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Case Report

A challenging fibrosarcomatous variant of dermatofibrosarcoma protuberans of head and neck: a case report

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is an unusual tumour of soft tissue with local invasive property with high rate of recurrence after surgical treatment. DFSP is frequently seen on the trunk and proximal extremities, although a 10-15% cases accounts in head and neck region. Mainstay for success of surgery in DFSP remains complete removal of tumour with adequate surgical margins of approximately 3cms. In head neck region challenges associated with surgery may be inadequate surgical margins, poor functional and cosmetic outcomes. We present a case of a 35 years old male reported to our hospital with complaint of rapidly growing mass over right cheek and retroauricular region causing facial deformity. Incisional biopsy diagnosis was made DFSP. The patient underwent wide local excision of tumour followed by adjuvant radiotherapy. Final histopatholgy showed fibrosarcomatous transformation of DFSP. After 24 months of post-op follow up, patient recovery was satisfactory without any signs of recurrence. This case is presented for its rarity and it highlights the need for proper diagnosis and treatment plan.

Keywords: DFSP, Fibrosarcoma, Head and neck region, Sarcoma

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a most frequent skin sarcoma and accounts as low grade malignant tumour.[1] DFSP originates from dermal stem cell or an undifferentiated mesenchymal cell with fibroblastic, neurological and muscular features.[2] DFSP reports less than 0.1% of all malignancies and ranges between 1-2% of all soft tissue sarcoma, with 4.2 per million annual incidence rate.[3] It typically represents as a small, painless, slow-growing firm patch of skin which is usually purplish, reddish, or flesh colored in the appearance. Early slow growth of tumour is seen followed by the accelerated growth with the typical protuberant nodules.[4]

In 1924 Darier and Ferrand detailed clinicopathological description of DFSP as a distinct disease process. Hoffman coined the term DFSP in 1925, which till date is preferred terminology for this disease.[5]

DFSP is locally aggressive tumour with 2-6% of distant metastasis to the lungs.[6] The 5-10% of DFSP cases show fibrosarcomatous transformation, it shows usually more aggressive with high chances of local recurrences occurring in 50% of patients and 10-15% of patients have distant metastasis.[7] Even though fibrosarcomatous variant of DFSP is aggressive but overall survival is satisfactory if adequate surgical margins are taken during surgical excision.[6] In cases of unresectable tumours or tumour near critical structures where these are high possible chances of close or positive margins, metastatic or

recurrent cases literature advocates adjuvant radiotherapy and systemic therapy.[8]

We present rare case of fibrosarcomatous variant of DFSP of right cheek and retroauricular region which was treated surgically followed by adjuvant radiotherapy.

CASE REPORT

A 35 year old male reported to department of head and neck oncology with complaint of painless progressively growing nodular growth over right side of face and behind right ear causing facial conspicuous asymmetry of face (Figure 1). On examination there were two swellings one on right cheek region and another over retro-auricular region. Swelling of cheek was approximately 15×12 cm with bosselated surface and purplish discoloration of overlying skin with telangiectasia. Swelling of retro-auricular region was dome shaped single nodular ranging approximately 9×7 cm with displacing pinna anteriorly. On palpation swelling was non tender, firm, non pulsatile and non compressible in nature with restricted mobility in both horizontal and vertical planes. Facial nerve was functional. No cervical lymphadenopathy was present.

MRI revealed both masses were hypointense on T1W, heterogeneous hyperintense on T2W and heterogeneous post contrast enhancement. Retro-auricular mass was infiltrating overlying skin and the right sternocleidomastoid muscle was seen lying at the base of the lesion and it did not show any definitive infiltration. Mass over cheek region was infiltrating masseter muscle and there was displacement of submandibular gland seen with slight compression of buccal mucosa.

Incisional diagnostic biopsy was which showed a spindle cell neoplasm of infiltrative nature with cells arranged in a storiform pattern. The initial histomophological diagnosis was that of DFSP (Figure 2). Metastatic workup was done further which did not reveal any regional or distant spread. Patient was undertaken for surgical resection of the masses under GA. Wide elliptical incisions were taken keep in oncological safe margins of 2 cm, resection was extended to muscular, aponeurotic,and periosteal planes. Reconstruction of the right cheek defect was done by forehead flap and retroauricular defect was covered by split thickness skin graft (Figure 3).

The histopathology of resected specimen revealed a spindle cell tumour of infiltrative nature showing areas of storiform pattern and increased vascularity (Figure 4). The resected tumour was quite cellular and showed nuclear atypia with increased mitosis and area of tumour necrosis (Figure 5). These histomorphological features suggested of fibrosarcomatous transformation in DFSP. All the peripheral skin and deep soft tissue surgical margins of resected specimen were free of tumour. On immunohistochemistry the tumour cells showed expression of vimentin and CD 34. The tumour cells were negative for CK, desmin, SMA and S-100. MIB 1 proliferation index was around 10 to 15% (Figure 6). The patient underwent adjuvant radiotherapy treatment with a total radiation dose of 60 Gy in 30 fractions. Patient was followed up regular intervals and after 24 months of post-op follow up, patient recovery was satisfactory without any signs of recurrence.

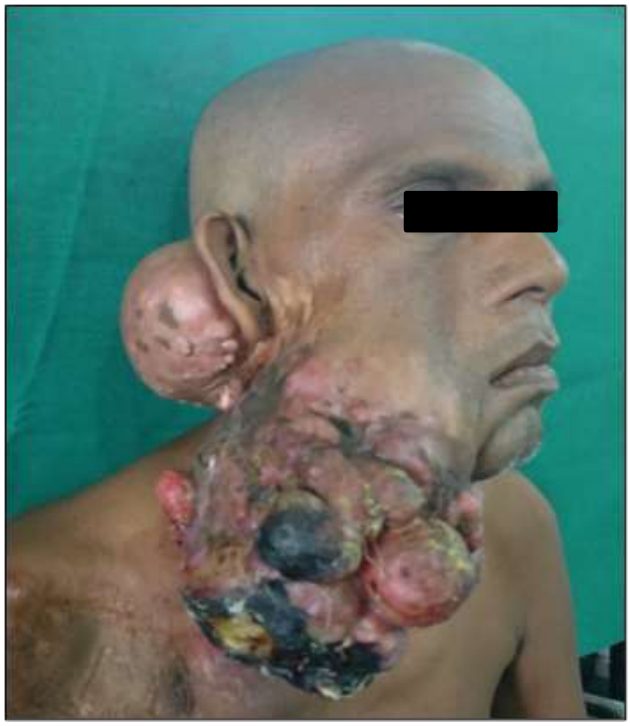

Figure 1: Clinical image of case showing a mass over right side of face and behind right ear.

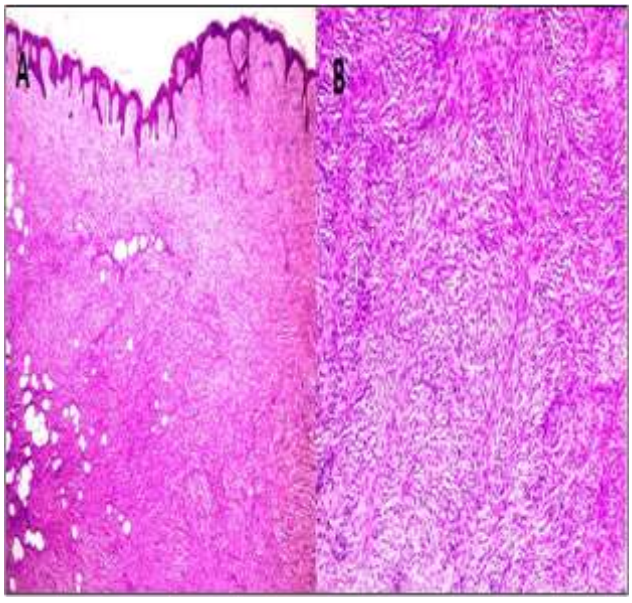

Figure 2 (A and B): H and E images from incisional biopsy of a cellular spindle cell tumour infiltrating the dermis and subcutaneous soft tissue spindle cells are arranged in a storiform pattern (×40, ×100).

Figure 3: Reconstruction of defect done with forehead flap and STSG.

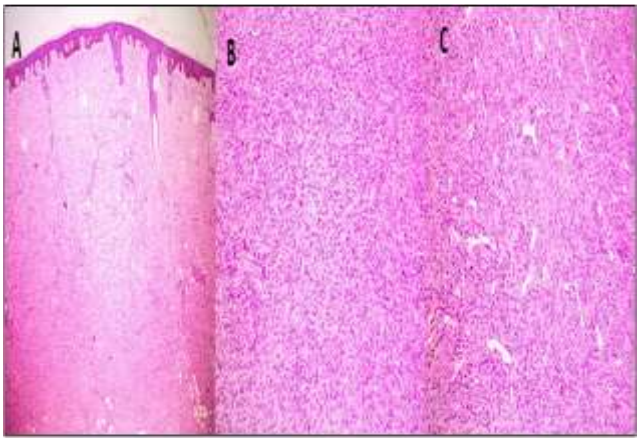

Figure 4 (A-C): H and E images from the resection specimen of a cellular spindle cell tumour of infiltrative nature, tumour shows increased cellularity and in areas shows an increased vascularity of staghorn type (×40 and ×100).

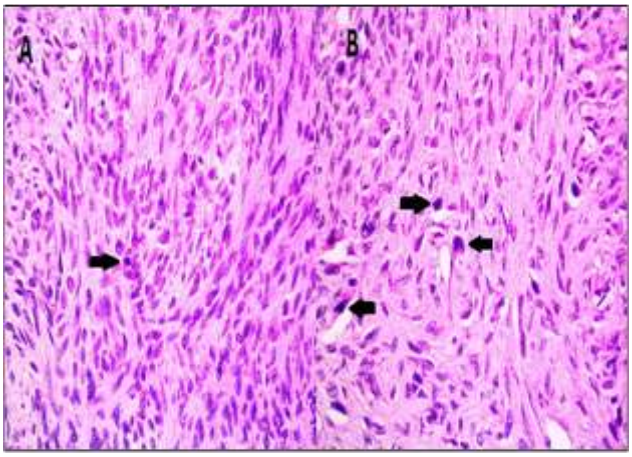

Figure 5 (A and B): H and E images showing increased cellularity, nuclear atypia and increased mitosis (marked by arrows) (×100).

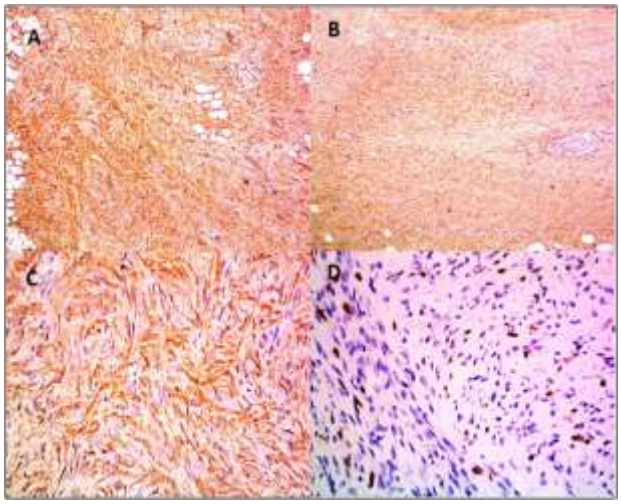

Figure 6 (A-D): IHC images of tumour cells expressing vimentin, CD 34, increased MIB 1 (Ki 67) proliferation index (×100 and ×400).

DISCUSSION

DFSP is low-grade tumour of mesenchymal cell origin which has low metastatic potential but shows high local invasiveness, which causes high rate of recurrence after surgical excision.[4] DFSP constitute of less than 0.1% of all malignancies and 1%-2% as soft tissue sarcomas of head and neck region.[3,5,9,10] It is commonly seen with adults in age group of 30-40 years with male preponderance.[4,5]

The classical presentation of DFSP cases manifests the emanation of a tiny, solid, and subtly stained skin nodule which increase in size over time and coalesce with adjacent nodules to form an indistinct skin plaque. If tumour is not treated on time, it shows slow growth with local infiltration of the surrounding tissues, including fascia, fat, muscles, periosteum, and neurovascular bundles.[8,11] Our case presents a 35 years male who had history of insidious onset painless nodules which gradually increased in size causing facial asymmetry.

Tumour pathogenesis in DFSP occurs due to chromosomal translocation t (17; 22) (q22; q13), which then leads to the fusion of platelet derived growth factor (PDGF) gene and collagen type 1A1 gene causing overproduction of PDGF causing cellular proliferation and tumour formation. This translocation is seen in 90% of DFSP cases.[12,13] A histological type of DFSP includes myxoid, pigmented, giant cell, giant cell fibroblastoma, granular cell, sclerotic, and fibrosarcomatous (FS).[13] Immunohistochemical staining of DFSP shows strong positivity for CD34 and vimentin.[1,13] In our case histopathology revealed a fibrosarcomatous transformation of DFSP of right cheek and retroauricular region. On IHC it showed expression of vimentin and CD 34. Various literature documents that progression of DFSP to FS-DFSP shows tumour progression or "dedifferentiation" along with increased risk of

metastasis, because of gains of p53 mutations and increased proliferation.[7,13]

There are two surgical modalities for DFSP; one is conventional WLE and another Moh's micrographic surgery (MMS). NCCN and European interdisciplinary group suggested that safe oncological margin of 2-4 cm as excisional criteria.[4,6,14] Low rate of recurrence is seen with proper safe margins of more than 2 cm; on contrary 60% recurrence is observed when there is improper excision due to irregular margins or tumour near vital structures.[4,15] MMS has gained acceptance in treatment of DFSP due to its reduce recurrence rate than WLE. It shows advantage of preservation of tissue over WLE. Main disadvantage of it remains longer surgical time, its complexity for larger lesions and precise need for frozen section. Review of 23 systemic non-randomized trials were conducted which stated that recurrence rate with MMS in DFSP was 1.11% and 6.32% with WLE.[16]

Radiotherapy (RT) is adjuvant treatment used in cases with close or positive margins as modality for local control where re-excision is not possible. Various literatures suggested that adjuvant RT post operatively reduces recurrence rate. In a retrospective study of 184 cases stated significant disease-free survival of 5 years in cases receiving adjuvant RT in comparison with cases that did not underwent RT (88.1% vs. 56.2%, p=0.044).[17] In cases of DFSP where disease is inoperable or there is metastasis use of Imatinib as chemotherapeutic drug as standard of care of treatment.[4] In a systemic review it was seen that when neoadjuvant imatinib was used in conjunction with surgery showed superior result for removal of tumours with negative margins.[4,18]

We performed wide local excision of lesion keeping oncologically safe margins in consideration and followed by adjuvant therapy because of extensiveness of tumour. Routine follow-up was carried every month for first six months then every three months to ensure disease free survival of patient.

CONCLUSION

Fibrosarcomatous transformation in DFSP is rare in head and neck region due to its indolent nature. Overall survival rate is excellent but is equally challenging considering size and location of tumour. Mainstay treatment remains wide local excision of tumour with margins of 2-4 cm as it reduces recurrence rate; however, this should be considered if appropriate reconstructive options. Adjuvant therapy with radiotherapy and imatinib should be considered in non resectable or extensive lesions and in palliative set up. Long-term follow up is essential in all patients.

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