

Original article

Clinicopathological study of Dermatofibrosarcoma Protuberans – An experience in a rural tertiary care hospital

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Abstract:

Background: Dermatofibrosarcoma Protuberans (DFSP) is an intermediate grade soft tissue tumor with rare metastasizing potential but frequent recurrence rate.

Material and Methods: The present study is a retrospective study of the clinicopathological features of 09 cases of histopathologically proven DFSP cases which had been treated in our institute over the last 8 years (2009-2016). Follow up period was 6 months to 86months.

Results/Observation: Nine cases of DFSP were noted. The demographic details with site of appearance were studied. Treatment protocols in the form of wide local excision with or without radiotherapy were applied 03 patients showed local recurrence. DFSP has a tendency of frequent local recurrence.

Conclusion : Though DFSP is an intermediate grade tumor, owing to its tendency to recur frequently, strict follow up is recommended. Outcome is excellent.

Key words: DFSP, Intermediate, Soft tissue

Introduction

DFSP is a rare intermediate grade malignancy soft tissue malignancy having a cytogenetic alteration t(17;22) (q22;q13). Taylor in 1890^{1,2} supposedly had described the first case of DFSP. Darrier and Ferrand in 1924 classified this entity.³ However, it was Hoffman who gave the present terminology of DFSP which was addressed as progressive and recurrent dermatofibroma by Darrier and Ferrand. Studies on DFSP have progressed ever the years in a stepwise manner⁴. Taylor and Helurig⁵ in 1962 studied the histological characteristics, whereas IHC was studied in detail in 1992.^{6,7}

It is a relatively rare tumor which occurs cutaneously which is its most common presenting site.⁸ Biologically, it is a low grade tumor in most of its cases, however few of the tumors contain a high

grade fibrosarcomatous component. Owing to these characteristics along with frequent recurrence, local spread along the dermis, subcutaneous tissue and dermis, DFSP has been termed as an intermediate grade malignancy in the soft tissue tumor family.⁹

Herein, we have analyzed cases of DFSP presenting to the department of Pathology with respect to clinicopathological features and our experience regarding the same.

Aim and objectives

To evaluate the clinicopathological features of DFSP.

Materials and Method

The Present work is a retrospective, descriptive study carried out in the department of pathology of a tertiary care rural hospital over a period of 08 years (2009 to 2016). All the cases of DFSP were reviewed. Data was obtained from the Institutional

data record section and database of the department of Pathology. Demographic and clinical findings such as age, gender, location, presentation, diagnosis, treatment given and recurrence were recorded.

All the patients presenting for the first time and the recurrent cases were included in the study with recurrent cases defined as those presenting with a tumor at the same surgical site or adjacent to it within or more than 6 months of initial excision.

All the cases were confirmed on histopathology with subsequent IHC studies. Patients were followed up for a period of 6 months to 86 months. Treatment given was also evaluated. However cytogenetic

studies could not be carried out due to financial constraints.

Observation and results

We reviewed a total of 09 cases of which 5 were males, whereas 4 were females. All were adults with a mean age of 40.1 years with M:F ratio of 1.2:1. The most common site of presentation was the upper extremity that is the forearm with 04 (44.4%) cases followed by the back and trunk with 02 cases of DFSP occurring over left anterior thigh. It was the only case found over the lower extremity of the 09 cases. We came across 03 cases of recurrence.(Table 1)

Table 1.Distribution of cases according to site and presentation.

Site/ presentation	Nodular non pigmented	Nodular pigmented	Plaque like
Back	01	01	01
Forearm	03	01	00
Trunk	01	01	00
Lower extremity	01	00	00

We classified the tumors as nodular non pigmented, nodular pigmented and plaque like. Almost all the cases were nodular, whereas a single case of a patient who presented with small brownish plaque like lesions over the right scapula was noted.

Provisional diagnosis of 04 cases (44.4%) matched with the IHC finding which showed strong CD 34 positivity and factor XIIIa negativity. The remaining

05 cases (55.5%) were diagnosed as soft tissue tumors of the 4 cases of DFSP which were diagnosed on light microscopy ,02 cases were pigmented DFSP ieBednar tumor. There were 05 (55.5%) cases of conventional DFSP, 02 (22.2%) of Bednar tumor, 01 case of plaque like DFSP and single case of myxoid DFSP which recurred. (Table 2).

Table 2. Distribution according to diagnosis

Diagnosis/ provisional diagnosis	Soft tissue	DFSP	Total
Conventional	03	02	05
Bednar	00	02	02
Myxoid	01	00	01
Plaque like	01	00	01

During the followed up period, 02 patients showed recurrence 17 months and 21 months after initial excision of the tumor with tumor free margins.(Table 3)

Table 3. Distribution of recurrent cases

Diagnosis/ Recurrence	Primary recurrence	Secondary recurrence
Conventional	01	01
Bednar	00	00
Myxoid	00	01
Plaque like	00	00

Discussion

DFSP is an indolent, intermediate grade soft tissue malignancy which has its origin from the dermis and it accounts for almost 1-2% of all soft tissue sarcomas.^{10,11,12,13} It is the most frequently occurring skin sarcoma with an annual incidence ranging from 0.9 to 4.5 per million.^{14,15,16}

Typically, affecting adults in their 3rd to 4th decade and rarely children, it has a slight male predilection.¹⁷The trunk and the proximal extremities are the preferred sites of presentation followed by distal extremities and head and neck region.¹⁸ In our study we had slight male preponderance of 1.2 with mean age of presentation in the 4th decade. The most common site of presentation was the upper extremity mainly the forearm followed by the trunk region.

The tumor evolves as a slowly growing indurated plaque like lesion eventually transforming into a

reddish brown painless nodule over time. The nodule formation indicates the latter stages of tumor evolution with the largest diameter of the tumor being less than or equal to 5 mm. Among the etiologies, preceding trauma, surgical scars and even burn scars have been documented.¹⁹In our study largest tumor was 4.5 cm in maximum diameter.

Various subtypes of DFSP depending on their histological characteristics have been described. Of the various subtypes, Fibrosarcomatous DFSP shows the highest recurrence rate and metastasis. We had a single case of myxoid DFSP which recurred after 21 months post excision.

The differential diagnosis of DFSP includes inflammatoryfibrosarcoma, myofibrosarcoma, angiosarcoma, epitheloid sarcoma, myxofibro sarcoma²⁰ and myxoid tumors of soft tissue

(especially when myxoid variant of DFSP is encountered).

Histologically, DFSP typically presents as monomorphic spindle shape cells arranged in storiform pattern with fibrous stroma, Scarce mitosis, absence of necrosis, perineural invasion and lymphovascular embolization. Immunohistochemically, DFSP shows strong CD 34 positivity, variable Ki67 and p53 expression and negative factor XIIIa which differentiates it from dermatofibroma. D2-40 can also be used as marker to differentiate DFSP from dermatofibroma.^{21,22,23}

RT-PCR and FISH are the molecular biology techniques which can be used in the diagnosis of the cytogenetic abnormalities in DFSP.

Complete surgical excision is the primary treatment which is employed. However wide local excision coupled with Mohs Micrographic Surgery (MMS), have proved to give the lowest recurrence rate, in

contrary to only excision of the tumor.²⁴ MMS is employed due to the infiltrative growth pattern of DFSP and is a highly effective technique. Newer molecular targeted therapies such as PDGFBR, imatinibmesylate, etc are being employed and the results are promising.

Conclusion

Being an intermediate grade malignancy, characterized by infiltrative growth pattern high local recurrence rate and rare distinct metastasis. DFSP is a tumor to watch out for when dealing with soft tissue tumor as many a times its benign appearance may alter the diagnosis. However, the treatment modalities currently practiced and those under study are promising. Further areas of improvement could be studies dealing with improved preoperative tumor extent estimation which could help in better prognostication.

Acknowledgements:

1. Former Professor and Head of Department of Pathology, Dr.Sushama R. Desai
2. Professor and Head of Department of Pathology, Dr.Sujata R. Kanetkar

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