# Multi-disciplinary Approach of a Pediatric Dermatofibrosarcoma Protuberans of the Scalp with Slow Mohs Micrographic Surgery and a Double Rotational-Advancement Scalp Flap

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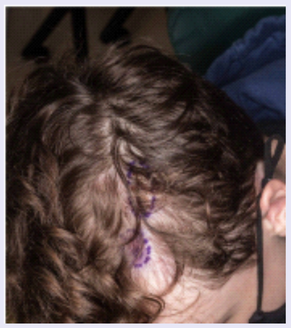
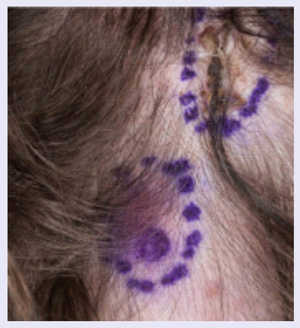
## Introduction

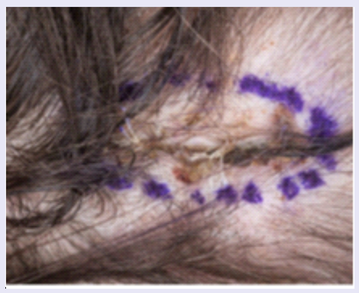
Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive soft-tissue tumor that accounts for 1-6% of all sarcomas. In patients under the age of 20, incidence is less than 1 in 1,000,000. [1,2]. DFSP most commonly presents on the trunk and extremities, with only 13% of cases arising in the head and neck region [2]. It is a slow-growing, locally aggressive malignancy that originates in the dermis and has low metastatic potential but is notable for high rates of local recurrence due to local invasion [3,4].

## Case Presentation

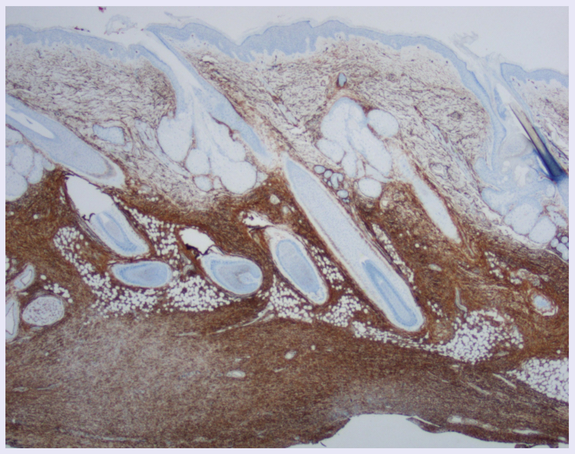
- An otherwise healthy 14-year-old male first noticed a non-tender swelling on his head present for 5 months.
- Initial differential diagnosis included trichilemmal cyst, atypical-appearing epidermal inclusion cyst, lipoma, fibrous lesion, or small complex fluid collection.

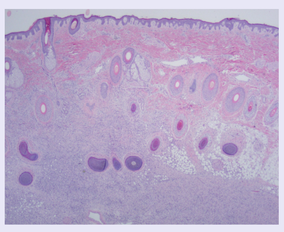


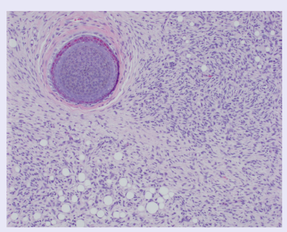




**Figures 1, 2 and 3:** Pictures of patient upon presentation to dermatology at NMCP (top left). 1.5 cm x 3.5 cm linear scar with sutures in place on left parietal scalp from previous lesion removal (bottom) as well as an adjacent 1.7 cm x 2.2 cm depressed plaque with atrophic skin and sparse hair (top right)







**Figures 4, 5, and 6:** (Top) CD34 staining showing strong CD34 positivity. (Bottom) H&E stain low power (left) and high power (right). Histopathologic exam significant for positivity for platelet derived growth factor subunit B (PDGFB) by fluorescence in situ hybridization (FISH) throughout the tumor. Of particular concern was a central cellular area with ovoid cells showing high nuclear to cytoplasmic ratios, speckled nuclear chromatin, and poorly-defined cytoplasmic borders. This region showed up to 3 mitoses per high-power field and a 10% proliferative index by Ki-67 staining. This area was felt to be concerning for high grade transformation, and DFSP with fibrosarcomatous transformation was diagnosed. Histologically, DFSP is characterized by relatively uniform spindled cells that show areas of storiform growth pattern and “honeycomb-like” infiltration of adipocytes.

## Discussion

- Varied approaches for excision, including Mohs micrographic surgery (MMS) as well as wide local excision (WLE) [5].
- Traditionally, excision of these lesions is carried out by WLE which has reported recurrence rates of 22%-47% [6].
- Recurrence rates can be high with WLE due to characteristic “Finger-like” projections of DFSP.
- Several studies have compared WLE to Mohs Micrographic Surgery (MMS), which report recurrence rates with MMS at 3% [3].

## Discussion Continued

- Verbruggen et al reported complete resection in 75% of patients treated for Head and Neck DFSP using Slow-Mohs micrographic surgery [7].
- Mohs micrographic surgery uses tangential sectioning of the peripheral and deep margins of the tumor [8].
- This allows examination of virtually 100% of tumor margins and gives the surgeon a clear three-dimensional image of the extent of the tumor, including the tentacle like projections common in DFSP.
- This unique approach has been shown to decrease the rates of recurrence in several studies [3,7-8].

## Reconstruction

- Defect of 97.75 $cm^2$ after clear margins obtained.
- Previously undescribed use of a double rotational-advancement scalp flap to reconstruct the defect [4,9,10,11,21].

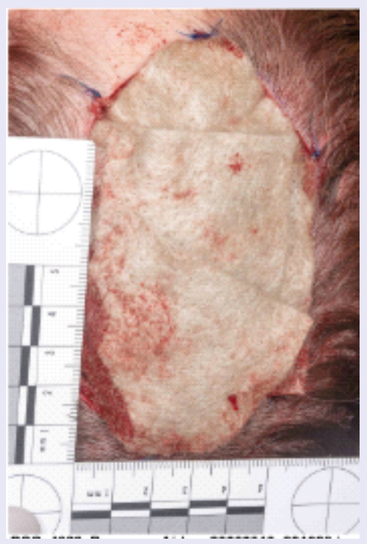
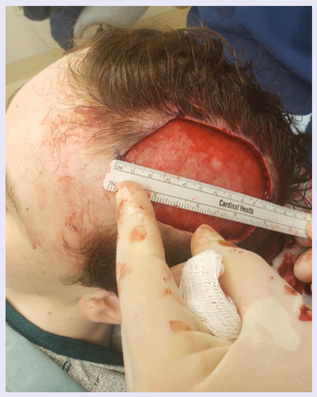
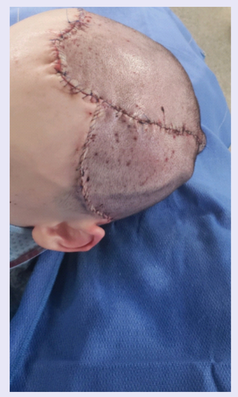


**Figure 7:** A four-stage excision was performed via four sMMS layers until clear margins were pathologically confirmed, which resulted in a temporoparietal defect of 8.5cm x 11.5cm down to and including the galea. The final defect after obtaining clear margins is pictured above with a dressing covering the remaining galea and pericranium.





**Figures 8 and 9:** Pre-reconstruction defect (left). Reconstruction (right) by a team of a plastic and a dermatologic surgeons was completed 2 days after the final Mohs procedure, utilizing a double rotational-advancement scalp flap to close the sizable defect. The patient tolerated all procedures well with an uncomplicated postoperative course.

## Conclusion

DFSP is a rare cutaneous malignancy that is even more uncommon in the scalp and the pediatric population, potentially leading to a missed diagnosis in our patient. Though the current standard of care for these lesions is WLE, MMS has been shown to have lower rates of recurrence and decreased margin width. Although our patient underwent MMS, he was left with a large scalp defect requiring complex advancement flap reconstruction. There is a preponderance of evidence favoring MMS for DFSP; however, further clinical trials should be performed in this area to strengthen the case for MMS.

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