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Scalp Dermatofibrosarcoma Protuberans: A Rare Localization Mimicking an Arteriovenous Malformation – A Case Report

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Abstract: Original Research

Background: Dermatofibrosarcoma protuberans (DFSP) is a rare, low-grade dermal sarcoma with a high risk of local recurrence. Scalp localization is extremely uncommon and may lead to diagnostic confusion. We report an unusual case of scalp DFSP in a young adult, initially misdiagnosed as an arteriovenous malformation (AVM), underlining diagnostic pitfalls and the importance of appropriate surgical management. ***Case Presentation***: A 20-year-old man presented with a painless, progressively enlarging occipital scalp mass evolving over 18 months. Initial clinical and radiological findings were consistent with a vascular lesion. The patient underwent six sessions of sclerotherapy for presumed AVM without improvement. Surgical excision revealed a fusocellular mesenchymal proliferation on histopathology, consistent with DFSP. Immunohistochemical staining showed diffuse CD34 positivity and was negative for PS100, AML, Desmin, and STAT6. Due to close surgical margins (<0.1 cm), a re-excision was performed with wide local margins and reconstruction by full-thickness skin graft. No residual tumor was detected on histological review. Follow-up at 3 and 10 months showed satisfactory healing and no signs of recurrence. ***Conclusion:*** This case highlights a rare presentation of DFSP mimicking a benign vascular anomaly. Accurate diagnosis through histopathology and immunohistochemistry is essential. Wide local excision remains the gold standard for treatment to minimize recurrence risk.

Keywords: Dermatofibrosarcoma protuberans, Scalp tumor, Arteriovenous malformation, CD34, Surgical margins, Case report.

BACKGROUND

Dermatofibrosarcoma protuberans (DFSP), also known as Darier-Ferrand tumor, is a rare dermal-origin sarcoma characterized by aggressive local infiltration and a high recurrence rate, despite its low metastatic potential. Its incidence is estimated at 4.5 cases per million per year. DFSP commonly occurs on the trunk and extremities; involvement of the scalp is exceptional. Due to its nodular appearance, DFSP may mimic benign conditions such as arteriovenous malformations (AVMs), as illustrated in our case.

CASE PRESENTATION

Clinical Presentation

A 20-year-old male with no significant medical history presented with a painless subcutaneous occipital mass that had gradually increased in size over 18 months. Clinical examination showed a firm, fixed mass without signs of inflammation or vascular bruit. The initial diagnosis was AVM, and the patient underwent six sessions of sclerotherapy with no improvement.

Preoperative Imaging

- Doppler Ultrasound: A subcutaneous, roughly oval, hypoechoic, heterogeneous, vascularized soft-tissue mass measuring 33×19 mm. Further sectional imaging was recommended.
- Contrast-enhanced Brain CT: A left parietal subcutaneous scalp lesion measuring 47.6×21.8×56 mm, isodense with homogeneous enhancement after contrast injection, abutting the external table of the parietal bone without bone lysis or erosion.
- Second CT scan: A large left occipito-parietal scalp mass measuring 9×4 cm, with intense post-contrast enhancement.

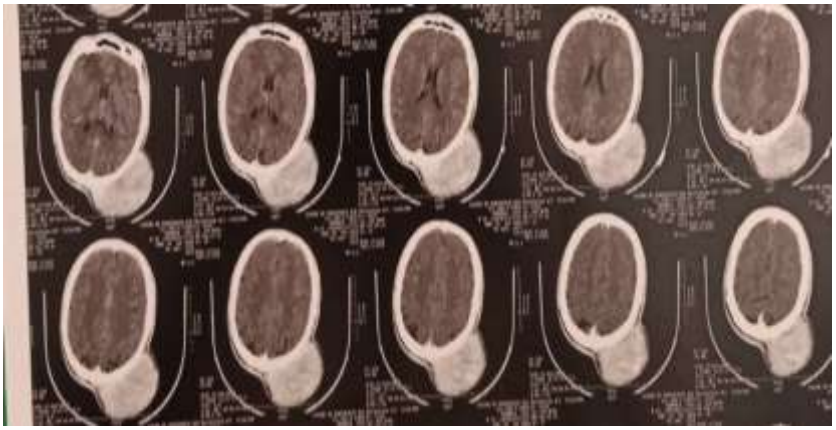


Figure 1: CT image showing the subcutaneous occipital lesion

Figure 2: Preoperative clinical image showing the occipital mass

First Surgery (January 9, 2024)

A complete excision of the mass was performed under general anesthesia. The wound was closed primarily to enable full histological analysis.

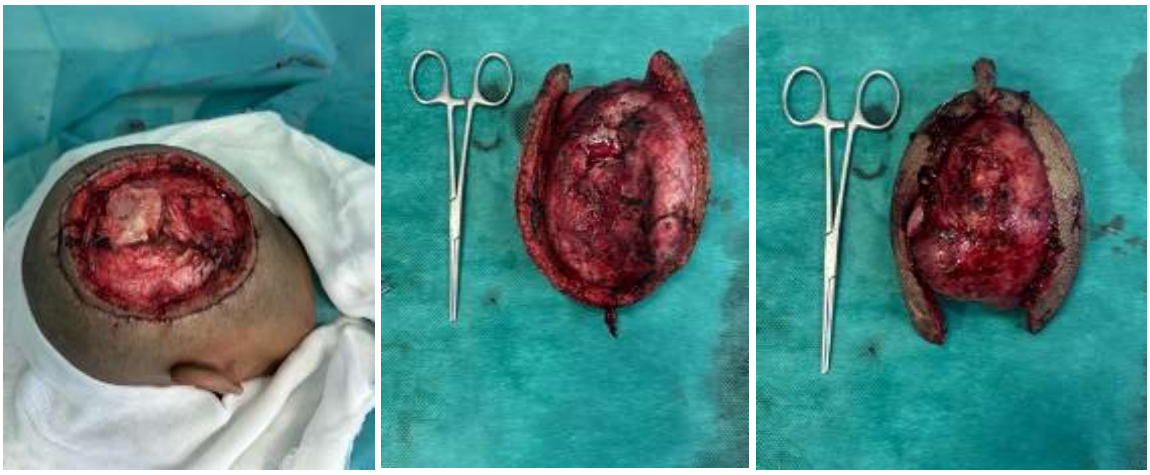


Figure 3: Intraoperative view after initial excision

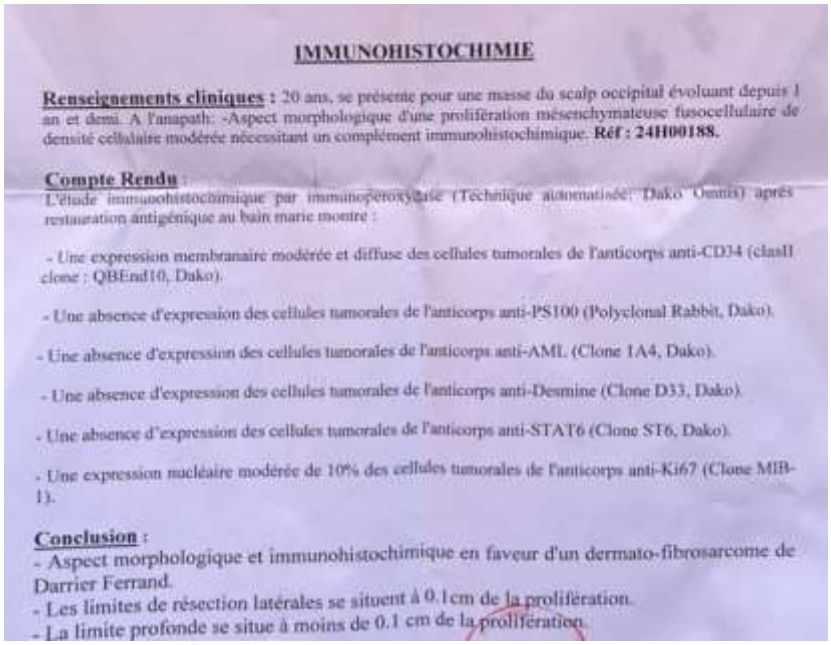
Histopathology

Histological analysis revealed a moderately dense fusocellular mesenchymal proliferation infiltrating the subcutaneous tissue. The deep and lateral margins were less than 0.1 cm from the tumor.

Immunohistochemistry

- CD34: Moderate, diffuse membranous expression
- PS100, AML, Desmin, STAT6: Negative
- Ki67 index: Less than 10%

These findings confirmed the diagnosis of DFSP.



IMMUNOHISTOCHIMIE

Renseignements cliniques : 20 ans, se présente pour une masse du scalp occipital évoluant depuis 1 an et demi. A l'anapath: -Aspect morphologique d'une prolifération mésenchymateuse fusocellulaire de densité cellulaire modérée nécessitant un complément immunohistochimique. **Réf : 24H00188.**

Compte Rendu :
L'étude immunohistochimique par immunopéroxydase (Technique automatisée: Dako Omnis) après restauration antigénique au bain marie montre :

- Une expression membranaire modérée et diffuse des cellules tumorales de l'anticorps anti-CD34 (classII clone : QBEnd10, Dako).
- Une absence d'expression des cellules tumorales de l'anticorps anti-PS100 (Polyclonal Rabbit, Dako).
- Une absence d'expression des cellules tumorales de l'anticorps anti-AML (Clone 1A4, Dako).
- Une absence d'expression des cellules tumorales de l'anticorps anti-Desmine (Clone D33, Dako).
- Une absence d'expression des cellules tumorales de l'anticorps anti-STAT6 (Clone ST6, Dako).
- Une expression nucléaire modérée de 10% des cellules tumorales de l'anticorps anti-Ki67 (Clone MIB-1).

Conclusion :
- Aspect morphologique et immunohistochimique en faveur d'un dermato-fibrosarcome de Darrier Ferrand.
- Les limites de résection latérales se situent à 0.1cm de la prolifération.
- La limite profonde se situe à moins de 0.1 cm de la prolifération.

Figure 4: Positive CD34 immunostaining

Second Surgery (April 17, 2024)

Due to the narrow surgical margins from the first excision, a wide re-excision of the scar and adjacent tissues down to the external table of the skull was performed. A full-thickness skin graft was used for reconstruction.

Figure 5: Intraoperative view during re-excision

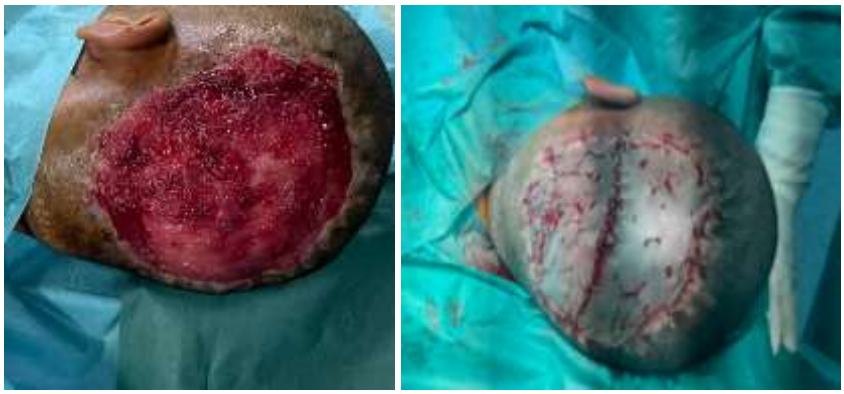


Figure 6: Placement of full-thickness skin graft

Histological Findings After Re-excision

Histopathological examination revealed a fleshy granulation tissue without residual malignancy. Chronic inflammatory changes and bone fragments were noted.

Postoperative Outcome

The skin graft healed without complication. Clinical follow-up at 1, 3, and 6 months showed good healing with no local recurrence.

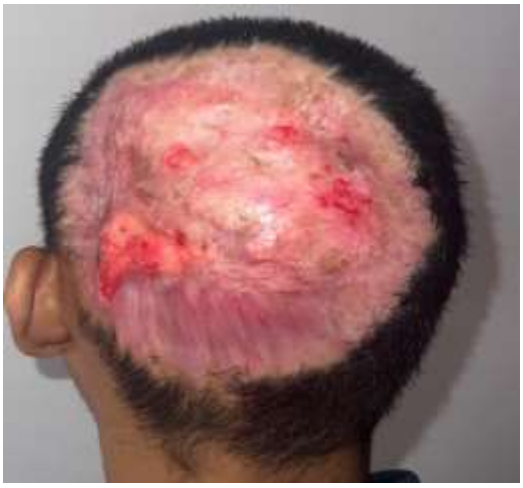


Figure 7: Three-month postoperative result

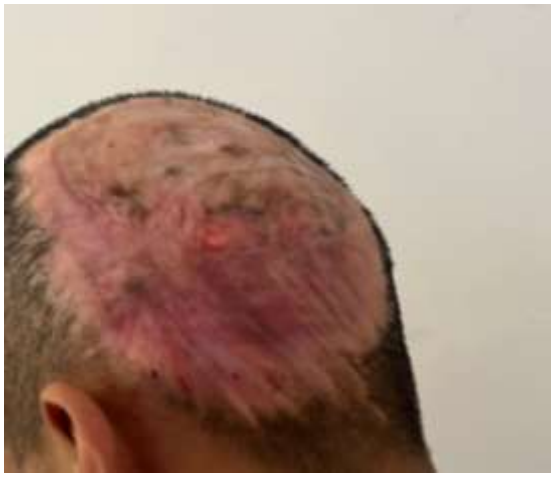


Figure 8: Ten-month postoperative result

DISCUSSION

DFSP is a rare neoplasm that can be diagnostically challenging, particularly in atypical locations such as the scalp. In our case, clinical and imaging features mimicked an AVM, leading to delayed diagnosis and ineffective treatment. This diagnostic confusion is reported in recent literature (e.g., Smith *et al*., 2018; Lee *et al*., 2020), emphasizing the importance of systematic preoperative biopsy in any atypical scalp mass.

Histopathology is essential for diagnosing DFSP, which is characterized by an infiltrative spindle cell proliferation. Diffuse CD34 positivity is a hallmark immunohistochemical finding (Garcia *et al*., 2019). A low Ki67 index supports the low-grade nature, although close margins are associated with high recurrence rates.

The mainstay of treatment is wide surgical excision with 2–3 cm margins, when anatomically feasible. In the scalp, achieving adequate margins may be technically difficult and require re-excision with complex reconstruction using grafts or flaps (Miller *et al*., 2017). In our case, a second surgery ensured clear margins and good outcomes.

Complete surgical excision remains the treatment of choice, with long-term follow-up essential due to recurrence rates reaching up to 50% in some series (Jones *et al*., 2016). Adjuvant therapies like radiotherapy or imatinib may be considered in recurrent or unresectable cases, although their role in initial management is limited (Brown *et al*., 2018).

CONCLUSION

This case highlights the need to consider DFSP in the differential diagnosis of any chronic subcutaneous scalp mass, particularly when vascular anomalies are suspected. Preoperative biopsy is crucial for accurate diagnosis. Optimal management requires wide excision with sufficient margins and close postoperative surveillance to prevent recurrence.

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