Case Report

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FNA Diagnosis of Primary Dermatofibrosarcoma Protuberans Fibrosarcoma Variant: A Rare Entity

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is an uncommon cutaneous mesenchymal tumor of intermediate malignancy. It has progressive but destructive growth pattern with propensity for focal recurrence and rare metastasis. Studies on fine needle aspiration (FNA) of DFSP are extremely rare.

We present a case of primary DFSP-Fibrosarcoma variant over left lateral leg in a 60 year old male, which was diagnosed on FNA. An accurate cytological diagnosis is crucial in establishing the adequate therapeutic approach, avoiding mutilating surgery. Correct sub typing of DFSP in FNA smears can be difficult due to its morphological overlapping with other spindle cell lesions occurring in the skin and soft tissue. The combination of characteristic morphological features of DFSP with clinical information is crucial to establish a correct diagnosis of such rare tumor.

The goal of this report is to emphasize the role of FNA in early and accurate diagnosis in spite of many overlapping features.

Keywords: Dermatofibrosarcoma Protuberans (DFSP); FNA; Leg; Soft Tissue Sarcoma.

Introduction

Dermatofibrosarcoma protuberans is an uncommon cutaneous soft tissue neoplasm with tendency to recur but rarely metastatize[1]. It occurs at almost any site but usually in the trunk and extremities [1]. DFSP represents 1 to 6% of all soft tissue sarcomas [2]. It accounts for < 0.01% of all malignancies and 0.1% of all the cutaneous neoplasm [2,3]. The most common presentation of DFSP is painless, long standing, slow growing subcutaneous nodule [4,5].

Studies on fine needle aspiration of DFSP are extremely rare and only handful of cases has been reported in the literature [6]. Its diagnosis may be difficult to render from cytological smears, as it shares some features with other spindle cell lesions occurring in the skin and soft tissue.

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We report this case of primary DFSP- fibrosarcoma variant in view of its diagnosis on FNA. Combinations of characteristic cytological features and clinical information are crucial to establish early and correct diagnosis of DFSP to avoid mutilating surgery.

Case Report

A 60 year old male patient presented to surgical OPD of our hospital with slow growing, painless mass over lateral side of left leg since 8 months. Local examination revealed firm, nodular mass with bosselation measuring 6x4 cms with superficial ulceration over lateral side of left leg (Fig 1). The mass was adherent to overlaying skin and was free from underlying structures. Past, personal and family history was not significant. The FNA of the lesion was advised. The FNA of left leg mass done by 23 gauge needle with 10 cc syringe. hemorrhagic aspirate was obtained and stained by pap stain. The cytological smears were cellular and showed atypical spindle cells

arranged in sheets, characteristic storiform patterns, fascicles and scattered singly (Figure 2). Individual spindle cells were long, elongated having highly pleomorphic hyperchromatic nuclei with occasional prominent nucleoli with pale to basophilic cytoplasm with tapering ends (Figure 3,4). Few of the spindle cells with bizarre, atypical nuclei with multinucleate on a hemorrhagic- myxoid background (Figure 5). The final cytological diagnosis of intermediate grade spindle cell tumor – Primary DFSP-fibrosarcoma variant was rendered. Incisional biopsy of the lesion was taken and the diagnosis of DFSP was confirmed. Afterwards the patient follow up was lost and not able to receive fully excised specimen with free margins.

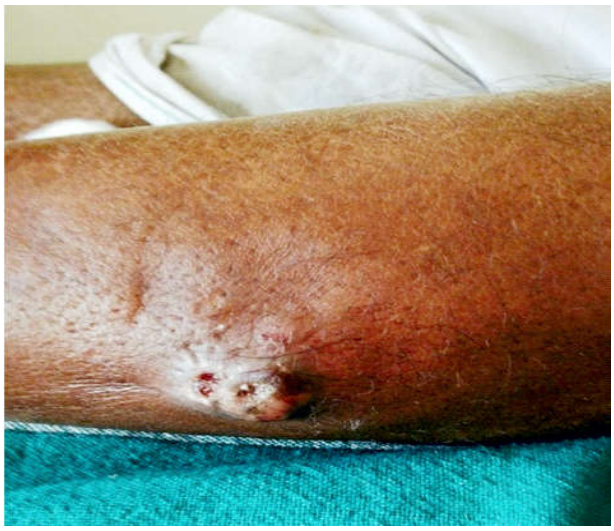

Fig. 1: Gross appearance of swelling over Left leg with multiple nodules with surface ulceration

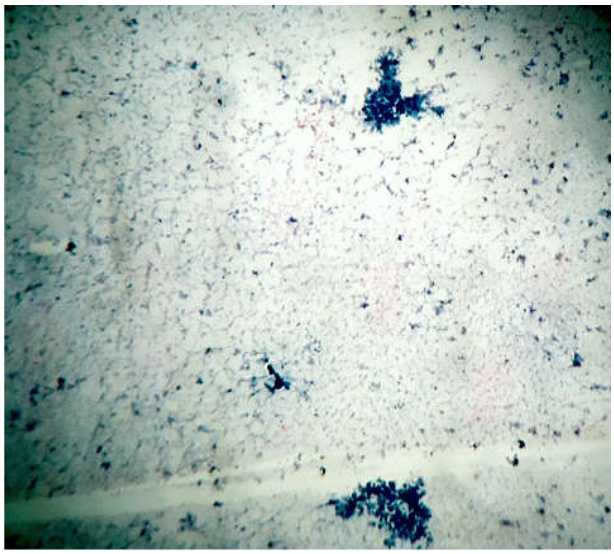

Fig. 2: Cytology showed atypical spindle cells arranged in sheets, characteristic storiform patterns, fascicles and scattered singly (Pap stain, x100)

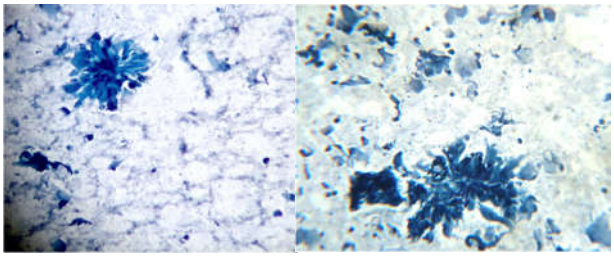

Fig. 3,4: Individual spindle cells having highly pleomorphic hyperchromatic nuclei with occasional prominent nucleoli with pale to basophilic cytoplasm with tapering ends (Pap stain, x400)

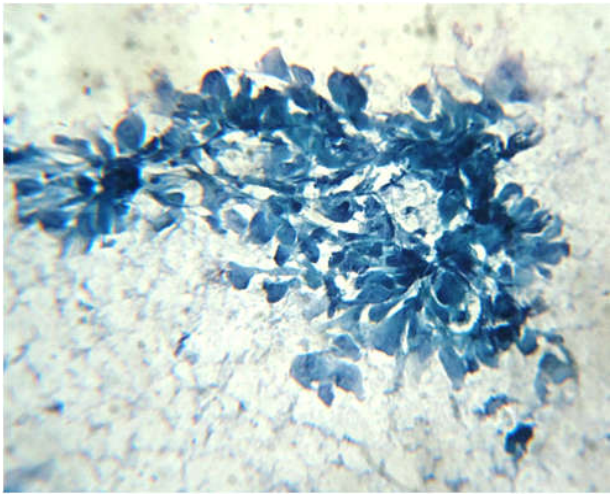

Fig. 5: Microphotograph showing spindle cells with bizarre, atypical nuclei with multinucleate on a hemorrhagic- myxoid background s (Pap stain, x400)

Discussion

Dermatofibrosarcoma protuberans is an uncommon low grade cutaneous sarcoma with a slowly progressive but destructive growth pattern with a propensity for local recurrence but metastasis is rare [6]. The standard treatment of the DFSP is excision with safe margins of 2- 3 cms [6].

Studies on fine-needle aspiration of DFSP are extremely rare; after review of literature, only 33 cases have been reported as per Klijanienko J et al [6]. DFSP shares morphological characteristics of some low grade spindle cell neoplasms. It should be differentiated from other benign, low and intermediate grade spindle neoplasm such as low grade fibrosarcoma, fibromyxosarcoma, low grade malignant peripheral nerve sheath tumor, nodular fasciitis and fibrous histiocytoma etc [6]. Hence the diagnosis of DFSP may be difficult to render from cytological smears. But due to characteristic cytological features of DFSP such as storiform , fascicular pattern of atypical spindle cells on myxoid /hemorrhagic background warrants its diagnosis. This was true

regarding this case. The combination of clinical information and characteristic cytological features is crucial to establish a correct diagnosis of DFSP [7].

Most of the times, it is said that DFSP is a pathologist's diagnosis and not the surgeon's, in view of its nature of local recurrence due to inadequate resection and rare metastasis [8]. The relatively infrequent occurrence of DFSP lessens its clinical awareness and diagnosis is often made on cytology and histology [9]. So recognition of this rare tumor is important because of excellent prognosis after adequate surgical excision [8,9].

DFSP was first described by Darier and Ferrand in 1924 and was referred to as a progressive and recurrent dermatofibroma. It was later officially termed as "Dermatofibrosarcoma Protuberans" by Hoffman in 1925 [10]. The histogenesis of DFSP is still controversial, even though it has been recognized entity for more than 60 years [11]. It is thought to be of histiocytic or neurogenic in origin [11]. There is t(17;22) involving COL1A1 (collagen type 1A1 gene) and PDGFb genes respectively [12].

Cytological preparations from DFSP cases are highly cellular and consist of large three dimensional aggregates of dispersedly placed short spindle cells with plump ovoid nuclei and scant basophilic cytoplasm. Minimal atypia with nuclei having dispersed chromatin and small nucleoli are seen. The hallmark of this condition is dispersedly placed shot spindle cells arranged in a distinct storiform pattern with minimal atypia [13]. Regarding morphological variants of DFSP,mainly–myxoid, atrophic, pigmented (Bedner),fibrosarcomatous and giant cell fibroblastoma was given in the literature [13]. In our case, characteristic features were noted with large number of atypical cells. The atypia was moderate and hence intermediate grade spindle cell tumor-Primary DFSP-fibrosarcoma variant was diagnosed on cytology.

Conclusion

Dermatofibrosarcoma protuberans is slow growing, rare, indolent low grade (mesenchymal) soft tissue tumor recognized for its progressive and locally infiltrative nature. Early and accurate diagnosis by FNA is paramount for good prognosis of the patient. The characteristic features of FNA can clearly diagnose DFSP, in spite of overlapping features with other spindle cell neoplasms. Cytological prompt and accurate diagnosis prevents the patient undergoing mutilating surgeries.

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