Dermatofibrosarcoma Protuberance (DFSP) of Parotid Gland – A Case Report

Afshan Aslam[1], Hira Andleeb[1*] and Shakil Aqil[1]

[1]*Department of E.N.T-Head and Neck Surgery, Liaquat National Hospital and Medical College, Karachi, Pakistan*

Abstract

Dermatofibrosarcoma protuberans (DFSP) is a rare slow-growing low-grade dermal sarcoma typically found on the chest or proximal extremities. Uncommon sites include the head, face, and neck area. This tumor is locally invasive but has low distant metastasis. DFSP shows CD34 immuno-reactivity and hence required tissue biopsy for diagnosis. The case describes an unusual occurrence of dermatofibrosarcoma protuberance arising from the parotid gland.

Keywords: *Dermatofibrosarcoma protuberance, parotid swelling, parotid malignancy, mononuclear spindle cells.*

INTRODUCTION

In 1924 Dermatofibrosarcoma protuberans (DFSP) was first named by Darier and Ferrand [1]. It is a rare slow-growing, dermal soft-tissue tumor. It accounts for < 1 percent of malignant head and neck tumors and 7 percent of all head and neck sarcomas [2, 3]. Its incidence is 1.14 times higher in women [1]. Nearly 90% of all DFSPs exemplify low-grade tumors, but the residual 10% encompasses a high-grade fibrosarcoma [3]. It has a high chance of local recurrence rate but less distant metastatic potential [3, 4]. On histology it has a spindle cell pattern, with a storiform arrangement and CD34 immunoreactivity [4]. The most frequently involved sites include the chest and proximal end of extremities [5]. Only seven cases are reported in the literature of Dermatofibrosarcoma protuberans arising in the parotid gland [1, 2]. The standard treatment is whole tumor excision with wide tumor-free margins and inoperable DFSPs are treated with radiation therapy [4].

CASE REPORT

A 61-year-old female with no known comorbid presented with right gradually progressive parotid region swelling for the last 3 years, with no history of pain or discharge from the swelling. On examination, there was a 7 x 7 cm soft non-tender swelling in the right parotid area, which was extended to the retroaurical region, involving the ear lobule and compressing the tragus with overlying skin showing prominent vessels. Fungating nodule of approx. 2 x 2cm with overlying skin fixed at the inferior anterior part of the main parotid swelling as shown in **Fig. (1)**. The facial nerve preoperatively was normal and the rest of the examination was unremarkable. She advised getting an ultrasound-guided fine needle aspiration that showed pleomorphic adenoma.

**Corresponding author: Afshan Aslam, Department of E.N.T-Head and Neck Surgery, Liaquat National Hospital and Medical College, Karachi, Sindh, Pakistan; Email: dr.afshan.aslam@hotmail.com*

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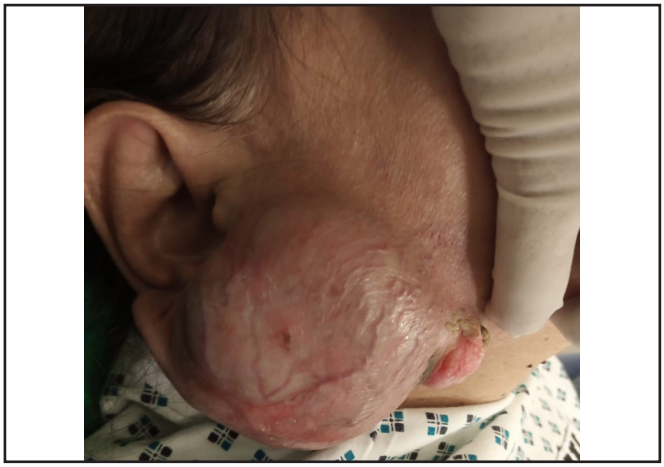

Fig. (1): shows a pre-operative picture of swelling of the right parotid region.

She had contrast-enhanced computed tomography (CT) of the head and neck that showed a mass measuring 4.9 x 3.7 cm involving the right parotid gland along with overlying skin and soft tissue, with deep lobe spared (**Fig. 2**).

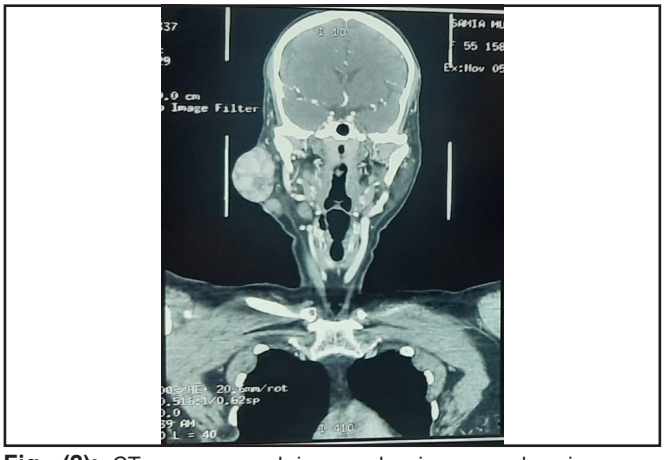

Fig. (2): CT scan coronal image showing an enhancing mass involving right parotid gland.

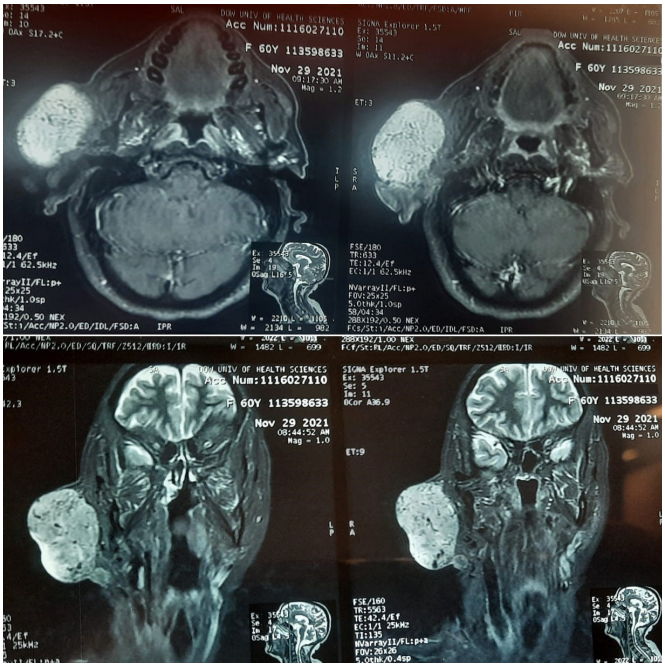

Fig. (3): MRI scan of a patient showing a hyperintense lesion involving the right superficial lobe of the parotid gland.

Contrast-enhanced MRA showed an oval-shaped mass measuring 7.3 x 4.7 x 7.0cm involving the superficial lobe of the right parotid gland, with a perilesional lobulated lesion measuring 1.5 x 1.8 x 1.6cm. No significant lymphadenopathy was noted (**Fig. 3**).

She was prepared for the right superficial parotidectomy. Her baseline workup was unremarkable. Modified Blair incision is given. Per operative, findings were tumor approx. 8x7cm involving the right superficial parotid gland extending up to the lobule of the ear. It was mixed with cystic firm components. Overlying skin adherent tightly to it and inseparable in middle. The overlying skin at the inferior of swelling was fixed, so removed along with the skin. All branches of the facial nerve were identified and saved but the buccal branch was involved by the tumor so scarified. The specimen was sent for histology (**Fig. 4**) and a free flap was done by the plastic surgery team. Final histology reported a Low-Grade Spindle Cell Neoplasm most likely Dermatofibrosacroma Protuberance with tumor size 8 x 6 CM, with following surgical margins

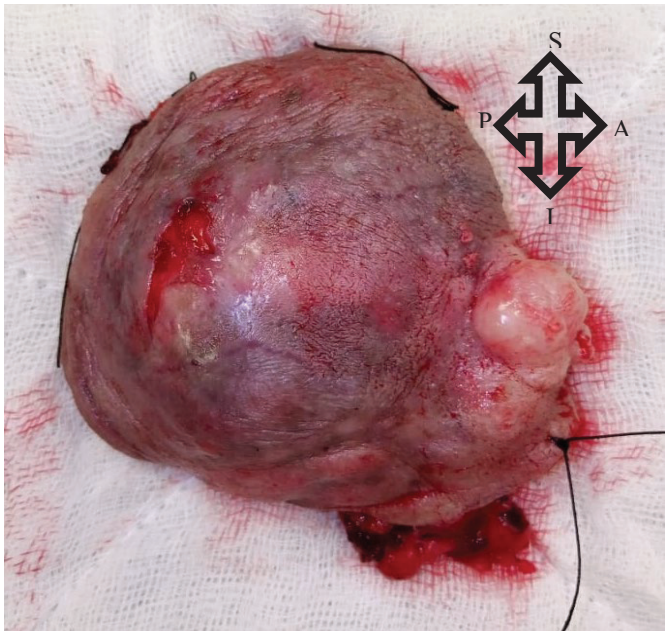

Fig. (4): Shows a per-operative picture of the specimen. S= superior, I = inferior, A= anterior and P= posterior orientation of specimen.

- Anterior: skin 0.6cm away — soft tissue 0.3cm
- Inferior: skin 0.2cm — soft tissue reaching–extended soft tissue 0.3cm
- Posterior: skin 0.2cm — soft tissue reaching–extended soft tissue tumor free
- Superior: skin tumor-free — soft tissue reaching – extended soft tissue tumor free
- Deep margin: 0.6cm

According to the American Musculoskeletal Tumor Society (MSTS), this classifies as Stage IA. The case was discussed in a multi-disciplinary tumor board meeting and was advised radiation therapy. For distant metastasis PET Scan was clear. No signs and symptoms of recurrence are up to date on the last follow-up visit in September 2022.

DISCUSSION

Literature suggested that to reduce the rate of recurrence one needs to clear margins of 3cm [3]. However, this cannot be followed in head and neck region involvement [1]. In head and neck excision, it poses difficulty in management due to the critical structures and esthetic difficulties in reconstruction, and excision tends to have smaller margins therefore the likelihood of positive margins is greater [1, 3]. The gold standard is wide surgical excision of the tumor, and in cases of positive or close margin or recurrence radiation therapy is advised [1, 3]. For distant metastatic disease chemotherapy role is suggested [6]. There is no established staging system for DFSP by American Joint Committee on Cancer [1]. The Short German guideline is the most widely used classification system for DFS, which is the stage I - Primary tumor, localized disease, Stage II - Lymph node metastasis, and Stage III - Distal metastasis [1, 3]. Another classification system suggested by the American Musculoskeletal Tumor Society (MSTS), that classify Stage IA tumors as low grade without involving the subcutaneous compartment, and stage IB tumors are low grade which extends extra-compartmental and involves the fascia, muscle, or causing bone erosion [3].

CONCLUSION

DFSP is a locally destructive soft tissue tumor arising from dermal tissue that also involves a subdermal layer of skin. We presented here a case of the parotid gland, in which the rarity of the site makes it difficult to diagnose.

However, DFSP is mainly diagnosed on histology by applying Immunohistochemical stains. The above-reported case has remained recurrence-free to date.

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CONSENT FOR PUBLICATION

Written informed consent was taken from the patient for publication of this case in the text. We have ensured to not report any potentially identifying information about the patient in the manuscript

DECLARATION OF CONFLICTING INTEREST

The author(s) declared no potential conflicts of interest.

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