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Case Report

A rare giant scalp dermatofibrosarcoma protuberans

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Abstract

Background: Giant dermatofibrosarcoma protuberans (DFSP) of the scalp is a rare case, which is an intermediate grade soft tissue neoplasm originating from the dermal layer of the skin, which usually occurs in adults.

Case Description: We describe such a case in a 26-year-old male. A wide local excision of the tumor with a generous tissue margin was performed; microscopic and immunohistochemical findings established the diagnosis of recurrent DFSP.

Conclusion: Our case is unique in that it is presented as a dermatofibrosarcoma protuberans of the scalp, which is an extremely rare clinical entity, and the patient remains well after 14 months with no further treatment, without any tumor recurrence.

Key Words: Giant dermatofibrosarcoma protuberans, recurrence, scalp

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is an intermediate malignancy neoplasm, most often diagnosed in individuals aged between 20 and 50 years, and is seen mainly on the trunk and the proximal extremities. This tumor is characterized by its aggressive local growth and marked propensity to recur after surgical excision. The classic histological features comprise of a monotonous storiform growth pattern of cytologically uniform tumor cells, with hyperchromatic and elongated nuclei and a characteristic honeycomb pattern of infiltration into the subcutaneous fat. Immunohistochemically, DFSP is characterized by a positive reaction for vimentin and CD34. This article reviews the incidence, clinical presentation, histologic features, immunocytochemical studies, prognosis, management, and follow-up recommendations for this unusual neoplasm.

DFSP is characterized clinically by its locally aggressive growth and a high rate of local recurrence, but distant metastases (1-4%) and tumor-related deaths are very rare. Recurrence is common in patients with DFSP and the literature suggests an incidence of 20-50%. There seems to be a poor correlation between the size of the tumor and the recurrence rate, but the completeness of excision and the distance of tissue excision margins from the tumor have been reported to affect recurrence rate. Several reports suggest that surgical excision with at least 2 cm of free margin (a so-called wide local excision) reduces the relapse rate significantly. Most of the variants of DFSP are not associated with significant differences in clinical behavior.[6-10]

CASE REPORT

We present a case of a 26-year-old male, who complained of headache with a raised, painless lesion on his left

frontal region [Figure 1a-d] since 2.5 years before admission. On physical examination, there was a single, skin colored, firm nodule, measuring 7 cm in diameter, with a fixated base. An excision biopsy was suggested to diagnose the tumor and to predict tumor behavior. A 2-cm margin of normal appearing scalp is incised down to the underlying periosteum, wide range of the incision was taken since the propensity of tumor spread along their facial planes; followed by undermining the defect with blunt-tipped scissors after removal of the specimen to minimize tension on the wound margin.[3,6] Subsequently, an excision biopsy was performed, and final diagnosis of DFSP was made based on the histopathological findings. Hematoxylin and eosin stained sections showed a densely cellular and poorly circumscribed tumor in the dermis layer, comprising of interwoven bundles and fascicles of uniform spindle-shaped cells arranged in "storiform" or "cartwheel" pattern [Figure 2a and b]. The tumor cells had monotonous appearance with oval nuclei, vesicular chromatin, inconspicuous nucleoli, and scanty to moderate cytoplasm [Figure 2b]. A panel of immunohistochemistry (IHC) comprising vimentin and CD-34 was applied. Tumor cells were positive for both vimentin and CD-34, respectively [Figure 2c and d].

He was discharged from our hospital uneventfully, and no recurrence of the mass was detected at 1-year follow-up.

DISCUSSION

DFSP is a slow growing, locally aggressive tumor of intermediate malignancy with a marked tendency for local recurrence, but rarely metastasizes. Genetically, DFSP is commonly associated with supernumerary ring of chromosome 11, often with amplified sequences from chromosomes 17 and 22. The resulting gene product involves the fusion of platelet-derived growth factors' beta chains (PDGF-β) with collagen type 1 alpha 1 gene (COL1A1), placing the PDGF-β under the COL1A1 promoter. Overproduction of PDGF-β has been reported to play a role in the development of dermatofibrosarcoma.[7] Although historically attributed to a fibroblastic origin, recent IHC evidence suggests that this tumor may arise from the dendritic cell in the skin. In 1924, Darier and Ferrand first described the entity of DFSP as a "progressive and recurring dermatofibroma", underscoring its predilection for local recurrence.[2] Hoffman reported three new cases and proposed the term DFSP in 1925.[5] This tumor is uncommon, usually present between 20 and 50 years of age, and is rare in children aged less than 16 years. It is most often found on the trunk and proximal limbs and is rare on the head and neck. The trunk and limbs are also most common sites in children.[1,13] It usually presents as a violet or bluish erythematous plaque or atrophic plaque or macule that is vascular in appearance. Tumor size varies from a few millimeters to a few centimeters. Clinically, it is also confused with other lesions such as sclerosing basal cell carcinoma, morphea, scar, and anetoderma. Clinical diagnosis of DFSP in infancy and childhood may be difficult because, in the early stages, the tumor often looks like a vascular malformation.[11]

The immunohistochemical demonstration of CD34 is an important feature for diagnosing DFSP.[4,12] In a case of conventional DFSP, areas of strong CD34 positivity were noted. The recurrence potential of DFSP is directly related to the extent of resection, although it can be very difficult to determine a positive margin because DFSP has fibroblast-like morphology. The risk of metastasis is greatly reduced if there is no recurrence, because metastasis never occurs without a preceding local recurrence. However, tumors that are removed by wide local excision with clear

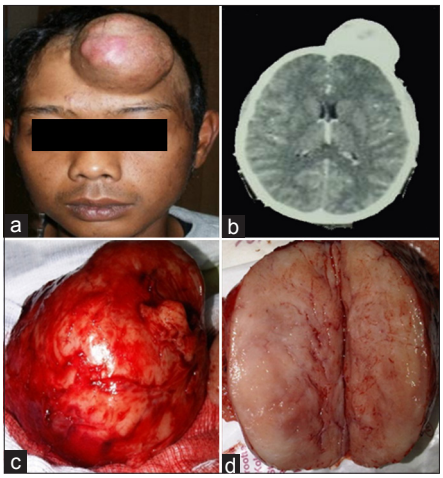

Figure 1: (a) Swelling in left frontal region in a 26-year-old male patient. (b) Contrast head CT scan. (c) Intraoperative finding showing soft tissue mass lesion in left frontal region. (d) Mass size 7 × 6 × 5 cm

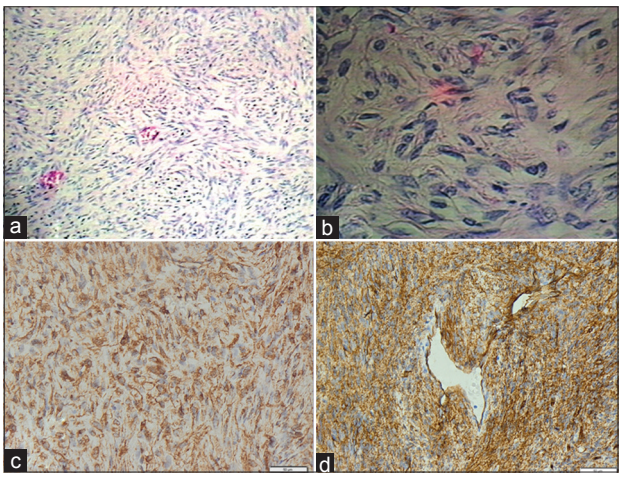

Figure 2: (a) Section showing spindle cells arranged in short fascicles and storiform pattern (Hematoxylin and eosin, x10). (b) Tumor cells with oval nuclei, vesicular chromatin, inconspicuous nucleoli, and scanty to moderate cytoplasm (Hematoxylin and eosin, x40). (c) Section showing tumor cells focally positive staining for Vimentin (x200). (d) Section showing tumor cells focally positive staining for CD34 (x200)

margins probably represent a less aggressive subset of lesions, as shown in high grade extremity sarcomas.[9]

CONCLUSION

A giant scalp DFSP is rare and difficult to diagnose, particularly when presenting at the head. Treatment of giant scalp DFSP is often delayed because of misdiagnosis, leading to local excision. Hence, we should be aware of this uncommon entity, and always perform a wide excision for these tumors to reduce the risk of recurrence.

Consent

Informed consent was obtained from the patient for publication of this case report and any accompanying images. His family was present at the time.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

MZA, FY, RHD, ABS, and AF had examined, treated, observed, and followed up the subject of this research. MZA, FY, and RHD performed the operation on the patient. BSH carried out the IHC studies and interpreted the results of the patient's biopsy sample. All authors participated in writing the manuscript. All authors has read and approved of the final manuscript.

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