Recurrent Dermatofibrosarcoma Protuberans of the Scalp: How wide should we go?

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Abstract

Objective: Dermatofibrosarcoma protuberans (DFSP) is a rare dermal tumor recurring after inadequate primary treatment. This tumour mostly located in the trunk region and DFSP of the scalp is very rare but well-known clinical entity. The preferred treatment of DFSP is wide resection, namely with margins of 3 cm beyond the evident disease and histological negative margins. All presented scalp cases in the literature were residual tumours.

Objective: To describe a case of DFSP of the scalp treated with wide excision including periosteum and outer layer of the cranium and reconstructed with skin grafting.

Method: In our patient a soft tissue tumor located on the scalp measured 11 cm X 9 cm X 5 cm. Neither cranium nor regional lymph nodes were affected. There was no distant metastasis. The tumor was excised with a 3 cm safety margin in the axial plan and the outer layer of the cranium was also shaved.

Results: The patient was followed up for 8 years and no recurrence or metastasis was detected.

Conclusion: Although skin grafting technique was not accepted an ideal reconstruction alternative of the scalp, skin grafting would be a good choice for monitoring of the possible recurrences rapidly and easily. Excision or shaving of the outer layer of the cranium should be included the specimen.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is considered a low grade dermal sarcomal malignant tumour of the skin[1]. DFSP has an uncertain histological origin. Trauma [2], immunizations[3], arsenic[4] were presented in the pathogenesis of the DFSP. Cytogenetic observations were done and some chromosomal anomalies were reported in etiology of the DFSP in the literature[5]. Specific COL1A1-PDGFB fusion genes were present in the cells of a DFSP case with an unusual chromosomal abnormality, a marker chromosome composed of segments from chromosomes 7, 8, 17, 21, and 22[6]. It is slow growing and locally aggressive spreading, rarely metastasizing to the regional node or distant sites and frequently recurs after resection, presumably owing to infiltrating tumor projections away from the central tumor mass[7]. Clinically, the tumor appears as an indurate plaque or nodule that may be violaceous, red-brown, or flesh-colored. 50%–60% of these lesions occur on the trunk, with less common involvement of the proximal extremities, the head, and neck[8]. DFSP is usually affixed to overlying skin and is firm and indurated. It can sometimes be attached to deeper structures, such as muscle and fascia[9]. The disease course is slowly progressive, with pain becoming increasingly prominent as the lesion increases in size. DFSP characteristically appears in the younger age group (20 –50 years), but it may also affect children and the elderly[10]. In our study, we described a female patient with residue DFSP on her scalp.

CASE REPORT

A 51-year-old female was referred to the Plastic and Reconstructive Surgery Department in Izmir Ataturk Training Hospital in 1993. She had a 3 year history of a large gradually swelling of the right temporal region of her scalp. The swelling was non-tender, firm, non transilluminant and non pulsatile. Interrogation of the patient and examination of the records revealed that she had undergone surgery 2 times during the past three years for

recurrent scalp tumour. Local flaps were used in her first operation. Recurrence was detected after 1 year of operation. Re-excision and skin graft technique was applied in her second operation.

Examination of the patient revealed a scalp tumor 11 cm X 9 cm X 5 cm in dimensions (figure 1 a and b). The tumour was tethered to the overlying skin. Systemic examination was unremarkable. No lymph node in the neck, occipital region, or axilla palpable. Her routine laboratory tests were normal. Radiological examination of the skull with plain X-Rays revealed periosteum reactions on the base of the mass. CT scan was performed. Soft tissue mass in right temporo-parietal region was detected in CT scan without bone involvement and intracranial extensions.

The patient was operated under the general anesthesia. The tumor was excised taking three cm of normal skin margin. The periosteum of the scalp was excised and the tabula externa of the cranium was shaved with a chisel. Primary cranioplasty was not done. Split thickness skin grafting was applied for the closure of the defects.

On histological examination, the tumor consisted of infiltrative spindle-shaped cells. The tumor cells were intermediate in size with pleomorphic vesicular nuclei, inconspicuous nucleoli, and variable cytoplasm; mitotic activity was greater than 5 per 10 high-power fields, and vascularity was prominent. There was a marked increase of cellularity in the microscopical examination of the tumor (figure 2). Surgical margins were tumor free. There was no early or late complication in post-operative period. Neither chemotherapy nor radiotherapy was applied to patient. Patient was followed 8 years. There was no evidence of re-growth on follow up after 8 years (figure 3). Patient did not accept the cosmetically reconstruction with tissue expander.

Figure 1

Figure 1 a and b: Pre-operative view of the patient.

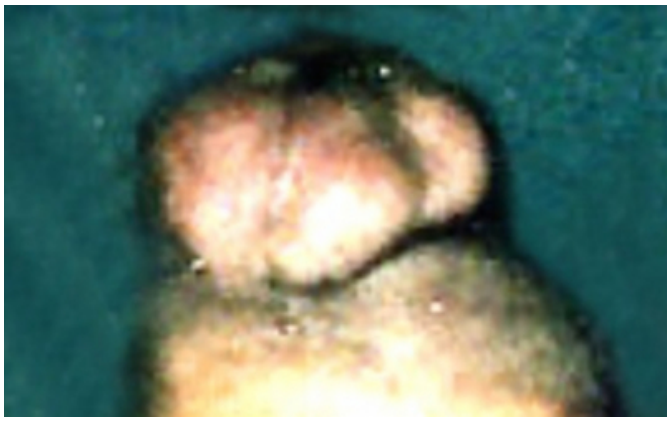


Figure 2

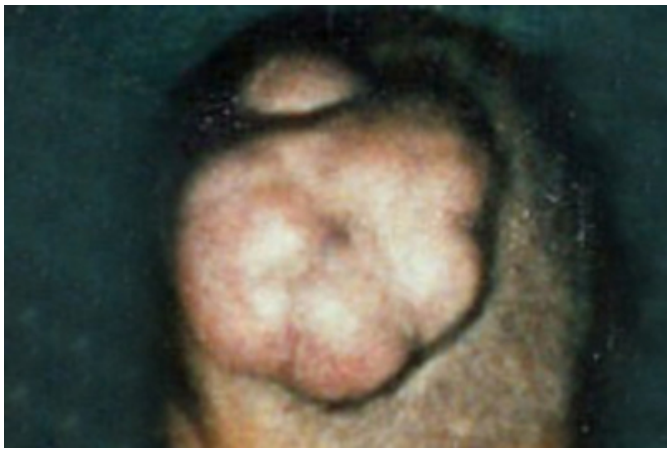


Figure 3

Figure 2: Microscopically view of the specimen (Hematoxylene Eosin staining, X 100 magnification)

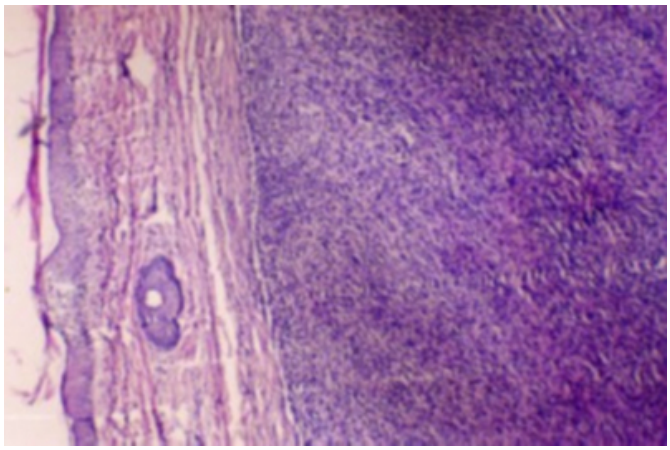
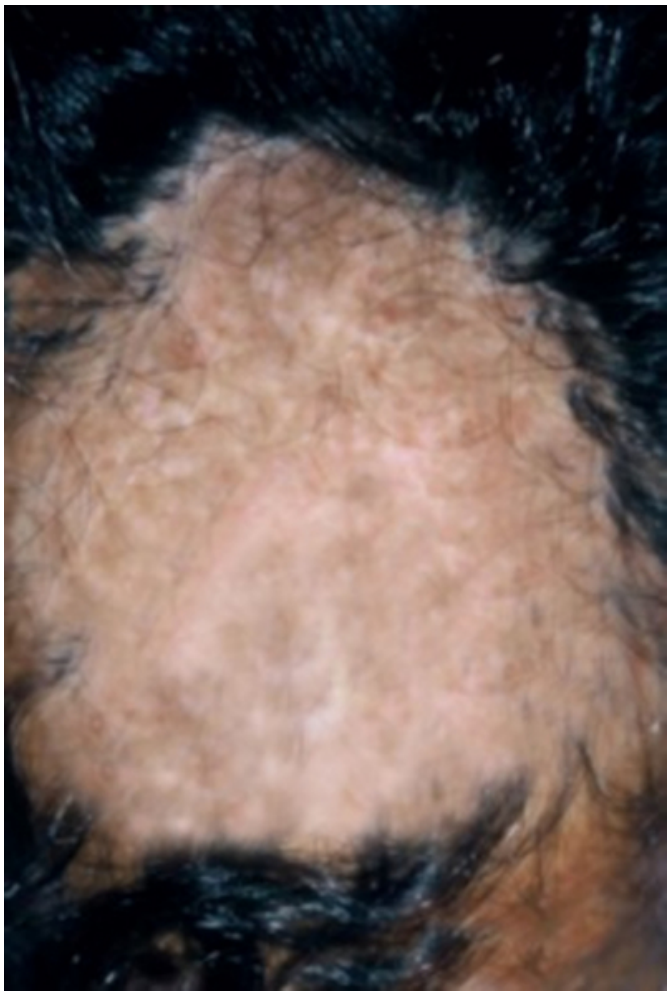


Figure 4

Figure 3: Post-operative 8 years view of the patient.



DISCUSSION

Dermatofibrosarcoma protuberans of the scalp is a very rare but well known clinical entity. D'Andree et al summarized the historical backgrounds of this entity in their study and according to their report Taylor first described a potentially relapsing neoplasia similar to a keloid. However, clinical classification was done by Darier and Ferrand in 1924 and finally by Hoffman who first named this tumour DFSP in 1924[11]. Wrotonski et al described this tumour as a cutaneous equivalent of malignant extracutaneous soft tissue histiocytoma[12]. DFSP has some synonyms as: hypertrophic morphea, progressive and recurrent dermatofibroma fibrosarcoma of skin and sarcamatous tumour resmbling keloid[13]. Bednar tumor is a rare pigmented variant of DFSP[14]. Also familial [15] and congenital[16] DFSP were described in the literature. This tumour is mostly located in the trunk region and vulva[17], penis[18], toe[19], breast[20], jejunum[21] were the rare reported sites of the tumour. Less than 70 cases with scalp DFSP were reported in the literature[13], [22,23,24,25].

The scalp and neck-supraclavicular fossa are the most common sites in the head and neck region. Sarcomas account for less than 1% of all head and neck malignancies. Mark et al reported that 7% of all sarcomas in head and neck region had the diagnosis of dermatofibrosarcoma protuberans after pathologic review[26].

Three major characteristic features of this tumour were listed by D'Andree et. al[11]:

1. The difficult histopathological classification,
2. The high tendency to recur after incomplete surgical excision
3. The potential progression into fibrosarcoma with disseminating metastasis.

Surgery is the treatment of choice for DFSP and the most significant prognostic factor has proved to be the extent of resection. In selected patients, Mohs micrographic surgery may further reduce local recurrence rate and an effective treatment of DFSP[27]. Classically, the preferred treatment of DFSP is wide resection, namely, margins 3 cm beyond the evident disease and histologically negative margins. But Arnaud et al[1] noted that it is important to excise a tumour with 5 cm margin during the initial procedure to improve treatment results of DFSP. Radiotherapy has been reported not to be successful as primary or adjuvant therapy for DFSP elsewhere on the body [28] but Suit et al concluded that Radiation in well-tolerated dose schedules is an effective option in the management of patients with DFSP[29].

Chemotherapy has not been reported as a therapy for head and neck DFSP. Ratios of the local recurrences of the tumors were reported as high as 49 to 80 % [30]. Especially the scalp is a critical anatomical region for DFSP because of its relationship with the brain. Das et al presented a case with intracranial extension of DFSP[24]. Their patient was operated three times. Since intracranial invasion would not be surprised, adequate treatment with aggressive approach should be performed in scalp DFSP. A major problem is the reconstruction of the scalp. The ideal scalp replacement is durable, the same thickness as the remaining scalp, hair-bearing, and reliable enough to allow for timely post-operative adjuvant therapy[31]. Early reports presented scalp reconstruction with skin graft and local flaps[32]. After the 70's, free tissue transfer for reconstruction of large scalp defects became popular[31]. Taniguchi et al presented a case with scalp DFSP with brain metastasis[33]. They performed a

wide excision and reconstruction with a latissimus dorsi myocutaneous flap. They concluded that a simple excision should not be performed as the initial treatment for DFSP of the head. They promoted a free flap reconstruction in scalp DFSP. But this technique requires technical expertise, more time and more intensive care in post-operative period. And free tissue transfer generally results in a hairless reconstruction. Skin grafts have major disadvantages such as missing hair and are prone the ulceration. Local flaps are more suitable for the reconstruction since they have similar characteristic features with the defect area but their relative inelasticity feature limits reconstruction without skin grafting of donor site.

Tissue expansion is well described for coverage of large scalp defects and offers the advantages of replacement with hair bearing skin[34]. But this technique is not suitable in early reconstruction of malignant tumors of scalp. Since the skin graft is very thin, visualization of any recurrences would be faster recognized in after reconstruction of the defect. We believe that skin grafting technique can be classified as unfavorable alternative in scalp reconstruction because of their disadvantages. But monitoring of the tumour site is so critical in scalp DFSP because of the high recurrence rate and the possibility of intracranial extension. Çakir et al managed their patient who had biggest DFSP on the scalp in the literature, with skin grafting because of the high possibility of recurrence[35]. We speculated treatment of the residue DFSP on the scalp should be treated with aggressive tumor excision and reconstruction with skin graft if there is a suitable donor site for the skin. If the cranium was involved and/or intracranial extension were detected a wide excision including the affected cranium, cranioplasty and reconstruction with free tissue transfer are the most suitable treatment approach.

But management of a scalp DFSP without local invasion is still controversial. Interestingly, all reports which presented scalp DFSP as a case report were residual tumours after incomplete treatment. Since there is no standard for pre or post-operative adjuvant therapy, incomplete treatment can only be explained with incomplete resection of the tumour in spite of tumor free margins. Diameters of the safety margins were discussed in the literature by the different authors but there is no data related to the depth of resection. We speculated that simple excision with 3 cm margins might not be the proper treatment in these patients. Excision of the periosteum of the cranium with shaving of the outer layer should be the first treatment. Reconstruction of the defect could be done with skin grafting for detection of the recurrence in the early period. Patients should be followed for a long time for recurrence and metastasis. After appropriate time, scalp reconstruction can be done for cosmetic reasons with tissue expanders.

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