Asian Journal of Case Reports in Surgery

7(1): 14-20, 2021; Article no.AJCRS.64757

Recurrent Dermatofibrosarcoma Protuberans of Head and Neck: A Rare Case Report

Richa Mehra[1], Munish Kumar Saroch[2] and Amit Saini[3*]

[1]Zonal Hospital, Dhramshala, HP, India.
[2]Department of Otolaryngology, Dr. Rajender Prasad Government Medical College, Tanda, HP, India.
[3]Department of Otolaryngology, Dr. Radhakrishan Govt. Medical College, Hamirpur, HP, India.

Authors' contributions

This work was carried out in collaboration among all authors. Authors AS and MKS designed the study, performed the statistical analysis and wrote the first draft of the manuscript. Author RM wrote the protocol, managed the analyses and literature searches of the study. All authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. Ashish Anand, GV Montgomery Veteran Affairs Medical Center, USA.
Reviewers:
(1) Takuya Maeda, Hokkaido University, Japan.
(2) Ligia Rios, Edgardo Rebagliati Hospital, Perú.
Complete Peer review History: http://www.sdiarticle4.com/review-history/64757

Case Study

Received 20 November 2020
Accepted 25 January 2021
Published 10 February 2021

ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is an uncommon neoplasm of dermis involving head neck region. This tumour is slow growing and has very high potential for local recurrence with less risk of metastasis. It is a very rare neoplasm of the dermis layer of the skin [1] and is classified as a cutaneous soft tissue sarcoma [1]. It accounts for approximately 1–2% of all soft-tissue sarcomas. About 85% to 90% of DFSPs are low-grade tumours. The remaining 10–15% exhibit a high-grade fibrosarcomatous component and are considered high-grade lesions associated with a higher risk of metastasis [2]. We report a case of 46 years old male patient who presented with third episode of DFSP. The first episode was on right side of fore head, the second one was on right frontoparietal region and latest lesion was on right preauricular region and right pinna. On all the three occasions the recurrence was away from primary site.

Keywords: Dermatofibrosarcoma protuberans; recurrence; head and neck.

**Corresponding author: E-mail: dr.saini@rediffmail.com;*

1. INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing, locally aggressive primary soft-tissue tumour with a high rate of local recurrence and low risk of metastasis. It is a very rare neoplasm of the dermis layer of the skin and is classified as a cutaneous soft tissue sarcoma [1]. It accounts for approximately 1–2% of all soft-tissue sarcomas and 85 to 90% of DFSPs are low-grade tumours. The remaining 10–15% exhibit a high-grade fibrosarcomatous component in more than 5% of tumour volume, and are considered high-grade lesions associated with a higher risk of metastasis [2]. Treatment of choice for DFSP is wide local excision with negative margins of 3 cm from the tumour border (including the underlying skin, the subcutaneous tissue and fascia); and includes the periosteum or even a part of the bone in more complex cases. The rate of inadequate initial surgical resection with positive margins can be as high as 83%, which significantly increases the risk of recurrence. In DFSP resected with wide margins, the likelihood of recurrence is <10%, but increases to >50% with positive margins.

The interval between treatment and development of recurrent disease is highly variable, ranging from 1 to 100 months, with the average time reportedly being around 35–40 months [2].

2. CASE REPORT

A 46 years old male patient presented to Otolaryngology clinic with a protuberant mass on right ear. The patient reported an increase in size of the mass in the preceding 3 months, there was no associated pain. Upon examination an approximately 6*6 cm large multinodular mass ,reddish purple coloured with few telengiectasias ,firm in consistency and non-tender on palpation was found on the right preauricular area and right pinna. There was past history of excision of a similar lump on the right fronto-parietal region two years back and another lump on the right side forehead three years back, which on histological examination were found to be Dermatofibrosarcoma protuberance. On both occasions the resection margins were negative for tumour.

Contrast enhanced computed tomography shows lobulated heterogeneously enhancing mass in the region of right pinna measuring 3.6*3.1*3.4 cm, extending into the right external auditory canal and the right superficial lobe of parotid. Few calcified foci are seen within the lesion, with loss of fat planes with the right Sternocleidomastoid muscle.

Patient underwent a wide surgical excision under general anaesthesia with a wide margin of 3 centimetres along with superficial parotidectomy.

3. DISCUSSION

DFSP is a rare neoplasm of intermediate malignancy. Taylor first described it in 1890 but Darier was credited with establishing DFSP as a distinct clinical pathological entity in 1924, and finally Hoffman established the term in 1925 [3]. Overall annual incidence has been estimated to be 4.2 per million and the tumour accounts for approximately 0.1% of all malignancies [3]. Its annual incidence is between 0.8 and 4.5 per million and represents the most common skin sarcoma [4]. The incidence is almost double among blacks compared to whites and women have a higher incidence rate than men; the highest age-specific annual incidence rates are observed between the ages of 30 and 50 years. Most occur on the trunk (42%), followed by the upper extremities (23%), lower extremities (18%), and head and neck (16%) [3]. It can develop at site of previous trauma, burn scar, site of vaccination and areas with previous radiotherapy. There is an association between DFSP and children with adenosine deaminase deficient severe combined immunodeficiency, with presentation in early age and often multicentric. The risk of metastasis in DFSP is about 3% [5].

Commonly presenting as a painless plaque, it often progresses to form a nodule. Initially fixed within the dermis, the tumour follows a radial growth pattern in finger-like projections, eventually invading underlying structures, and potentially ulcerating the epidermis [6]. The histologic features of DFSP are those of a monotonous storiform architecture of uniform bland spindle cells centred in the dermis and locally infiltrative. Fine needle aspiration is unlikely to yield diagnostic results owing to the inherent difficulty of acquiring suitable cellularity from densely fibrotic mass. Fibrosarcomatous DFSP is similar to fibrosarcoma and are more common in recurrent tumours and has high metastatic spread. The cytogenetic abnormality in DFSP is manifested as translocation t(17;22)(q22;q13).This results in fusion of *COL1A1* (also located at 17q21) and *PDGFB* genes [4].The resultant fusion gene disallows

normal repression of transcription and translation of *PDGFB.* Metastasis to lymph nodes and internal organs tends to be extremely rare in pure DFSP, but can occur in up to 13% cases of fibrosarcomatous transformation. The risk of local recurrence was 29.8% for the DFSP Fibrosarcomatous-group vs. 13.7% for DFSP; the risk for metastasis was 14.4 vs. 1.1% and death from disease was 14.7 vs. 0.8% [3].

The standard of care for DFSP is surgical resection with negative margins. Wide surgical resection of the lesion with 2-3 cm of surrounding healthy tissue is still the first-line therapy. It is reported that wide excision of 3cm margins decreases the rate of recurrence significantly [7]. Because the shape of these tumours is always irregular, with finger-like extensions that cannot be observed with the naked eye, the recurrence rates after surgical excision range from 11% to 53% [8]. Inadequate surgical margins lead to high recurrence rates. Even more, a recurrence rate of up to 20% with 3 cm surgical margins has been described. Tumours occurring in head and neck are challenging because of the risk of disfigurement and functional impairment. Positive margins are more prevalent in DFSPs occurring in the head and neck compared with other regions, due to the anatomical constraint [2]. Mohs' micrographic surgery which focuses on minimal removal of normal tissues also shows low recurrence. This is of particular significance in head and neck region [9]. Adjuvant radiotherapy has a role for unresectable DFSP and in those cases with positive margins when re-excision is not feasible. Imatinib, a tyrosine kinase inhibitor is the first effective systemic therapy for advanced DFSP and could potentially be used for reducing tumour size in those considered initially unresectable so surgery may be feasible [10]. HIV infected patients can also present with aggressive disease [11].

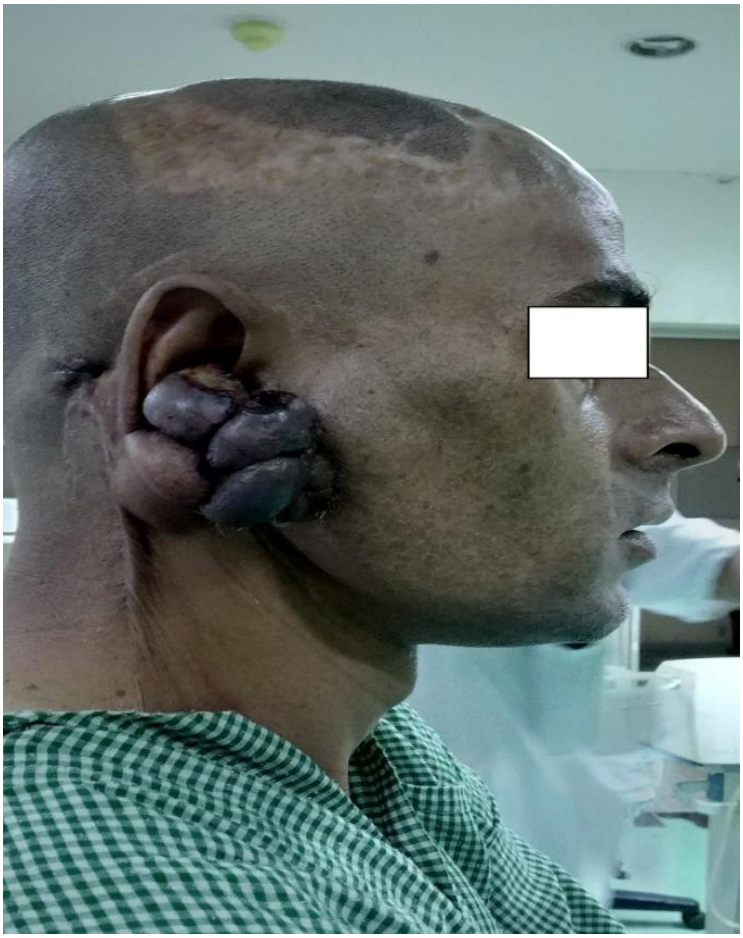

Fig. 1. Clinical photograph of the patient showing the tumour

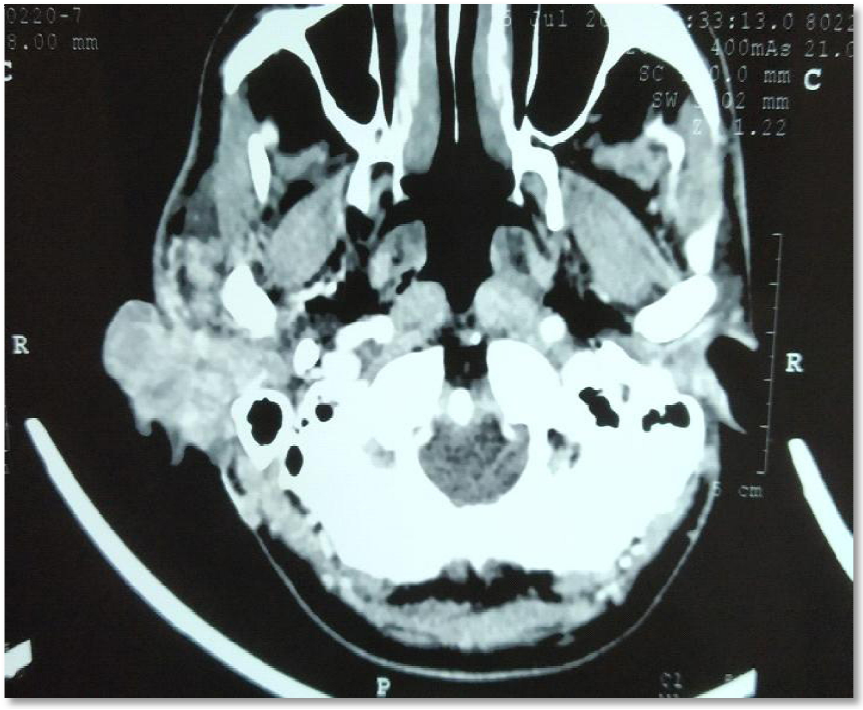

Fig. 2. Contrast enhanced axial computed tomography image showing the lobulated heterogeneously enhancing mass in the region of right pinna

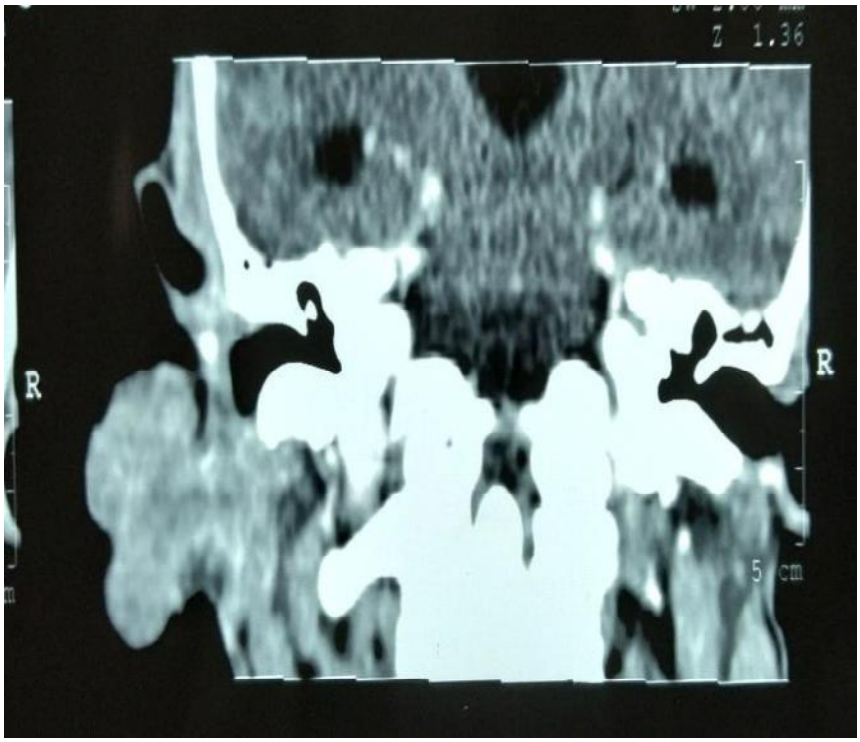

Fig. 3. Contrast enhanced coronal CT image showing the involvement of right external auditory canal and right parotid

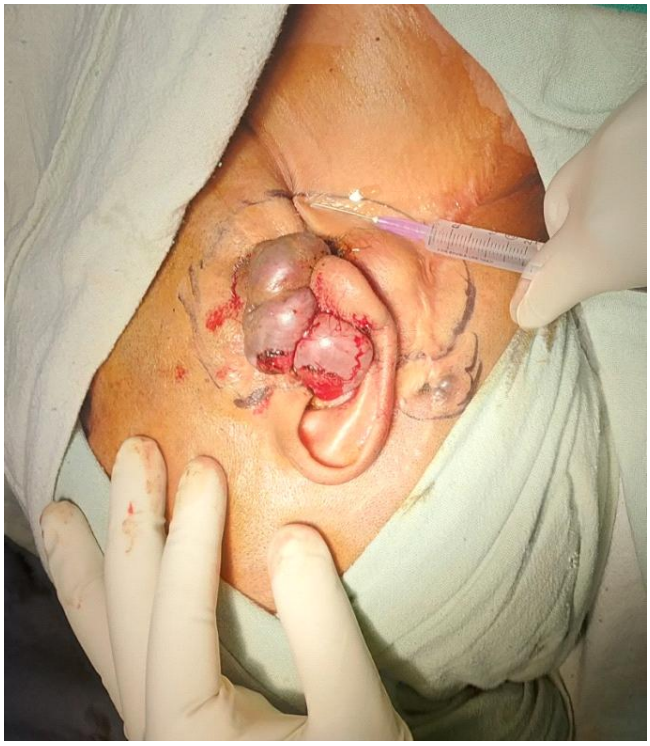

Fig. 4. Marked incision line taking wide safe margins

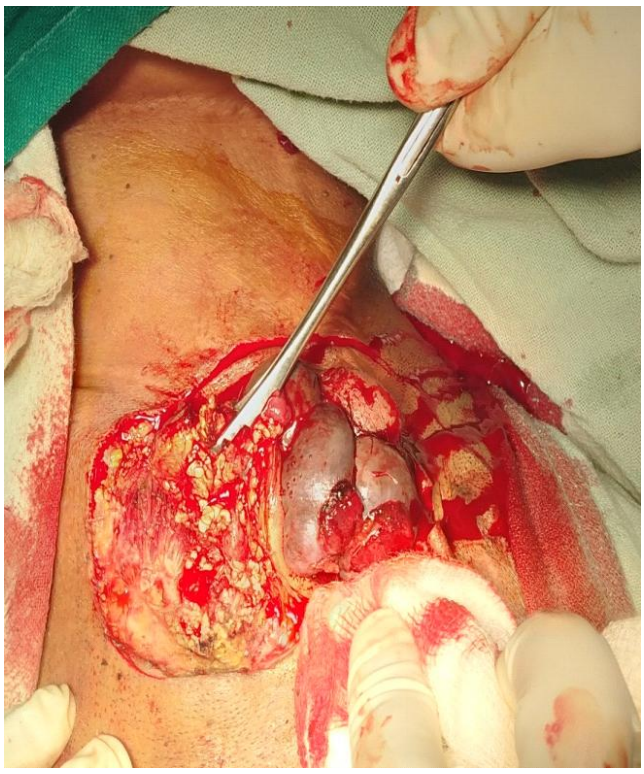

Fig. 5. Wide surgical resection done along with superficial parotidectomy

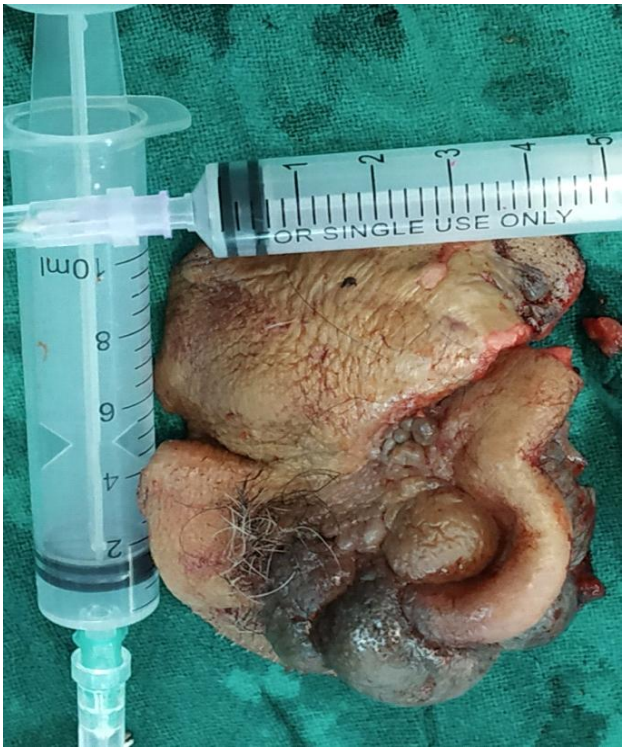

Fig. 6. The excised tumour

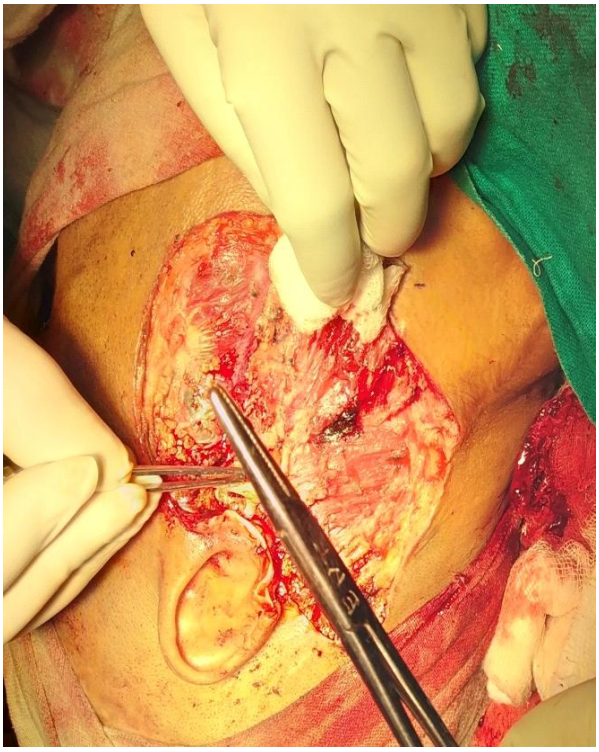

Fig. 7. Defect after removal of enblock tumour with wide surgical margins

4. CONCLUSION

Dermatofibrosarcoma protuberans is a rare tumour of head neck region with high potential of local recurrence and low risk of distant metastasis. The present case report involves the patient with third regional recurrence though the resection margins were negative in previous surgeries. This raises the possibility of regional spread in case of DFSP.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Terro W, Hamadeh N, Terro M. Management of dermatofibrosarcoma protuberans of the face using lower trapezius musculocutaneous pedicle flap reconstruction: A case report. Journal of Surgical Case Reports. 2018;6:1–3.
2. Tan YG, Chia CS, Loh WL, Teo M. Single-institution review of managing dermatofibrosarcoma protuberans. Royal Australasian College of Surgeons; 2015.
3. Acosta AE, Velez CS. Dermatofibrosarcoma Protuberans. Curr Treat Options in Oncol. 2017;18:56-60.
4. Perez GC, Arias C, Luna P, Soren I, Mazzuoccolo LD. A Challenging Giant Dermatofibrosarcoma Protuberans on the Face: Case report. Case Reports in Dermatological Medicine; 2016.
5. Llombart B, Serra C, Requena C, Alsina M, Morgado-Carrasco D, Través V, Sanmartín O. Guidelines for Diagnosis and Treatment of Cutaneous Sarcomas: Dermatofibrosarcoma Protuberans. English, Spanish. Actas Dermosifiliogr. 2018;109(10):868-877. DOI: 10.1016/j.ad.2018.05.006. PMID: 30539729.
6. Perez G, Glenda L, Montague, Bhayani MK. New Facial Weakness after 5 years of facial asymmetry. JAMA Otolaryngology–Head & Neck Surgery. 2016;142:1120-1122.
7. Hegde U, Shetty S, Shreesahyla H, Doddawada V. Dermatofibrosarcoma Protuberans –A Recurrent Lesion with Unusual Presentation in the Parotid Region. Journal of Clinical and Diagnostic Research. 2014 ;8(3):130-131.
8. Jiongyuan X, Jialiang L, Xulong Z, Jianying Z, Fei Y, Yuan W, et al. Cryotherapy for local recurrent dermatofibrosarcoma protuberans: Experience in 19 patients. Cryobiology. 2014;68:134–138.
9. Gloster HM, Harris KR, Roenigk RK. A comparison between Moh`s micrographic surgery and wide surgical excision for the treatment of dermatofibrosarcoma protuberans. J Am Acad Dermatol. 1996 ;35:82.
10. Allen A, Ahn C, Sangüeza OP. Dermatofibrosarcoma protuberans. dermatol clin. 2019;37(4):483-488. DOI: 10.1016/j.det.2019.05.006. PMID: 31466588.
11. Al-Mendalawi MD. Dermatofibrosarcoma protuberans: Two rare variants. Indian J Dermatol Venereol Leprol. 2019;85(4):412. DOI: 10.4103/ijdvl.IJDVL_234_19. PMID: 31134900.

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/64757