CASE REPORT

Dermatofibrosarcoma Protuberans of Scalp - An Unusual Site of Occurrence

Jyotsna,V Shruthi Kamal, Shivarahini, Ruban Kumar

Abstract

Dermatofibrosarcoma protuberans [DFSP] is a low to intermediate soft tissue sarcoma arising from the skin dermis. DFSP of the scalp is very rare and is often confused with an epidermoid cyst. Patient Concern: Women reported with a swelling on the scalp for 6 months. Rational: The purpose of the report is to highlight the variable presentations of DFSP. Report: We report a case of DFSP of the scalp in a middle-aged woman. Diagnosis: The lesion histologically showed spindle-shaped cells and was positive for S100 and Ki 67 and negative for CD34. Outcome: The lesion was excised and the margins were negative for tumor cells. The patient was followed up for 2 months and there were no recurrences reported. Lessons: One must be aware of this uncommon entity and always perform a wide excision for these tumors to reduce the risk of recurrence

Keywords

Dermatofibrosarcoma Protuberans, Darier-Hoffmann Tumor, DFSP, Fibrosarcomatous

Introduction

In 1924 Darier and Ferrand were the first to describe the Dermatofibrosarcoma protuberans [DFSP], but it was Hoffmanin who established the definition of "DFSP" in 1925. [1] The estimated incidence is 0.8 to 4.5 cases per million per year. [2] It is a rare, low to intermediate-grade soft tissue sarcoma arising from the skin dermis. Initially, it presents as a slow-growing, painless, skin-colored plaque with discolorations. [3] Later, it becomes protuberant. It accounts for almost 1% of all soft tissue sarcomas and <?0.1% of all cancers. [2] Affects mostly the adults of 20-50 years. Equal gender predilection.[4] 42-72 % DFSP occur in trunk, 20-30 % proximal extremities and 10-16 % in head and neck.[5] Of all DFSP, DFSP of the scalp accounts for less than 5% . [4] Though it is a low-grade malignancy it is locally invasive, leading to bony erosion, and invasion into the brain, especially with recurrent tumors. The local recurrence rates for all cases of DFSP of the head and neck are extremely high, ranging from 50 to 75%. Studies reported 5-15% fibrosarcomatous changes [FS-DFSP] with a high risk of recurrence and metastasis. [5, 4]

Case Report

A 39-year-old woman came to the surgical outpatient clinic with complaints of swelling in the scalp for the past 6 months. She did not have any other co-morbidity. The swelling had a gradual onset which progressed to the

Department of General Surgery, Saveetha Medical College And Hospital Thandalam Chennai - 602105

Correspondence to: Dr Jyotsna, Post graduate Department of General surgery Saveetha Medical College And Hospital Thandalam Chennai - 602105

Manuscript Received: 19.08.2022; **Revision Accepted**: 01.11.2022;

Published Online First: 10 July, 2023

Open Access at: https://journal.jkscience.org

Cite this article as: Jyotsna, Shruthi Kamal V, Shivarahini, Kumar R . Dermatofibrosarcoma Protuberans of Scalp - An Unusual Site of Occurrence JK Science 2023;25(3):194-96.

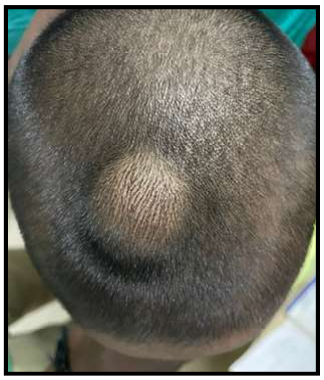

Fig 1. Swelling on the right parietal region of the scalp

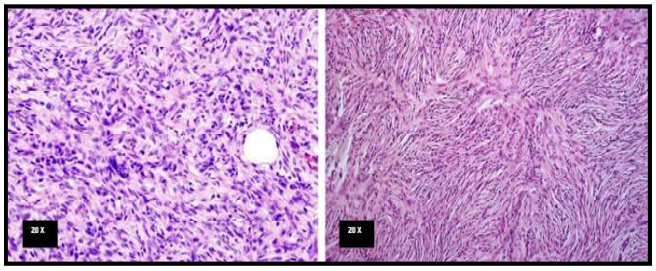

Fig 3. Histopathology showing: A. Fibroblast showing spindle shaped cells and B. Focal areas of spindle cells arranged in fascicles

current size. It was not associated with trauma, pain, or discharge. There was no other swelling elsewhere in the body. On physical examination, there was a spherical-shaped swelling, about 2 cm x 2 cm located in the right parietal region of the scalp [*Fig 1*]. It was non-tender, firm in consistency, non- Trans illuminant, and non-pulsatile. Impulse on cough was absent. There were no palpable lymph nodes noted. The skin over the swelling was normal. Her blood investigations were within the normal range. Serology for Human ImmunoVirus HIV, Hepatitis B surface antigen [HbsAg], and Hepatitis C Virus [HCV] was non-reactive. Radiographic examination of the skull [Anteroposterior and Lateral view] showed no intracranial extension [*Fig 2*]. With the clinical and radiographic findings, a diagnosis of a dermoid cyst on the scalp was given. The swelling was excised under local anesthesia and sent for histopathological examination. The hematoxylin and eosin stained sections showed features suggestive of spindle cell neoplasm, probably dermatofibrosarcoma protuberans [*Fig 3*]. In

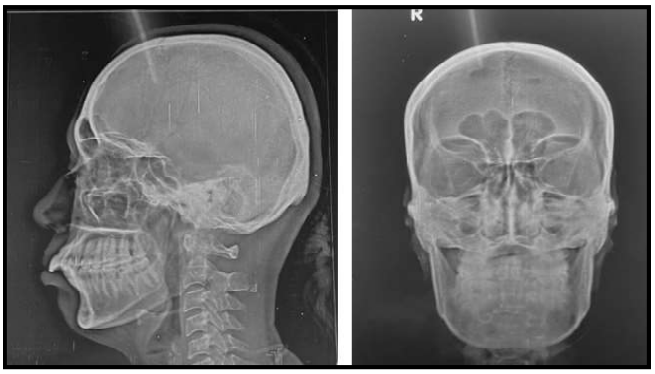

Fig 2: Radiograph of skull: A .Lateral View and B. Antero-posterior view

Immunohistochemistry [IHC], the sections showed positivity for S100, Ki67, and negative for CD34, and a final confirmatory diagnosis of low-grade spindle cell sarcoma was made. After obtaining informed consent, a wide local excision with rotational flap under general anesthesia was performed. Since the lesion was diagnosed as DFSP, the margin status ought to be confirmed. Hence, a re-excision of margins was performed and they were reported to be negative for malignancy. The patient was followed up for 2 months without any recurrences.

Discussion

DFSP of the scalp is rare and aggressive. Here we presented a case of DFSP of the scalp. DFSP presents as a violaceous, pink, or reddish-brown plaque with slow growth, initially confined to the skin. At later stages, the tumor develops multiple "protuberant" nodules infiltrating the surrounding subcutaneous tissue, fascia, muscles, and bone. [5] A nonprotuberant tumor takes 7.6 [± 9.3] years to become the protuberant variants. [7] Since the present case was a 6-month-old, suggestive of a benign lesion presented as a well-localized, with the absence of secondary changes and without any infiltration into the surrounding tissues, a clinical diagnosis of the dermoid cyst was considered with the differential diagnosis of keloid, lipoma, dermatofibroma, and nodular fasciitis. [8] Imaging of DFSP is usually done to know the extent of the lesion and to identify any distant metastasis. The radiography of the present case there was no cranial

involvement nor was there any metastasis noted. Since dermoid cyst was the clinical diagnosis made, the need for MRI was not anticipated. Moreover, the literature says that MRIs cannot be relied on in such cases as they may not always distinguish DFSPs from other soft tissue sarcomas. [9] Therefore, histopathology remains the gold standard for the diagnosis of DFSPs. Therefore, complete wide excision was performed. The H&E stained sections revealed a spindle cell neoplasm with a probability of dermatofibrosarcoma protuberans. Hence, the tissues were studied with S100, Ki67, and CD 34 markers. It was positive for S100 and Ki67 thus confirming the fibrous nature with its high proliferative capacity. And CD 34 negativity suggests that it was an aggressive fibrosarcomatous variant of DFSP. Generally, the histopathology of DFSP shows a distinct "storiform" or "cartwheel" arrangement of uniform appearing fibroblasts. The four main types of DFSP are ordinary, fibrosarcoma, pigmented, and myxoid. [6] The fibrosarcomatous variant [DFSP-FS] in IHC shows areas of fibrosarcoma are characterized by increased cellularity and loss of CD34 positivity. [11] Mitotic count, necrosis, and areas of fibrosarcomatous change should be correlated with its aggressive clinical behavior and poor survival;[11] Hemorrhagic foci resembling aggressive vascular neoplasms and carry a higher metastatic potential. [4] Previously 21% of DFSP with fibrosarcomatous changes showed metastasis. [11] Chromosomal aberrations are responsible for the transition from DFSP to DFSP-FS. [12]

Simple excision of DFSP is not the treatment of choice owing to its infiltrative behavior. [4] The most accepted treatment of DFSP is wide local excision with 2 cm margins and Mohs micrographic surgical technique. [13] Since the lesion was excised anticipating it to be a dermoid cyst, margins were not excised with the lesion. The lesion was confirmed histopathologically as a DFSP, the margins of the lesion were to be removed later to know the margin status. Margins were negative in the present case. The recurrence rate for wide excision is 20% and for simple excision is 44%. [6] This case was followed up for 2 months and no recurrences were reported.

Conclusion

Scalp DFSP is rare and difficult to diagnose. Treatment of 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.

References

1. Paramythiotis D, Stavrou G, Panagiotou D, Petrakis G, Michalopoulos A. Dermatofibrosarcoma protuberans: a case report and review of the literature. Hippokratia 2016; 20:80-83.
2. Natali S, Borelli C, Shogan P. Dermatofibrosarcoma Protuberans. J Am Osteopath Assoc 2020; 120:362.
3. Eguzo K, Camazine B, Milner D. Giant dermatofibrosarcoma protuberans of the face and scalp: a case report. Int J Dermatol 2014; 53:767-72.
4. Loss L, Zeitouni NC. Management of scalp dermatofibrosarcoma protuberans. Dermatol Surg 2005; 31[11 Pt 1]:1428-33.
5. Stivala A, Lombardo GA, Pompili G, Tarico MS, Fraggetta F, Perrotta RE. Dermatofibrosarcoma protuberans: Our experience of 59 cases. Oncol Lett 2012;4:1047-55
6. Du K, Li J, Tang L, Lin X, Kong X, Liao X, et al. Role of postoperative radiotherapy in dermatofibrosarcoma protuberans: a propensity score-matched analysis. Radiat Oncol 2019; 14:20.
7. Martin L, Piette F, Blanc P, Mortier L, Avril MF, Delaunay MM, et al. French Group for Cutaneous Oncology. Clinical variants of the preprotuberant stage of dermatofibrosarcoma protuberans. Br J Dermatol 2005; 153:932-36.
8. Angouridakis N, Kafas P, Jerjes W, Triaridis S, Upile T, Karkavelas G, et al. Dermatofibrosarcoma protuberans with fibrosarcomatous transformation of the head and neck. Head Neck Oncol 2011;3:5
9. Paramythiotis D, Stavrou G, Panagiotou D, Petrakis G, Michalopoulos A. Dermatofibrosarcoma protuberans: a case report and review of the literature. Hippokratia 2016; 20:80-83.
10. Llombart B, Serra-Guillén C, Monteagudo C, López Guerrero JA, Sanmartín O. Dermatofibrosarcoma protuberans: a comprehensive review and update on diagnosis and management. Semin Diagn Pathol 2013; 30:13-28.
11. Gatlin JL, Hosch R, Khan M. Dermatofibrosarcoma protuberans of the scalp with fibrosarcomatous degeneration and pulmonary metastasis. J Clin Imaging Sci 2011; 1:55.
12. Wang J, Morimitsu Y, Okamoto S, Hisaoka M, Ishida T, Sheng W, Hashimoto H. COL1A1-PDGFB fusion transcripts in fibrosarcomatous areas of six dermatofibrosarcomas protuberans. J Mol Diagn 2000;2:47-52.
13. Love WE, Keiler SA, Tamburro JE, Honda K, Gosain AK, Bordeaux JS. Surgical management of congenital dermatofibrosarcoma protuberans. J Am Acad Dermatol 2009; 61:1014-23