous stroma contains small blood vessels. By immunohistochemistry, tumour cells stain positively for CD34 and may show expression of EMA [8].

The diagnosis of DFSP on FNAC can be challenging due to morphological overlapping of other spindle cells lesions. Studies have shown that combination of clinicopathological features and ancillary techniques are crucial in establishing a correct diagnosis.

The differential diagnoses considered were dermatofibroma, nodular fascitis, fibromatosis, neural tumor and fibrosarcoma. In the present case the smears showed monomorphic spindle cell population without any inflammatory cells, giant cells and hemosiderin -laden macrophages in contrast to dermatofibroma. The possibility of nodular fasciitis was ruled out in view of slowly growing mass, absence of inflammatory cells, pleomorphic cells and ganglion - like cells. Fibromatosis is a deeply located lesion with sparse cellularity and more collagenous stromal fragments in contrast to DFSP, hence was not consistent with the diagnosis of fibromatosis. IHC is more helpful in problematic cases. In the present case IHC studies were done on histological sections for confirmation of diagnosis.

The treatment of choice is excision with safe margin of 2-3 cms. DFSP show recurrence in 50% of the cases. Metastases are reported in only 1.5% of cases. Recurrent DFSP often behaves in a more aggressive fashion with metastasis in few cases [10].

Conclusion

Distinguishing dermatofibrosarcoma protuberans from other spindle cell tumors and fibrohistiocytic lesions may pose significant challenges to the pathologist. However, in the appropriate clinical setting and applying strict diagnostic criteria, fine needle aspiration cytology is a reliable tool in establishing the diagnosis of this neoplasm.

Conflict of Interests

The authors declare that there is no conflict of interests.

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References

1. Tumors and tumors-like proliferations of fibrous and related tissues, in Weedon's Skin Pathology, D. Weedon, Ed., 2010, pp. 833-835. Elsevier, View at: Google Scholar
2. Garcia C, Clark RE, Buchanan M. Dermatofibrosarcoma protuberans. Int J Dermatol 1996;35:867-71.
3. Rosai and Ackerman's surgical pathology 11th edition Goldblum JR. CD34 positivity in fibrosarcomas which arise in dermatofibrosarcoma protuberans. Arch Pathol Lab Med. 1995 Mar;119(3):238-41.
4. Diagnostic cytology- pranab dey 3rd edition.
5. Fine needle aspiration cytology of recurrent dermatofibrosarcoma protuberans presenting as a chest wall mass.
6. Jadhav MN, Kempula G, Haravi RM, Kittur SK, McPeak CJ, Cruz T, *et al*. Dermatofibrosarcoma protuberans: an analysis of 86 cases-five with metastasis. Ann Surg. 1967 Nov;166(5):803-16.
7. WHO Classification of Tumours, 5th Edition, Soft Tissue and Bone Tumours.
8. Orell & Sterrett's Fine Needle Aspiration Cytology 5th edition.
9. Filipowicz EA, Ventura KC, Pou AM, Logrono R. FNAC in the diagnosis of recurrent dermatofibrosarcoma protuberans of the forehead. A case report. Acta Cytol. 1999 Nov-Dec;43(6):1177-80.

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