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Original Research Paper

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DERMATOFIBROMA SARCOMA PROTUBERANS OF SCALP – A OVERVIEW AND CASE REPORT!

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ABSTRACT

Background- Dermatofibroma sarcoma protuberans(DFSP) of scalp is a rare, low grade, locally infiltrative sarcoma. It originates from the fibroblasts in the dermal layer of skin.

Case description- We describe such a case in a 39- year old male for whom wide local excision of the lesion with adequate margin was done. Microscopic and immunohistochemical findings established the diagnosis of DFSP.

Conclusion- This case is significant as DFSP presenting on the scalp is very rare and accounts for less than 5% of all DFSP occurrence.

KEYWORDS : Dermatofibroma sarcoma protuberans, wide excision, CD34, imatinib.

Case report-A 39 year old male presented to the OPD complaining of a diffuse discoloration and swelling on his scalp since 4 years. The patient gave a history of two recurrent surgeries been done on the scalp within the past one year which was unsuccessful. The recent excisional biopsy gave a histopatholgical diagnosis of dermatofibrosarcoma protuberans. The CT scan showed no periosteal involvement of the skull and no significant neck nodes were seen. The chest xray and USG abdomen were unremarkable.

Wide excision of the lesion with rotational flap and skin graft coverage was planned for the patient. An anterolateral thigh free flap was kept as standby, to be used in case the coverage deemed insufficient.

The patient was scrubbed and draped according to standard protocol followed by nasotracheal intubation. Careful marking was made with a safe margin of 1.5cm. Complete excision of the lesion was done sparing the periosteum. A superiorly and inferiorly based rotational flap was attempted which successfully closed the defect. A split thickness skin graft was harvested from the thigh and was used to cover the lateral temporal defect created by the rotational flap. A pressure bandage was placed and the operative area was immobilized for 10 days. Post-operative healing was uneventful.

Fig 1 – Preoperative marking

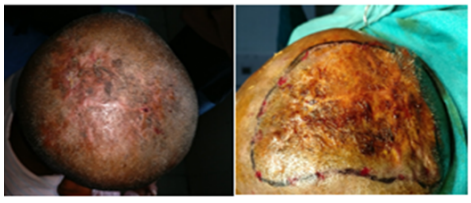


Fig 2 - Post Excision defect

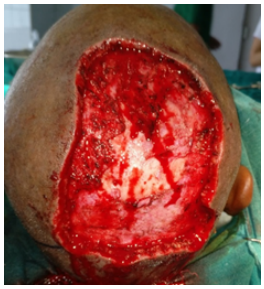


Fig 3 – After Rotationtal advancement and skin graft placement

Fig 4 - Tumor Specimen

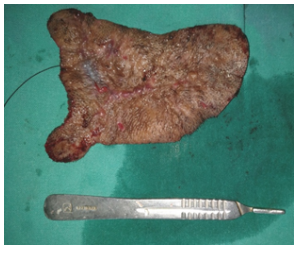
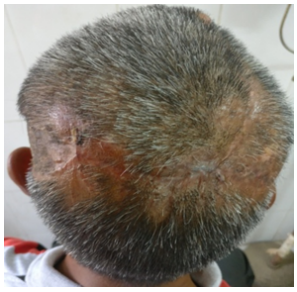


Fig 5 – Post –operative 24 months



The patient has completed 24 months of follow-up and is disease free.

DISCUSSION-

DFSP is a locally aggressive, tumor with a marked propensity for recurrence. In 1924, Darier and Ferrand first described the entity of DFSP as a "progressive and recurring dermatofibroma", underscoring its predilection for local

recurrence (1). Hoffman reported three new cases and proposed the term DFSP in 1925.(2)

DFSP has been seen to be associated with rearrangement of chromosome 17 and 22 and they are characterized by a supernumerary ring chromosome composed of hybrid material derived from t(17:22).(10,11). This rearrangement leads to continuous activation of PDGF receptor β protein tyrosine kinase ,via autocrine and paracrine production of its functional ligand.(3)

The tumor usually presents between 20-50 years. The tumor first appears as a single, red to bluish, blanchable, firm cutaneous nodule. During the late stages, the rate of growth accelerates, producing the characteristic protrusion from the skin. The growth rate is variable. Lesions may remain stable for many years or they may grow slowly with periods of accelerated growth. Local recurrences occur in 20-55% of the cases (4).

Diagnosis may be suspected on the basis of the tumor's clinical appearance, while physical examination may assess the extension of the tumor. Lymphatic or haematogeneous dissemination is rare , however lymph nodes are assessed by palpation (5).

Murphey et al. and Moureau-Zabottoa et al. studied the clinical characteristics at early stage and they classified 3 different forms of non-protruding DFSP: (i) morphea-like, characterized by the formation of a white or brown indurated plaque with the appearance of a scar, morphea, morpheaform basal cell carcinoma, or dermatofibroma plaque; (ii) atrophoderma- like, characterized by a soft depressed white or brown plaque that appears similar to atrophoderma or anetoderma; and (iii) angioma-like, the least common form, made up of indurated or soft, red or violaceous plaques that have a clinical appearance similar to vascular malformations or such as morphea-like plaques, and congenital cases, such as atrophoderma-like, are more common, particularly when the lesions are located on the trunk(7).

Differential diagnosis includes dermatofibroma, epidermal inclusion cyst, keloid and hypertrophc scar, melanoma, morphea, lymphoma and fibrosarcoma(8).

Table 1- Immunohistochemical difference between dermatofibroma and dermatofibrosarcoma protuberans (DFSP)

	DFSP	Dermatofibroma
CD34	+	+/-
Cd44	-	+
XIIIa	-	+
P75	+ (95%)	-

DFSP demonstrates strong CD34 staining with immunohistochemistry. In the pigmented

variant of DFSP, also known as Bednar tumor, the melanin-containing dendritic cells are scattered between the neoplastic spindle-shaped cells.(9)

Table 2 – immunohistochemical and cytogenetic markers of dermatofibrosarcoma protuberans(DFSP)

Markers	COMMENTS
Cd34and vimentin	Immunostaining helps in diagnosis of DFSP
Cd117	Immunostaining used only for GIST susceptibility To Imatinib
Apolipoprotein B	Immunostaining new marker for diagnosis of DFSP
COL1A1- PDGF-β	FISH Confirms probable suspectibilty to imatinib;diagnosis of DFSP if loss of CD34 is observed.
t(17:22)	RT-PR Confirms probable susceptibility to imatinib; diagnosis of DFSP if loss of CD34 is observed

Magnetic resonance imaging (MRI) is very useful for the estimation of the tumor invasion, mainly in cases of large tumors or large recurrent lesions (6). Conventional T1-weighted images show hypointense lesions compared to subcutaneous fat however it can be hard to separate DFSP from fat on conventional T2 weighted images without fat saturation. CT scan is not indicated for DFSP diagnosis except when underlying bone involvement or pulmonary metastasis is suspected (5).

DFSP is a locally aggressive tumor characterized by high capacity for local invasion and low rate of metastasis. In the early 1990s, the treatment of choice was resection with wide margins of 3-5cms, but the recurrence rate was about 43%. The idea of using more wider margins led to the loss of more healthy tissue and histomorphometric studies showed DFSP doesn't follow a concentric growth pattern and therefore a vertical section might not remove a potentially asymmetric part of the tumor.In 1970's Moh's introduced the micrographic surgical technique which improved the treatment for DFSP.(12)

MOHS MICROGRAPHIC TECHNIQUE-

Mohs technique is an attractive surgical option as it is an intra-operative procedure which gives a continuous surgical margin and will allow sparing of disease free tissue (13, 14). MMS is very well suited in aesthetic zones where wide resection may not be advisable. Recurrent DFSP's tends to grow deeper and MMS is not indicated in those cases. When the lesion is very close to the bone, a portion of the periosteum as well as underlying bone needs to be removed (15).

DFSP is a radiosensitive tumor but sufficient literature could not be seen on its use as a adjuvant treatment modality (16, 17). In MD Anderson Cancer centre, a study was done involving 19 patients who received adjuvant radiotherapy after surgical resection. The local control rate achieved was 95% at 10 years (18).

Imatinib at a dose of 800 mg/day was granted approval as a single agent in the treatment of DFSP by the FDA in 2006. Previously , Imatinib was used in the treatment of gastrointestinal stromal tumors(GIST) and chronic myelogenous leukemia. Imatinib is an orally active selective tyrosine kinase inhibitor (19,20).

CONCLUSION-

DFSP of the head and neck region can be challenging to the head and neck surgeon when extensive margins needs to be balanced with aesthetics and vital structures. Surgical resection with adequate margins is the gold standard in the management of DFSP's. Advances in head and neck reconstructive surgery has helped to counter this difficulty but the question of "what would be the exact margins" still remains to be answered. Moh's micrographic surgery (MMS) when available will help to reduce the resection of healthy tissue margins. In recurrent DFSP'S adjunctive radiotherapy after surgical resection should be considered. Imatinib has shown promising results and can be considered when it comes to DFSP's in the head and neck region as well as peripheral limbs where enbloc resection may be difficult.

REFERENCES-

1. Cabo H, de Montes de Oca NF, Palmiero G, Tzovanis MC, Lutzky F, Bacaloni E, et al. Darier-Ferrand progressive recurrent dermatofibroma. Med Cutan Ibero Lat Am. 1986;14:186–92.
2. Laskin WB. Dermatofibrosarcoma protuberans. CA Cancer J Clin. 1992;42:116–25.
3. Maire G, Pédeutour F, Coindre JM. COL1A1-PDGFB gene fusion demonstrates a common histogenetic origin for dermatofibrosarcoma protuberans and its granular cell variant. Am J Surg Pathol. 2002;26:932–7

4. Asuquo ME, Umoh MS, Ebughe G (2007) Dermatofibrosarcoma protuberance: case reports. Ann Afr Med 6: 80-83.
5. Softah A. Recurrence of Dermatofibrosarcoma Protuberans: A Case Report and a Review of Literature. Indian J Chest Dis Allied Sci 2003;45:261-263
6. Murphey M, Arcara L, Fanburg-Smith J. Imaging of musculoskeletal liposarcoma with radiologic-pathologic correlation. RadioGraphics 2005;25:1371-1395
7. Heuvel S, Suurmeijer A, Pras E, Van Ginkel R, Hoekstra H. Dermatofibrosarcoma protuberans: Recurrence is related to the adequacy of surgical margins. Eur J Surg Oncol 2010; 36: 89-94
8. Hirata K, Kanemitsu S, Nakayama Y et al. Novel Germline Mutation of MSH2 in a Hereditary Nonpolyposis Colorectal Cancer Patient with Liposarcoma. Am J Gastroenterol 2006;101:193-196.
9. Quigley EA, Marghoob AA, Busam KJ, Chen CS. A firm red-brown plaque on the arm. Dermatofibrosarcoma protuberans(DFSP), pigmented variant (Bednar tumor. Arch Dermatol 2009;145:589-594.)
10. Murphey M, Arcara L, Fanburg-Smith J. Imaging of musculoskeletal liposarcoma with radiologic-pathologic correlation. RadioGraphics 2005;25:1371-1395.
11. Lisovsky M, Hoang M, Dresser K, Kapur P, Bhawan J, Mahalingam M. Apolipoprotein D in CD34-positive and CD34-negative cutaneous neoplasms: a useful marker in differentiating superficial acral fibromyxoma from dermatofibrosarcoma protuberans Mod Pathol 2008;21:31-38.
12. Yu W, Tsoukas MM, Chapman SM, Rosen JM. Surgical Treatment for Dermatofibrosarcoma Protuberans. The Dartmouth Experience and Literature Review. Ann Plast Surg 2008;60:288-293
13. McArthur G. Bone and Soft Tissue Sarcomas Dermatofibrosarcoma Protuberans: Recent Clinical Progress. Ann Surg Oncol 2007;14:2876-2886.
14. Wente MN, Schwarzbach MH, Hinz U et al. Perioperative outcome in sarcoma surgery. Langenbecks Arch Surg 2007;392:83-93.
15. Park TH, Seo SW, Kim JK, Chang CH. Reconstructive challenge of dermatofibrosarcoma protuberans in the female breast. World J Surg Oncol 2011;9:1.
16. McArthur G. Bone and Soft Tissue Sarcomas Dermatofibrosarcoma Protuberans: Recent Clinical Progress. Ann Surg Oncol 2007;14:2876-2886.
17. Chen X, Chen YH, Zhang YL, Guo YM, Bai ZL, Zhao X. Magnetic resonance imaging and mammographic appearance of dermatofibrosarcoma protuberans in a male breast: a case report and literature review. J Med Case Report 2009;3:8246.
18. Ballo MT, Zagars GK, Pisters P, Pollack A. The role of radiation therapy in the management of dermatofibrosarcoma protuberans. Int J Radiat Oncol Biol Phy 1998;40:823-827.
19. Kim GK. Status Report on the Management of Dermatofibrosarcoma Protuberans: Is There a Viable Role for the Use of Imatinib Mesylate? In Which Cases May It Be Therapeutically Helpful and in Which Cases Not ? J Clin Aesthet Dermatol 2011; 4:17-26.
20. FDA approves imatinib mesylate (Gleevec) as a single agent for the treatment of multiple indications 5/11/2009, www.fda.gov.