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Case Report

Giant scalp dermatofibrosarcoma protuberans at mid-occipital scalp: A rare case report

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ABSTRACT

Background: Dermatofibrosarcoma protuberans (DFSP) is a rare type of skin cancer that arises in the deeper layers of the skin, most commonly on the trunk and limbs. The presentation of DFSP on the scalp is worth documentation due to its extremely rare occurrence in the literature.

Case Description: We describe a case of a 41-year-old female presented with a lump on the back of the head with gradual enlargement. It was solitary with a size 19 × 12 × 10 cm with purulent discharge and intermittent pain. A computed tomography-scan showed an isodense mass at mid occipital without intracranial involvement. Tumor extirpation and the free flap were performed collaborated with plastic surgery. The patient was discharged unremarkably on the 7th postoperative day without any neurological deficits.

Conclusion: Our case is distinct because, after 24 months without any adjuvant therapy and without any recurrence, the patient is still doing well. This is a very rare clinical entity.

Keywords: Dermatofibrosarcoma, Head neoplasm, Resection margin

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare type of skin cancer (1–2% of all soft-tissue sarcoma) that arises in the deeper layers of the skin, most commonly on the trunk and limbs.[14] On the other hand, it can also happen on the scalp, which poses special difficulties for diagnosis and treatment. It makes up around 6% of soft-tissue sarcomas and has an incidence rate of 0.8–4.5 cases/million persons per year.[8] While some DFSPs include a high-grade sarcomatous component, most DFSPs are low-grade.[17] It typically affects the dermis, or middle layer of skin, along with subcutaneous fat and, in rare cases, muscle and fascia.[3,21] DFSP is characterized by a considerable tendency of local recurrence following surgical resection, notwithstanding the slow likelihood of metastasis.[10] Patients with DFSP often experience recurrence, with a reported prevalence of 20–50%. There appears to be a poor association between tumor size and recurrence rate, although completeness of excision and distance of tissue excision margins from the tumor has been shown to influence recurrence rate. Surgical excision with a free margin of at least 2 cm, known as a wide local excision, has been shown to considerably lower relapse rates, according to multiple studies. The majority of DFSP variations do not show significant differences in clinical

behavior.[11,15,16,18,19] We are presenting this case report because the presentation of DFSP in the scalp is rare, not to mention the prevalence of DFSP *per se*.

CASE PRESENTATION

We described a case of a 41-year-old female who initially presented with a lump on the back of her head that had gotten bigger overtime since a year ago, which was initially a diameter of 1 cm and now became over 10 cm. She had not visited any physicians during the meantime until the lump obstructed her daily activities. The complaint was accompanied by wound and pus on the lump and intermittent pain in the lump since 5 months prior. On physical examination, vital signs and other local examinations were within normal limits. As with the mass, there was an irregularity of skin color and lobulation. The mass was solitary, 19 × 12 × 10 cm diameter, with a fixated base followed by discharge and a smelly odor [Figures 1a-e]. Other than DFSP, we came up with a differential diagnosis of scalp hemangioma and basal cell carcinoma.

Head contrast computed tomography-scan showed mix density mass at mid occipital extracalvaria without intracranial involvement. 3D angiography imaging revealed that the tumor received blood supply from both the superficial temporal arteries and bilateral occipital arteries [Figures 2a,b]. Due to its high vascularity, the possibility of a hemangioma still could not be ruled out. We performed a joint meeting with plastic surgery, radiology, and anatomical pathology of our hospital before proceeding with the surgery.

Under general anesthesia, the patient was positioned prone with the head elevated at approximately 20°. A wide radical excision with a 4 cm distance from the neoplasm margin and translational skin lines were drawn. After a wide radical excision was performed, the skin defect was then reconstructed by translational skin flap and split-thickness skin graft. An irregular, skin colored, multiple lobulated mass measuring 19 × 12 × 10 cm in diameter was excised along the surrounding skin in a radius of 4 cm from the neoplasm edge [Figures 3a-g]. Since there were no immediate complications, the patient was admitted after the operation to intensive care for 4 days and then moved to the general ward. The patient was discharged remarkably on the 7th postoperative day without any neurological deficits. We did not plan for adjuvant therapy because the mass was completely resected with a wide margin.

As in histopathological findings, using hematoxylin and eosin stained sections showed a densely cellular and poorly circumscribed tumor in the dermis layer, comprising interwoven bundles and fascicles of uniform spindle shaped cells arranged in a "storiform" or "cartwheel" pattern. The tumor cells had a monotonous appearance with oval nuclei, vesicular chromatin, inconspicuous nucleoli, and scanty-to-moderate cytoplasm [Figures 4a-c]. The patient was followed up on 3 months, 1 year, and 2 years after the visit; where no recurrence has been noted.

Figure 1: (a-e) Swelling in mid occipital region in a 41-year-old female patient, with the mass size of 19 × 12 × 10 cm.

DISCUSSION

DFSP is a rare soft-tissue neoplasm first identified by Sherwell and Taylor in 1890.[20] In 1924, Darier and Ferrand defined it as a recurrent, progressive dermatofibroma. Hoffmann gave it the name DFSP a year later; it had previously been described under several different headings, such as skin fibrosarcoma, hypertrophic morphea, progressive and recurring dermatofibroma, and keloid-like sarcomatous tumors.[12,13] It is believed to have started from a dermal stem cell or an undifferentiated mesenchymal cell that was fibroblastic, muscular, or neurologic.[5]

Standard treatment of DFSP is wide local excision with margins of at least 2 cm and many now advocate Moh's micrographic surgical technique as achieving better results.[21,22] Even though total resection has a great prognosis if sufficient surgical margins are not achieved, there is a noticeable tendency for the tumor to return. Due to the tumor's demonstrated radiosensitivity, radiation therapy is often a part of the treatment plan. Chemotherapy using selective tyrosine kinase inhibitors, like imatinib mesylate, has also demonstrated promising outcomes.[15]

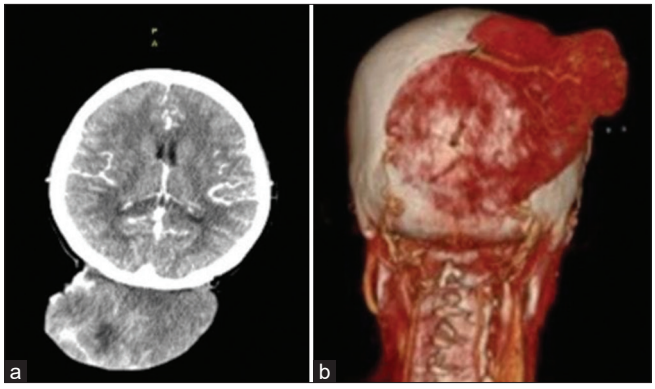

Figure 2: Computed tomography-scan showed (a) Mix density mass (b) 3D Imaging showed received blood supply from both the superficial temporal arteries.

In terms of genetics, DFSP is frequently linked to chromosome 11's supernumerary ring, which frequently contains amplified sequences from chromosomes 17 and 22. The final gene product is the consequence of the fusion of the collagen type 1 alpha 1 gene (COL1A1) and platelet-derived growth factors'-beta chains (PDGF-β), with the PDGF-β being expressed under the COL1A1 promoter. It has been suggested that PDGF-β overproduction contributes to the development of dermatofibrosarcoma.[16] Despite being historically linked to a fibroblastic origin, new immunohistochemistry data indicate that the tumor might originate from the skin's dendritic cells. The DFSP was initially characterized as a "progressive and recurring dermatofibroma" by Darier and Ferrand in 1924, highlighting the condition's propensity for local recurrence.[7,22] In 1925, Hoffman introduced the term DFSP and described three new instances.[6,17] This tumor is rare; it typically manifests between the ages of 20 and 50, and it seldom affects children under the age of 16. It is uncommon on the head and neck and most frequently affects the trunk and proximal limbs. In children, the limbs and trunk are likewise the most frequently seen areas.[2,22]

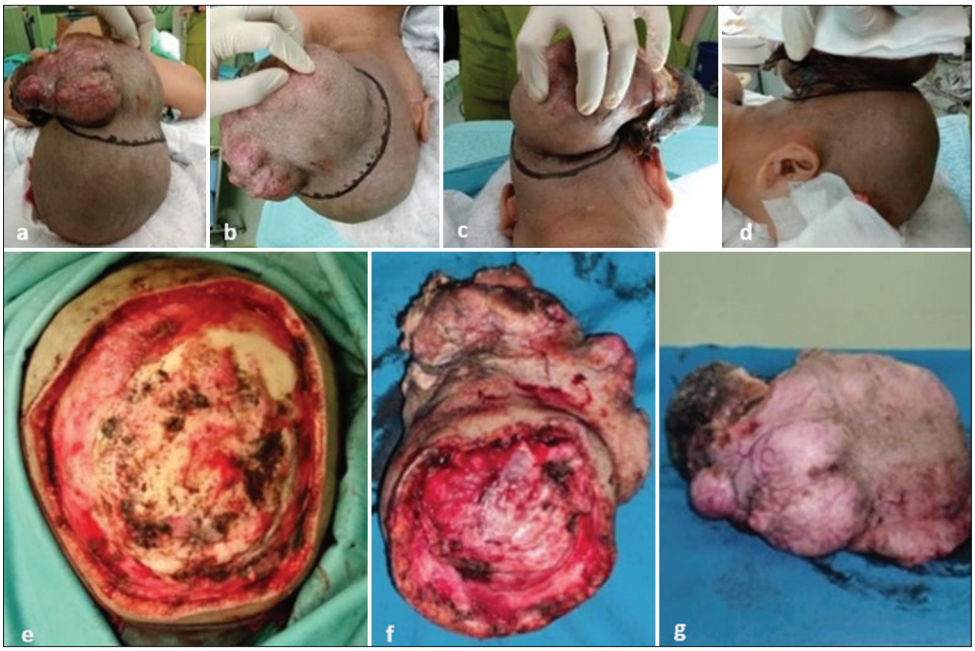

Figure 3: (a-d) A wide radical excision with 4 cm distance from neoplasm margin (e) tumor is removed with periosteum (f-g) An irregular, skin colored, multiple lobulated mass, measuring 19 × 12.

Table 1: Summary of case reports of scalp DFSP in literature, adapted from Kuhlmann *et al.*

No.	Sex and age	Main location	Tumor volume cm^3	Treatment	Outcome
1	F 33	Temporooccipital	31.9	2× resection	None
2	F 47	Frontal	9	3× resection	None
3	F 30	Parietal	16	2× resection	None
4	M 33	Occipital	72	1× resection	None
5	F 63	Frontal	Not specified	1× resection	None
6	M 26	Frontal	Not specified	3× resection	None
7	M 57	Parietooccipital	7.3	1× resection	Prostate metastasis
8	M 19	Occipital	9.9	1× resection	None
9	F 68	Occipital	Not specified	2× resection	None
10	M 41	Frontal	9.7	2× resection	Not specified
11	M 40	Occipital	Not specified	1× resection	None

DFSP: Dermatofibrosarcoma protuberans

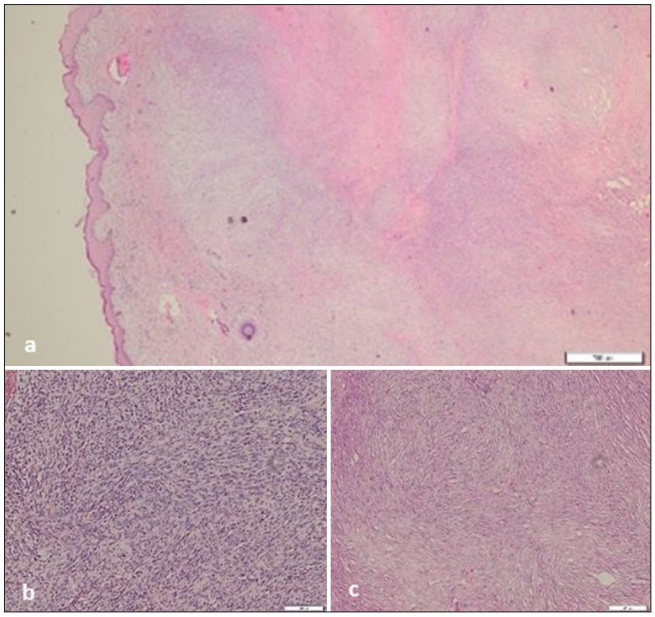

Figure 4: (a) Section showing spindle cells arranged in short fascicles and storiform pattern (Haematoxylin and Eosin, ×20). (b-c) Tumor cells with oval nuclei, vesicular chromatin, and inconspicuous.

In their earlier publications, Faried and Arifin[1,4] described three cases, two of which had significant recurrences before the other's free recurrence after 14 months. Due to this, the best course of action for this patient's condition requires careful handling of this situation to avoid a recurrence.[3] There appears to be a poor association between tumor size and recurrence rate, although completeness of excision and distance of tissue excision margins from the tumor has been shown to influence recurrence rate. Surgical excision with a free margin of at least 2 cm, known as a wide local excision, has been shown to considerably lower relapse rates, according to multiple studies. The majority of DFSP variations do not show significant differences in clinical behavior.[11,15,16,18,19]

Kuhlmann *et al.*, [Table 1] compiled the case reports of DFSP of the scalp and summarized them.[9] They concluded that while local recurrences had not been observed, a meticulous margin assessment should be performed before and during the resection to maximize safety while still preserving healthy tissue.

CONCLUSION

It is uncommon and challenging to detect a huge scalp DFSP, especially when it manifests at the head. Due to misdiagnosis, treatment for massive scalp DFSP is frequently delayed and results in local excision. Therefore, to lower the chance of recurrence, we should be aware of this unusual entity and always perform a wide excision for these tumors. Achieving local control and preventing cosmetic and functional deficits are essential for effective therapy and care; full excision with negative margins should be the goal. Our case is distinct because, after 24 months without any therapy and without any tumor recurrence, the patient is still doing well, and the condition is diagnosed as DFSP of the scalp, a very rare clinical entity.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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