

Surgical Management Of A Dermatofibrosarcoma Protuberance Of The Face

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Abstract

Background: Dermatofibrosarcoma Protuberance (DFSP) is a rare cutaneous tumor of unknown etiology. The lesion frequently develops on the extremities (mostly the lower legs) trunk, and rarely above neck. It is usually asymptomatic, although pruritus and tenderness are not uncommon. DFSP is a cutaneous malignancy that arises from the dermis and invades deeper subcutaneous tissue (eg, fat, fascia, muscle, bone).

Two cases of dermatofibroma of the face presented to us of which is a pigmented variety also called bednar [6] . WE report these cases as these tumors are rare and though an entity known for about 9 decades still much research is to be done regarding the management.

Materials & Methods : Two cases of Dermatofibrosarcoma Protuberance of face of which one was pigmented variety, presented to Plastic Surgery OPD between July 2003 to December 2005.

These cases were managed by wide excision and reconstruction by local rotational flaps to achieve cosmetically good results.

INTRODUCTION

Primary mesenchymal tumours with localization outside the skeleton, parenchymatous organs or hollow viscera are generally referred to as soft tissue sarcomas. These are slow growing tumors Dermatofibrosarcoma protuberans (DFSP) is a relatively unusual, highly invasive locally aggressive cutaneous tumour of intermediate malignancy.

Usually this is reported to occur in limbs and trunks, incidence on face is very rare. Moreover the pigmented variety is still a rarer entity. [7]

The age of patients vary between second to forth decade, but may present as late as 80 years. These tumors are locally invasive, but recurrence rate is found to be very high.

Surgery forms the mainstay of management, though radiotherapy has got some role incases of involved margins. Chemotherapy has no role in management.

HISTORY

Two female patients with exfoliative growth at face presented to us between July 2003 to December 2005.

On histopatholgy these lesions came out to be pigmented

dermatofibrosarcoma protuberance a locally invasive malignant tumor which was cosmetically unacceptable in these patients.

CASE 1 Mrs ABC a 60 years old female presented to us with a huge mass of size of 13 cm. x 9 cm arising from the bridge of nose and inner canthus of the left eye. It was covering both the eyes as a curtain. It was pink colored with areas of necrosis. No cervical lymph nodes were palpable. Histopathology revealed pigmented dermatofibrosarcoma protuberance. [Photo 1]

Figure 1

Photo 1: DFSP arising from nose pink colored



Figure 3

Photo 3: post operative



CASE 2 Mrs xyz a 56 year old female presented to us with a mass arising from rt upper eyelid and supraorbital region. The size of mass was 10cm. x 11cm. Her eyesight of rt eye was lost with complete destruction of eyelid. It was skin colored. No cervical lymph nodes were palpable. Histopathology revealed dermatofibrosarcoma protuberance. [Photo2]

Figure 2

Photo 2: PDFP arising from eyelids



Figure 4

Photo 4: post operative result (right eye had to be sacrificed)



Tissue histopathology was done and margins were free of tumor.

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) is a rare variety of mesenchymal tumour arising from dermis. It is locally aggressive and highly recurrent malignant neoplasm.

DFSP was first described by Darier and Ferrand as a distinct clinical entity in 1924, it was Hoffmann who first coined the term DFSP. [1] Other terms used to describe this neoplasm are hypertrophic morphea, progressive and recurrent dermatofibroma, fibrosarcoma of skin and sarcomatous tumour resembling keloid.

Pathophysiology: The precise mechanism for the

development of DF is unknown. Cultured DFSP tumor cells have increased growth response to platelet-derived growth factor BB (PDGF-BB). Cytogenetic studies may reveal specific lesions in DFSP tumor cells, such as reciprocal translocations of chromosomes 17 and 22, t(17;22), and supernumerary ring chromosomes composed of interspersed sequences from chromosomes 17 and 22. These rearrangements fuse the platelet-derived growth factor B-chain (PDGFB, c-sis proto-oncogene) and the collagen type I alpha 1 (COL1A1) genes. PDGFB and COL1A1 fusion noted in DFSP may contribute to tumor development through the ectopic production of PDGF-BB and the formation of an autocrine loop.

DFSP usually presents as a large indurated plaque several centimeters in diameter. It is composed of firm, irregular nodules varying in color from flesh to reddish brown. Sometimes, it may present as a morphea-like, atrophic, sclerotic, violaceous plaque without nodularity that may ulcerate as it slowly increases in size .[2]

Frequency: In the US: DFSP constitutes less than 0.1% of all malignant neoplasms and approximately 1% of all soft tissue sarcomas. The incidence has been estimated to be 0.8-5 cases per 1 million population per year in 2 separate studies. It occurs mostly on trunks extremities. The incidence on face is still rarer.[3,7]

HISTOLOGIC FINDINGS

In the plaque type of DFSP, slender tumor cells with large, spindle-shaped nuclei are embedded fairly uniformly in the collagen stroma, parallel to the skin surface. Mitotic figures are sparse. The more characteristic findings are seen in the nodular type. These findings include the high cellularity and irregular, short, intersecting bands of tumor cells forming a storiform pattern. Also typical are cells radiating from a central hub of fibrous tissue forming a cartwheel pattern. The degree of cellular atypia is higher in nodular lesions than in plaque lesions. Occasionally, DFSP may show focal fibrosarcomatous changes with a characteristic herringbone pattern. The cellular atypia is then even more prominent with hyperchromatic nuclei and more mitotic figures.[2]

Investigations : No imaging study is required . Only skin biopsy is required .

Medical Care: no medical treatment is there apart from some response to radiotherapy specially when margins are positive.

Surgical Care : Surgery forms the mainstay of treatment. Excision with a margin of 3 – 5 mm is necessary. But at cosmetically important areas as face reconstruction becomes a seemingly big problem. In our series both the patients were managed by mobilizing flaps from peripheries after resection and cosmetically acceptable results were achieved.[4,5] One of the surgical techniques used to prevent recurrence is Mohs method. The Mohs micrographic surgery is an excisional technique with complete histological margins' control. This technique allows the complete removal of the neoplasm, examining all its margins, while conserving as much uninvolved tissue as possible.

Radiotherapy has a limited role; it may however be useful, combined with surgery when margins are positive or even alone in cases of non-operable tumours. Chemotherapy is not indicated being reserved for metastatic disease

Ours case was managed by wide local excision with a single stage reconstruction using local rotational flaps, and has yielded good results so far with no recurrences.

Follow up Since it is a locally invasive tumor regular close follow up is necessary. In our series both the patients were initially assessed 3 monthly for 2 year. Now they are on annual follow up with no evidence of recurrence.[2]

Thus we can conclude that though a rare and locally invasive entity, these lesions, by knowledge of the condition, its clinical appearance, course and histopathology can be dealt with by optimized management strategies.

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