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# Recurrent Dermatofibrosarcoma Protuberans - An Institutional Case Series over a Span of 7 Years

**Fathima Seles. M**

Post Graduate, Institute of Pathology, Madras Medical College, Chennai-3, India

**Abstract:** ***Background: Dermatofibrosarcoma protuberans (DFSP) is a rare, superficial sarcoma of fibroblastic origin, tends to recur locally. This study demonstrates the incidence, distribution and recurrence rate of DFSP. Methods: Over a span of 7 years, from January 2012 to December 2018, 11 cases of Dermatofibrosarcoma protuberans were reportedin Tertiary hospital, Chennai. Out of which 8 cases were recurrent tumors and 3 cases were primary tumors. Results: 8 out of11 patients presented with RecurrentDFSP. One patient had history of five times recurrence over a duration of 20years. One patient presented with the lesion since childhood. Among them 5 cases were male and 3 cases were female. 7 patients underwent wide local excision and 1 patient underwent excision biopsy. Post excision reconstruction included direct closure in 4 cases, split skin graft in 1 case and local or free flap in 2cases. Of the 8 patients, one received post operativeradiotherapy. Conclusion: Wide local excision with adequate circumferential and deep resected margins can effectively control the recurrence rate.***

**Keywords:** Dermatofibrosarcoma, Dermatofibrosarcomaprotuberans, DFSP, recurrent DFSP.

## 1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare neoplasm, first reported by Darier and Ferrand in 1924[1]. Later, in 1925 Hoffman describedthe tendency of dermatofibroma to develop into dermatofibrosarcoma protuberans[2]. In 1962, Taylor and Helwig studied the histological characteristics of DFSP[3]. In 1992, immunopositivity for CD34 and immune negativity for factor XIIIa was discovered. In 1997, Simon et al identified the reciprocal translocation between chromosome 17 and 22[4]. Other terms used to describe this neoplasm are hypertrophic morphea, fibrosarcoma of skin, progressive and recurrent dermatofibroma and sarcomatous tumour[5].

DFSP is a rare malignant neoplasm accounting for <0.1% of all cancers and <2% of all soft tissue sarcoma[6]. It is a low grade neoplasm of mesenchymal origin, arising from the dermis with the tendency to recur locally and rarely metastasizes to vital organs[6]. It typically appears between 20 – 50yrs of age and has equal sex distribution. it usually occurs in the trunk (50%), extremities (35%), head & neck (6%) and DFSP of scalp occurs in <5% of the cases[7].

## 2. Methods

Over a span of 7 years, from January 2012 to December 2018, 11 cases of Dermatofibrosarcoma protuberans were reported in tertiary hospital, Chennai. Out of which 8 cases were recurrent tumors and 3 cases were primary tumors. Their medical records were assessed. Among the 11 cases reported, 72.7% cases were recurrent. The 3 cases with primary lesions were excluded and the rest 8 cases with recurrent tumor are discussed below. The mean age was 34 ± 21years (13 to 55 years). Most of them between the age of 20-40 years. The majority of the cases were presented by the male(62.5%) followed by female(37.5%). The recurrence period between the primary tumor and the current lesion ranged from 1 to 10years. The surgical procedure done for the patients with recurrent dermatofibrosarcoma are listed in the Table I:

**Table I:** Surgical procedure done for the current lesion:

| Surgical procedure for the recurrent tumor | No. of Cases |
|---|---|
| **Wide local excision + direct closure** | **3(37.5%)** |
| **Wide local excision + flap reconstruction** | **3(37.5%)** |
| **Wide local excision + split skin graft** | **1(12.5%)** |
| **Simple excision** | **1(12.5%)** |

Only one patient who presented with recurrent DFSP for 5 times over the span of 20years, underwent post operative radiotherapy.

## 3. Results

Among the 8 patients, 3patients (37.5%) presented with lesion in head and neck region, of which 2 were in the scalp. 2patients (25%) with lesion in the trunk. 1 patient (12.5%) in the upper limb and 2 patients(25%) in the lower limb (Figure 1 )

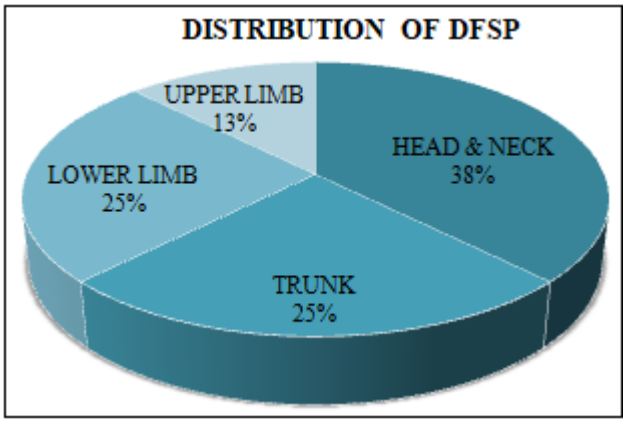


**Figure 1:** Pie chart showing the distribution of recurrent Dermatofibrosarcoma protuberans

Fine needle aspiration was done for 3 patients and was reported as spindle cell neoplasm. 5 tumors were lesser than 5 cms in dimension and 3 tumors were greater than 5cms, measuring upto 10cm in their maximum dimension. Grossly all the 8 tumors were nodular in appearance.


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The details of the eight recurrent dermatofibrosarcoma protuberans are given in table II:

**Table II:** Details of the recurrent dermatofibrosarcomaprotuberans cases

| S. NO | BX. NO | Age/Sex | Location | Duration | Current Procedure | Recurrence Period |
|---|---|---|---|---|---|---|
| 1 | 1239/12 | 22/F | Back | 3 months | Wide local excision with direct closure | 2 years |
| 2 | 2819/13 | 30/M | Rt forehead | 7 months | Excision | 3 years |
| 3 | 5014/13 | 32/F | Lt Buttock | 1 year | Wide local excision with direct closure | 5 years |
| 4 | 9032/14 | 55/M | Anterior Abdominal wall | 5 months | Wide local excision with direct closure | 8 years |
| 5 | 11360/14 | 13/M | Rt Thigh | 2 months | Wide local excision with microvascular flap | 5 years |
| 6 | 10916/16 | 36/F | Scalp | 1 month | Wide local excision with split skin graft | 1 year |
| 7 | 5634/17 | 43/M | Lt Arm | 2 months | Wide local excision with direct closure | 10 years |
| 8 | 11370/18 | 45/M | Scalp | 2 months | Wide local excision with microvascular flap | 5 times over a period of 20years |

On histopathological examination, 5 tumors were reported as the classical DFSP, 2 tumors were reported as Myxoid variant of DFSP and 1 tumor was reported as DFSP with intermediate grade malignancy.The margin status of the 8 tumors are given in the Figure 2:

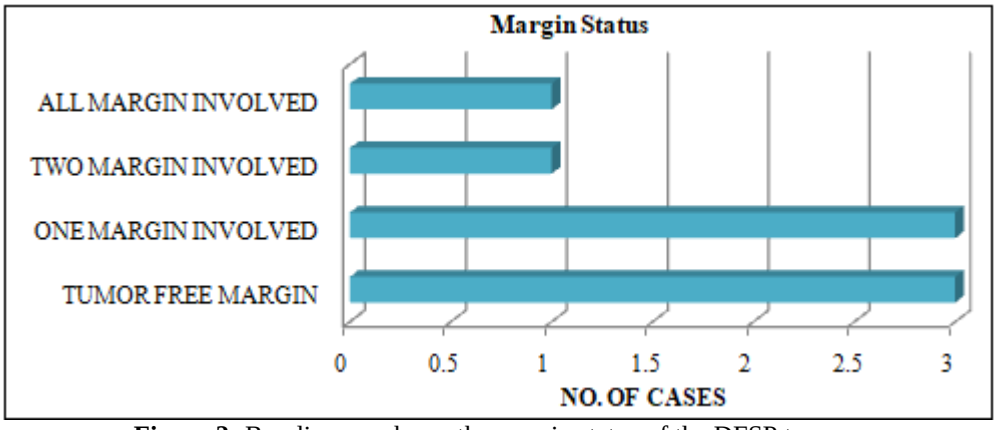


**Figure 2:** Bar diagram shows the margin status of the DFSP tumors

**Case report of the patient who presented with Multiple Episodes Of RecurrenT DFSP:**

A 45 year old man came to the OPD with complaints of swelling over the scalp on the left tempero-parietal region associated with pain and bleeding. He gave a past history of similar complaints over the occipital region 20years back for which he underwent wide local excision in the year 1998. In 2012 he developed recurrent lesion over the operated site and underwent wide local excision. In 2014, he again developed recurrent swelling and underwent wide local excision. In May 2015, he developed frontal scalp swelling, was operated and histopathology reported as dermatofibrosarcoma protuberans scalp with all resected margins positive for malignancy. Later, he completed 30 cycles of radiotherapy of 60Gray in July 2015.

On local examination, there was a 5x4cm ulceroproliferative mass in the left tempero-parietal region of scalp.

CT Brain showed evidence of heterodense mass lesion measuring 3.4x5cm noted in the left tempero-parietal region with lytic lesion involving the left frontal bone and scalloping of the outer table noted anterior to the split skin graft of parietal region. Dermatofibrosarcoma protuberans of left parietal bone with recurrent lesion in the frontal bone (Figure 3). MRI Brain showed evidence of well defined lobulated T2 iso to hypointense lesion of size 5.3(antero-posterior) x3.2(transverse) x5(cranio-caudal)cm with 7.4(antero-posterior)x 1.6(transverse)x 7.7(cranio-caudal)cm fatty component eroding the adjacent parietal bone. Report was given as features suggestive of recurrent dermatofibrosarcoma (Figure 4).

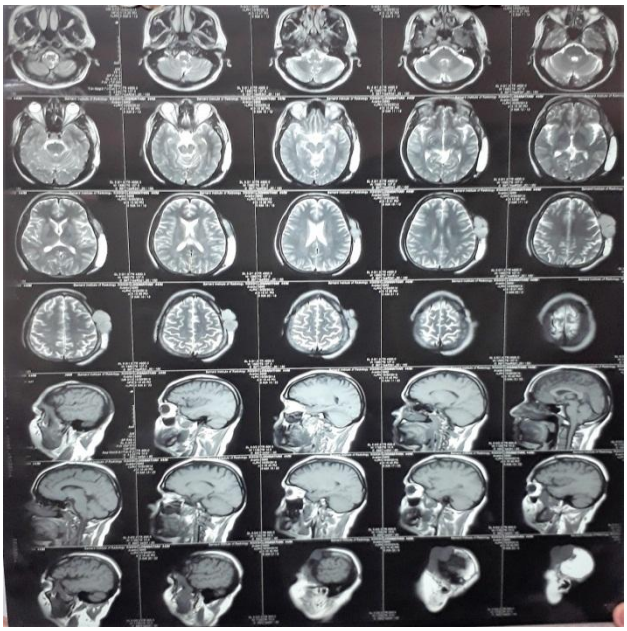

**Figure 3:** CT Brain shows heterodense mass lesion and lytic bone lesion at left tempero-parietal region


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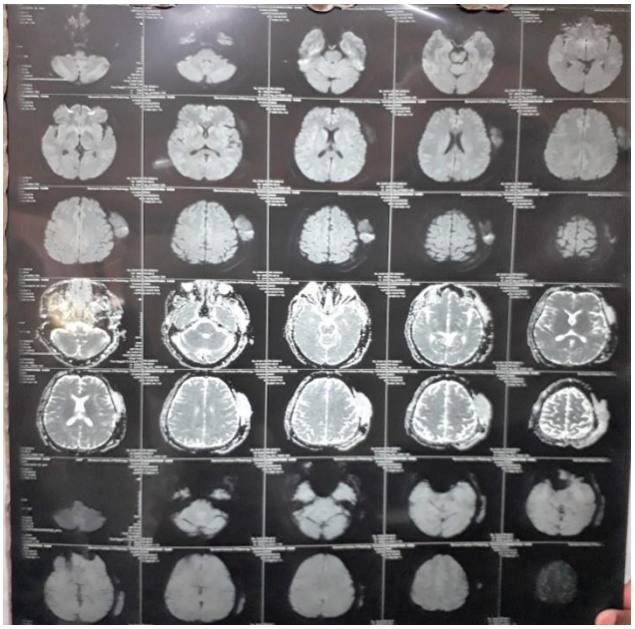

**Figure 4:** MRI Brain shows iso to hypointense mass lesion in left tempero-parietal region

Under general anesthesia, incision was made around the lesion and deepened , 5 burr holes were made in the left fronto-temporo-parietal region. Bone flap was removed along with the lesion. Inner table of left frontal region was removed, intact dura was noted and gelfoam was kept. After hemostasis was achieved, skin flap from left thigh was kept and sutured. Anastomosis was made between descending branch of lateral circumflex femoral artery to left superficial temporal artery and vena comitantes of lateral circumflex femoral artery to the left external jugular vein. The excised lesion with the underlying bone was sent for histopathological examination.

On gross examination, wide local excision specimen measured 11.5x10x4cm with attached skull bone on the posterior aspect measuring 9.5x4.5cm. External surface showed two ulcerated nodules adherent to the bone, largest measuring 4x3x1.5cm and smallest measuring 3cm in diameter. The nodules were 3cm from the anterior margin, 2.5cm from the lateral margin, 1.5cm from the medial margin and adherent to the deep bony margin. Grossly skull bone showed no tumor infiltration. (Figure 5)

**Figure 5:** On macroscopic examination
Wide local excision specimen shows two nodules

On microscopy, section showed skin with underlying dermis showing a neoplasm composed of spindle shaped cells arranged in storiform pattern, fascicles and sheets. The cells are spindle shaped with scant to moderate eosinophilic cytoplasm, plump ovoid to spindle shaped nucleus with mild to moderate atypia. 1-2 mitosis per high power field. Tumor cells are interspersed with thin strands of collagen and infiltrates the subcutaneous fat (Figure 6). The stroma shows necrosis in occasional foci. All margins are free from tumor infiltration. Immunohistochemical (IHC) staining revealed diffuse strong positivity for CD34 (figure 7), diffuse positivity for vimentin (figure 8), focal scattered positivity for CD68 (figure 9) and negativity for S100.

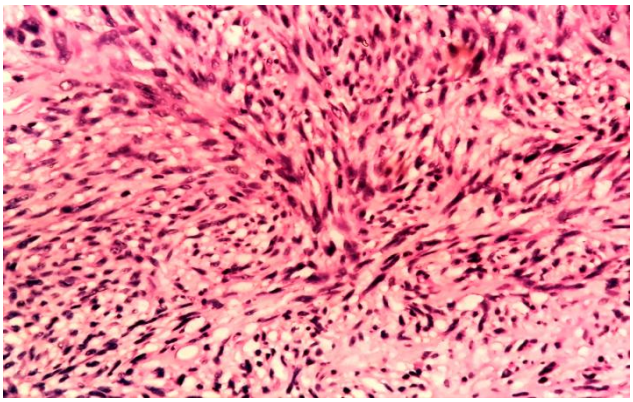

**Figure 6:** Microscopically, on H&E staining spindle shaped cells arranged in storiform pattern [100X]

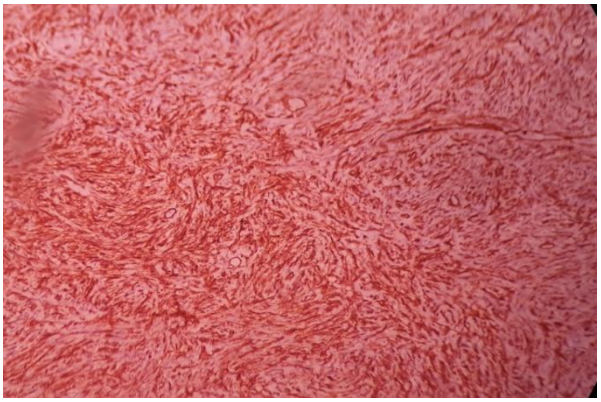

Figure 7: IHC staining of CD34 shows diffuse strong positivity [40X]

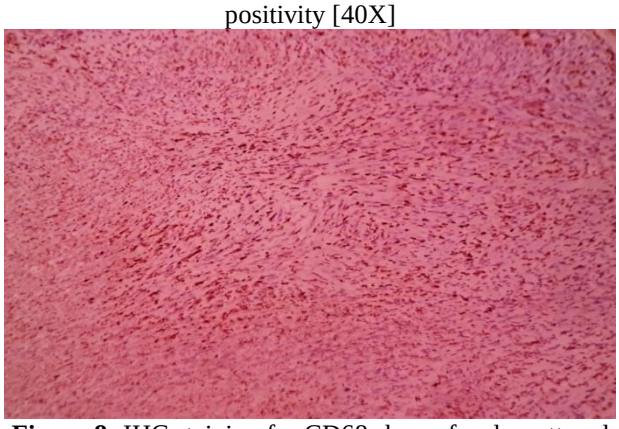

**Figure 8:** IHC staining for CD68 shows focal scattered positivity [40X]


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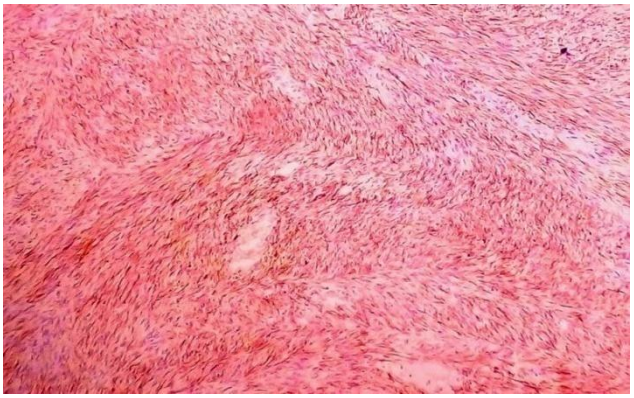

**Figure 9:** IHC staining for Vimentin shows diffuse positivity [40X]

## 4. Discussion

DFSP is a rare malignant neoplasm accounting for <0.1% of all cancers and <2% of all soft tissue sarcoma[6]. It is a low grade neoplasm of mesenchymal origin, arising from the dermis with the tendency to recur locally and rarely metastasizes to vital organs[6]. It typically appears between 20 – 50yrs of age and has equal sex distribution. it usually occurs in the trunk (50%), extremities (35%), head & neck (6%) and DFSP of scalp occurs in <5% of the cases[7].

DFSP presents as asymptomatic purple to pink, plaque or nodule with history of slow but persistent growth[8]. It gradually enlarges and evolves into ulcerative protuberant tumor[9]. Scalp fixation may occur in early stages caused by periosteal attachment due to thickness of skin and subcutaneous tissue in this area[9].

Ultrasonography shows a well marginated, slightly lobulated, hypoechoic matrix with discrete areas of increased hypoechogenicity and small echogenic foci and posterior enhancement[10]. CT and MRI are used to determine the the extension of larger masses, myxoid degeneration, presence and degree of fibrosarcomatous change[10].

Fine needle aspirate from DFSP shows cellular smear with clusters of spindle shaped cells embedded in a collagenous, occasionally myxoid matrix and fascicles of spindle cells in storiform pattern maybe seen[11].

Histopathology shows well circumscribed tumor consisting of slender, pleomorphic spindle cells arranged in cartwheel-like or storiform growth pattern, inconspicuous vascular network, low mitotic activity and absence of necrosis. The tumor cells arranged around foci of collagen or vascular spaces, radiating outward like the spokes of a wheel. Infiltration around fat cells may produce a honeycomb appearance of the fat[12].

Immunohistochemistry show positivity to CD34 and negativity to s100, desmin& factor XIIIa[13].The histologic subtypes are pigmented DFSP (Bednar tumor), granular cell variant of DFSP, sclerosing DFSP, atrophic DFSP, giant cell fibroblastoma(GCF) and fibrosarcomatous DFSP[14].

American Musculoskeletal Tumor Society Staging System is widely used. Stage IA tumors are low grade, intracompartmental and Stage IB tumors are low grade, extracompartmental[15]. Ugurel et al proposed a staging system according to German guidelines. Stage I is primary tumor stage, Stage II is primary tumor with regional lymph node metastasis and Stage III is tumor with distant metastasis[16].

Cytogenetically, DFSP is characterized by the presence of a reciprocal translocation t(17;22)(q22;q13). As a consequence of this translocation, fusion occurs between the gene for collagen type I alpha 1 (COL1A1) on chromosome 17q and the gene encoding platelet-derived growth factor beta polypeptide (PDGFB) on chromosome 22q[17].

Treatment options include complete surgical excision, including wide excision and Mohs micrographic surgery, radiation, and imatinib mesylate[18]. Mohs micrographic surgery (MMS) is recommended compared to wide local excision (WLE)[19]. Studies showed recurrence rate upto 6.6% after MMS whereas increased recurrence rates of 11-35% after WLE[19]. Since regional metastasis is very rare, prophylactic lymph node dissection is not recommended[20]. DFSP is highly radiosensitive, therefore adjuvant radiotheraphy is recommended. Imatinib can also be administered if translocation of chromosome 17 and 22 is present.

DFSP is often misdiagnosed as benign lesion and inadequately excisied leading to local recurrence[21]. Risk factors for DFSP recurrences are age >50years, margin <1mm/ positive margin status, increased cellularity, high mitotic rate and fibrosarcomatous change[22]. Due to increased rates of local recurrence of DFSP, regular follow-up of the primary site must be conducted once in every 6-12months and biopsy of any suspicious lesion[23].

Metastasis is rare and have been reported in 1-6% of cases. Recurrent tumors have the highest risk of metastasis and lung is the most common site, via hematogenous spread[24].

## 5. Conclusion

Dermatofibrosarcoma protuberans is a very rare sarcoma which is locally aggressive in nature. It can be often misdiagnosed and inadequately excised leading to increased rates of local recurrence. Therefore proper diagnosis, adequate excision should be ensured and adjuvant radiotherapy is recommended to prevent recurrence.

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