

Case Report

Dermatofibrosarcoma protuberance of the oral cavity: Does it exist?

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Abstract

Dermatofibrosarcoma protuberance (DFSP) is a low grade spindle cell malignant tumour that is locally aggressive especially if incompletely excised.

A 64-year-old man presented with intra-oral buccal mass of 34 year duration with accelerated increase in size in the last two years. CT scan showed well-circumscribed tumour with no relation to the overlying skin. Fine needle aspiration cytology revealed a highly cellular mitotically active spindle cell neoplasm with recommendation of excision with safety margins. Histological examination of the excised mass showed typical dermatofibrosarcoma protuberance with cytoplasmic positivity for Vimentin and CD34.

Intra-oral sarcomas are rare and to the best of our knowledge only a single case of DFSP has been described in literature. We present another case for its unusual site, presentation, duration and surgical approach.

Introduction

Dermatofibrosarcoma protuberance (DFSP) is a low grade malignant dermal tumour that is locally aggressive especially if incompletely excised. It usually affects middle-aged individuals with the preferential location being the trunk and lower extremities and it clinically appears as a plaque like or nodular growth with extremely infiltrative and locally destructive growth pattern.

The most frequent presenting location of DFSP is trunk with 42-72% of cases and involves proximal extremities in 16-30% of cases and head and neck region in 10-16% of cases. DFSP of the head and neck accounts for 7% of all head and neck sarcomas¹⁻⁴ Intra-oral sarcomas are rare and to the best of our knowledge only a single case of DFSP has been described to occur in the oral cavity.⁹

We present this case for its unusual site, presentation, duration and surgical approach.

Case Report

A 64-year-old man presented with intra-oral buccal mass of 34 year duration with accelerated increase in size in the last two years. The mass was located in the left cheek bulging beneath the skin (Figure-1A and 1C). CT scan showed a well-demarcated mass extending from the facial muscles and raising

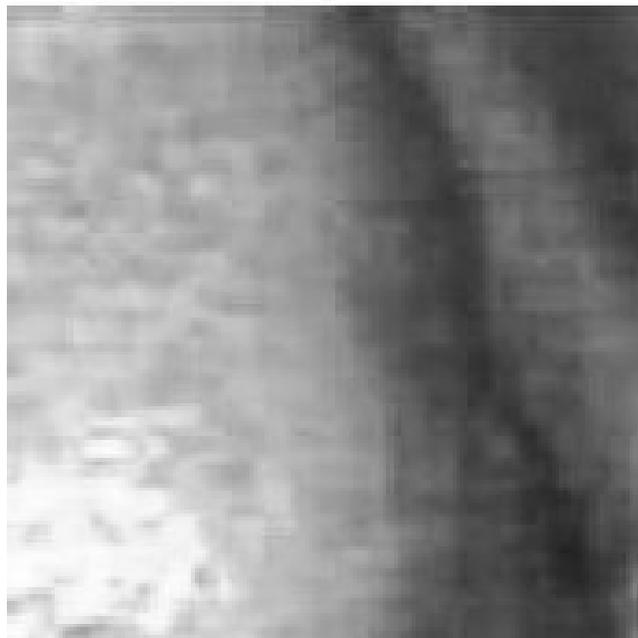


Figure-1: a) Clinical appearance of the patient with left cheek bulge. b) CT scan showing well-demarcated mass extending from the facial muscles and raising the skin without signs of infiltration to the underlying buccal mucosa or the overlying skin. c) The tumour is located in the left cheek mucosa bulging into the oral cavity. d) Gross appearance of the tumour.

the skin without signs of infiltration to the underlying buccal mucosa or the overlying skin (Figure-1B).

Fine needle aspiration cytology was performed percutaneously from the left cheek by the pathologist using 25 x 7/8 needle without negative pressure. Specimen adequacy was assessed using the diff quick stain. The rest of the smears were stained with diff quick, haematoxylin, eosin and papanicolau stain. The material was not sufficient for cell block preparation. Fine needle aspiration [FNA] showed cellular smears formed of uniform spindle cells that were of intermediate size (Figure-2A). The cells were arranged in sheets and individually with crushing artifact. The nuclei were hyperchromatic with increased N/C ratio. No mitoses or necrosis was seen. It was reported as spindle cell neoplasm with cytological atypia with recommendation of excision with safety margins.

The tumour was enucleated under local anesthesia. Grossly the mass was rounded, well circumscribed and firm measuring 3.5 x 3.0 x 2.0 cm (Figure-1D). Three micrometer-thick sections were cut from the paraffin block and stained with haematoxylin and eosin. Microscopically, the tumour was solid composed of densely packed monomorphic plump spindle cell arranged in a storiform pattern (Figure-2B and C). The spindle cells infiltrated the surrounding adipose tissue and skeletal muscle. They had large vesicular nuclei and conspicuous nucleoli. Scattered mitotic figures with occasional atypical forms were seen (5 mitoses per 10 high power field [HPF]). No necrosis was seen. The tumour extended to the margins of excision.

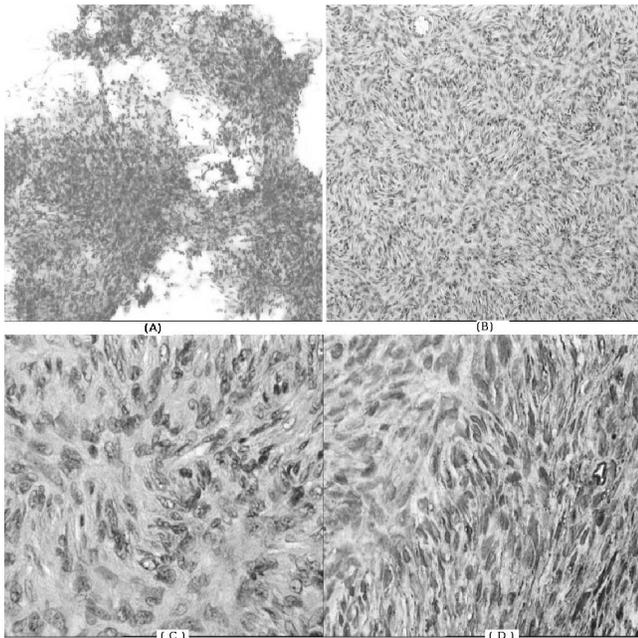


Figure-2: a) FNA showing sheets of spindle cells with nuclear pleomorphism. b) Figure demonstrates spindle cell neoplasm exhibiting prominent storiform pattern (H&E x 200). c) Higher magnification showing pleomorphism and hyperchromasia of nuclei.(H & E x 400) d) CD 34 staining the cytoplasmic membrane.

Immunohistochemical studies were performed using the standard LSAB technique and the microwave heating antigen retrieval method. Immunohistochemical studies showed positive staining of the tumour cells for Vimentin and CD34 (Figure-2D). Staining for S100, Desmin, Actin, Cytokeratin AE1/AE3, CD68, CD31, Factor VIII and HMB45 was negative. The histological and immunohistochemical features were characteristic of DFSP.

Discussion

DFSP typically presents as a nodular cutaneous mass found most frequently on the trunk and proximal extremities. It is a superficial sarcoma of fibroblastic/ myofibroblastic differentiation of the dermis and underlying soft tissue with a high local recurrence rate and little metastatic potential. It predominantly occurs in adults between 20 and 40 years and has no sex predilection. Congenital and childhood DFSP are also reported. DFSP of the head and neck accounts for 7% of all head and neck sarcomas.¹⁻³ 10-15% of DFSP cases involve the head and neck. Five cases have been reported in the face.⁴ Among the other unusual locations are upper lip, parotid, scalp, breast and vulva.⁵⁻⁸

Intraoral sarcoma is rare and only a single case has been described to occur in the oral cavity.⁹ DFSP has a characteristic storiform pattern with infiltrative growth pattern. Differential diagnosis of the spindle cell lesions in the oral cavity includes benign and malignant fibrous histiocytoma, demoplastic melanoma, schwannoma, leiomyosarcoma, kaposi's sarcoma and haemangiopericytoma. Immunohistochemical studies are helpful in this regard; CD34 is a good marker and is consistently positive DFSP and the recurrent tumours retain the positivity even in the fibrosarcomatous areas.¹⁰⁻¹² Incomplete excision increases the risk of recurrence. Post operative radiotherapy was adopted in some cases.

Fine needle aspiration cytology is a good tool to achieve diagnosis or direct therapy. It is being used increasingly in soft tissue tumours with little or no negligible risk of needle tract seeding. Immunohistochemistry is very useful in excluding other possible differential diagnosis and confirming the final diagnosis.^{10,11} The cell of origin of DFSP is debatable. It was thought to be of fibrohistiocytic origin based on the morphologic similarity to benign and malignant fibrohistiocytoma, however, some ultrastructural features suggest neural differentiation similar to Schwann cells. This is further supported by the positivity for CD34 which is expressed in the endoneurium and some nerve sheath tumours.

In this case, it was debatable whether to consider this case as a soft tissue DFSP of head and neck or as oral cavity DFSP which is an atypical location that created difficulty in the original diagnosis. Combining the radiologic, intra-operative and pathologic findings, we regarded it as intra-oral DFSP. The

duration of this lesion was 34 years with doubling of size in the last two years which is a long history compared to usual dermal DFSP. This could be attributed to the location of being intraoral while dermal DFSP attracts patient attention earlier.

Recurrence rates of head and neck DFSP is highest of any body site ranging from 50%-75% because of the difficulty of achieving wide margins. The high recurrence rate of DFSP is attributed to the infiltrative growth pattern in the tissue that clinically appears normal. Primary treatment of DFSP is complete surgical resection but the occult spread of tentacle like projections of tumour cells makes the complete removal difficult. The most important prognostic indicator is the extent of surgical resection. The aim of surgical excision in these tumours is total excision with at least 3 cm safety margin. Tumour size and location dictate the most appropriate surgical procedure. Generous margins are not possible when the tumour involves distal extremities and the head and neck region specially face where it is located near critical facial structures. This was not possible in our case due to the location, cosmetic and functional reasons and the patient's general condition. Alternative to wide resections, Mohs micrographic surgery with precise histological resection margin control is propagated to be a good surgical method in the treatment of DFSP.¹³

DFSP is a radiosensitive tumour. Post operative radiotherapy has reduced the risk of local recurrence in some cases.¹³ No radiation was given to our patient in the other institution that he is following till present. The role of chemotherapy in the management of DFSP is not established.

In more than 90% of DFSP a specific chromosomal aberration involving chromosomes 17 and 22 is described which leads to activation of platelet derived growth factor receptor (PDGFR) followed by continuous stimulation of

tumour cell growth. The use of targeted inhibitors of PDGFR is a good therapeutic option.¹³ Few studies, have shown the role of Imatinib in unresectable, metastatic or recurrent DFSP.

DFSP is a dermal tumour that has never been described in areas away from dermis including the oral cavity. This case further supports that the progenitor cell of DFSP is not present only in the skin but also in the submucosal tissue of the oral cavity.⁹ Our patient was kept on close clinical and radiologic follow-up for local recurrence. The surgical approach was challenging to the surgeon who removed the tumour under local anaesthesia through oral route.

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