CASE REPORT

Recurrent Dermatofibrosarcoma Protuberans of the Parotid: A case report and review of literature

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Abstract

In 1924, Darier and Ferrand described Dermatofibrosarcoma Protuberans as a progressive and recurring dermatofibroma. It is a locally aggressive sarcoma originating from dermal and subdermal tissue of the skin. It usually begins as a small plaque that grows over a period and later manifests as multiple small subcutaneous nodules. It is more commonly found in females as compared to males and typically occurs in between 2nd and 5th decades of life. Most frequently involved regions of the body are torso and proximal ends of extremities and very rarely head and neck region is the site of involvement. The mainstay of treatment of this entity is surgery. The rate of recurrence of this disease is very high in about 50% of the cases and it may also express rare distant metastasis. It is a radiosensitive tumour and radiation may play a role in reducing risk of recurrence. We present a case of a 35 years old male with recurrent Dermatofibrosarcoma Protuberans of right parotid gland.

Keywords: Parotid gland, Dermatofibrosarcoma, Spindle cell lesion, Recurrence, Salivary gland.

Introduction

The history of dermatofibrosarcomaprotuberans (DFSP) dates back to 1924 when Darier and Ferrand, in 1924, initially defined it as "progressive and recurring dermatofibroma".[1] It originates from dermal and subdermal tissue of the skin and is locally aggressive soft tissue sarcoma.[2] The annual incidence rate of DFSP is 4.1 persons per million person-years. Incidence among women is 1.14 times higher than in men,[3] and it typically occurs in early mid adult life i.e.; 2nd to 5th decade. It can involve any part of the body but the torso and proximal end of extremities are the most frequent sites of its incidence.[4] Head and neck region is a quite rare region for this tumour to occur.[5,6] Recurrence rate of DFSP is about 50% and it also expresses rare distant metastasis.[7] The gold standard treatment for this locally aggressive tumour is surgery.[8]

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Case Report

A 35-year-old man came to the Otolaryngology outpatient clinic at Aga Khan University Hospital, Karachi in November 2016 with history of a slowly enlarging mass in the right pre-auricular region for one and half year. On physical examination, the mass was around 4 x 5 cm in size, located in the right parotid region and it was firm, mobile and non-tender. There was no associated pain, discharge, fever, or trismus and no remarkable findings on oral examination. Facial nerve was also intact. The rest of the head and neck examination was normal. The chest X-ray and laboratory examination were unremarkable with no evidence of metastatic disease. Initially an ultrasound was done that revealed irregular, heterogeneous mass of 5 x 4.2 cm lying superficial to the parotid gland. The fine needle aspiration biopsy of the mass showed spindle cell lesion.

The past history of the patient is significant in a view that five years back he was operated for a mass in right parotid gland. The final histopathology showed solid and smooth cut surface of gray-white color, measuring 3 x 2 x 1 cm. Final histopathology reported it to be spindle/fusiform cell sarcoma, consistent with low grade fibrosarcoma.

Keeping all the parameters in view, the patient was treated

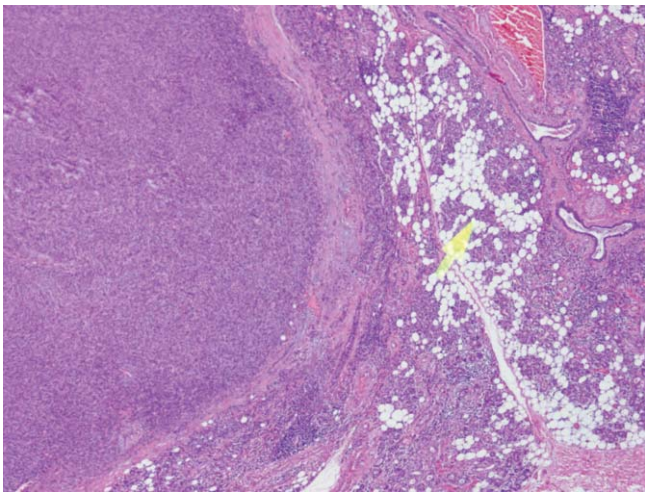

Figure-1: Photomicrograph showing a cellular lesion closing abutting the salivary gland parenchyma. (H&E staining, X40).

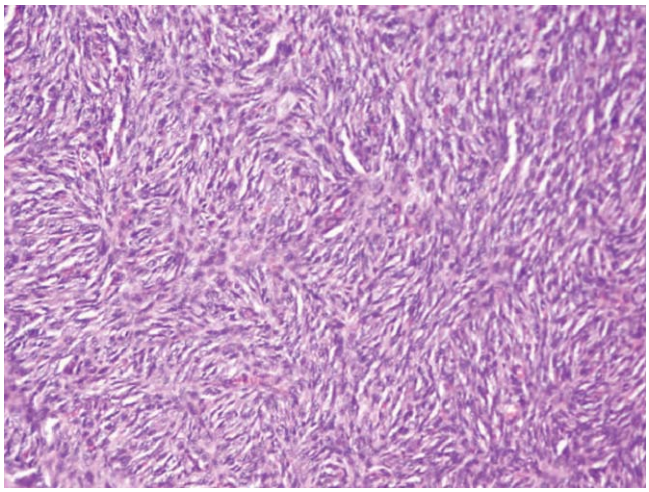

Figure-2: Photomicrograph showing cellular spindle cell lesion with storiform pattern. (H&E staining, X200).

with right superficial parotidectomy. Post-operative period was uneventful and facial nerve of the patient was saved. Gross examination revealed a tan white firm lesion measuring 5X4.5X3.5cm. The tumour was 0.1 cm away from inked excision margin. Light microscopic examination revealed a highly cellular tumour closely abutting the salivary gland tissue (Figure-1). The tumour showed plump monomorphic spindle cells with vesicular nuclei, inconspicuous nucleoli arranged in a storiformpattern (Figure-2). Mitotic count of 5-6/10HPF was noted in cellular areas. There was lesional entrapment of adipose tissue. The immunohistochemical finding of diffuse positivity of tumour cells with CD34+ helped in arriving at the final conclusive diagnosis of Dermatofibrosarcomaprotuberans (DFSP). The patient was followed in the clinic after one year and has had no recurrence till date along with intact facial nerve.

Discussion

Firstly defined to be "progressive and recurring dermatofibroma" by Darier and Ferrand in 1924, DFSP begins usually as a plaque that slowly grows over several years. It later manifests as multiple small subcutaneous nodules. Therefore, the "protuberant" appearance of DFSP can only be witnessed in the fully developed lesions.[9] Usually males between 20 and 50 years get affected by DFSP. The most common location of its occurrence is trunk (47%), followed by lower extremity (20%), upper extremity (18%), and then head and neck (14%).[10]

DFSP is a locally aggressive tumour; it tends to grow in a more infiltrative manner. The tumour can assume irregular shapes and extend in a villous or finger-like manner.[11] These irregular, tentacle-like extensions of DFSP are supposed to be responsible for the local recurrence following inadequate resection.[12]

Histopathologically, DFSP is poorly circumscribed with tumour cells infiltrating diffusely into dermis and subcutis. The tumour cells are composed of uniform population of monomorphic spindle cells arranged in storiform pattern with mitotic figures not exceeding 5 per 10 HPF.[13]

The American Joint Committee on Cancer has not established a staging system for Dermatofibrosarcoma Protuberans (DFSP). The following staging system published in "Short German guidelines: dermatofibrosarcomaprotuberans" is helpful clinically,[14]

- Stage I - Primary tumour, localized disease
- Stage II - Lymph node metastasis
- Stage III - Distal metastasis[14]

The mainstay of treatment for DFSP remains to be surgery with wide local excision with gross margins of 2 centimeters. But this may not be followed in the cases where it involves the head and neck region, where excision tends to have smaller margins and therefore likelihood of positive margins is greater.[15] DFSP is rarely seen to be metastasized, but the chances of metastasis increase with the increase in diameter. Tumours with size 10cm or above have greater chances to metastasize.[16]

Conventional chemotherapy appears to offer little utility, however, treatment with the molecular targeted therapy Imatinib, has yielded some limited but encouraging results to date.[12] DFSP is known to be a radio-sensitive tumour and several studies have inspected the use of radiotherapy in the treatment of DFSP. Haas et al. studied 21 patients that were treated only with surgery had a local control of 67% while 17 patients who were treated with combined therapy (surgery and radiation therapy) had local control of 82%.[17] Radiation therapy may be recommended for patients if the margins of resection are positive or for situations in which adequate wide excision alone may end up in major cosmetic or functional deficits. Williams et al. reported outcomes of 14 patients with DFSP who were treated with radiation therapy with curative intent, 12 patients remained disease free from 1.0 to 23.5 years after treatment (11 received postoperative radiation therapy and 1 received radiation therapy alone). No severe complications occurred after radiation.[18] Postoperative adjuvant radiation therapy may reduce the risk of recurrence when clear surgical margins are not obtained.[19] In 2015, De liz et al reported a case of 37 years old female

who underwent surgery followed by radiation therapy and remained free of disease after 5 years of follow up.[20] Hedge et al reported a case of 42 years old lady with DFSP of parotid region who developed recurrence after 9 months of surgical excision. Re-excision of the tumour was performed and patient was referred for radiation therapy.[21]

Conclusion

Dermatofibrosarcoma Protuberance is a locally aggressive soft tissue sarcoma originating from dermal and subdermal tissue of the skin. It rarely occurs in the head and neck region. We report here on a case of DFSP of parotid gland in a 35 year old gentleman who underwent surgery and remained recurrence free till date.

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Conflict of Interest: This case report has no conflict of interest.

Ethical Approval: All procedures performed on the patient were in accordance to the ethical standards of the institution research committee.

Informed Consent: Written informed consent was obtained from the patient for publication of this case in the text.

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