A Rare Case of “Facial Dermatofibrosarcoma Protuberance”

Ashutosh G Pusalkar*, Haritosh Velankar**, Sharad Bhalekar***, Yogesh Dabholkar+, Jyoti Singh**

Abstract
Dermato-fibrosarcoma protuberance (DFSP) is a relatively uncommon soft tissue neoplasm with intermediate-to-low grade malignancy. It is a very slow growing tumour. Although metastasis rarely occurs, DFSP is a locally aggressive tumour with a high recurrence rate. It commonly occurs on the trunk and extremities but rarely above the neck. Surgical wide excision remains the mainstay of treatment for DFSP. We report a case of Facial Dermato-fibrosarcoma protuberance.

Introduction
DFSP have been reported in the literature as early as 1890, Darier and Ferrand first described it in 1924 as a distinct cutaneous disease entity called progressive and recurring dermato-fibrosarcoma.1 Hoffman officially coined the term dermato-fibrosarcoma protuberance in 1925. DFSP is a cutaneous malignancy that arises from the dermis and invades deeper subcutaneous tissue (e.g. fat, fascia, muscle, bone). The cellular origin of DFSP is not clear at this time. Evidence exists that supports the cellular origin being fibroblastic, histiocytic, or neuroectodermal. It usually presents as a large indurated plaque composed of firm, irregular nodules varying in colour from flesh to reddish brown. It commonly occurs on the trunk, extremities but rarely above the neck.2 Currently the cause of DFSP is unknown. Laboratory studies have shown that chromosomal aberrations may contribute to the pathogenesis of DFSP; however no evidence of hereditary or familial predisposition exists.3 In 10-20% of patients with this tumour, trauma at the site seems to be incriminated. Surgical and old burn scars and sites of vaccinations have all been reported as sites of DFSP.

We report a rare case of facial dermatofibrosarcoma protuberans

Case Report
A 72 year female presented to our hospital with a huge 10 cm × 10 cm × 8 cm painless swelling protruding from forehead (Fig. 1). Patient was apparently alright 30 years back when she developed a small papule on forehead which started growing slowly to the present size obscuring nasal field of vision. On examination it was a firm non-tender well defined swelling with 4-5 satellite small lesions around it. FNAC was suggestive of vascular tumour because of engorged superficial veins. Biopsy was taken from the lesion which was conclusive of dermato-fibrosarcoma. C.T. Scan (P+C) was suggestive of a heterogeneously enhancing lesion arising from forehead skin (Fig. 2) involving glabella, nasal bone and frontalis muscle. Patient was posted for excision of the swelling under general anaesthesia. Although the swelling was obscuring anaesthetist's vision patient could be intubated. The swelling was excised with safe 3 cm margins all around. Primary wound closure was possible hence it was done (Fig. 3). Histopathology report showed slender tumour cells with large, spindle shaped nuclei are embedded in

*Professor Emeritus; **Professor and Head; ***Assistant Professor, +Associate Professor; Department of ENT, Padamshree Dr D.Y.Patil Medical College, Sector-7, Nerul, Navi Mumbai - 400 706.
Fig. 1: A patient with swelling protruding from forehead.

Fig. 2: CT scan showing lesion arising from forehead skin.

Fig. 3: After operation.

The collagen stroma, parallel to the skin surface. These findings included the high cellularity and irregular, short, intersecting bands of tumour cells forming a storiform and cartwheel pattern. Immunohistochemistry showed moderate to strong staining of human progenitor cell antigen CD-34 in tumour cells confirming Dermato-fibrosarcoma protuberance.

Discussion

DFSP accounts for less than 0.1% of all malignant neoplasms and approximately 1% of all soft tissue sarcomas. The annual incidence of DFSP is reported as low as 3 cases per million populations. It is characterized by its aggressive local invasion. The tumour invades local tissue by extending tentacle like projections underneath healthy skin rendering complete.

Removal of the tumour very difficult. Incomplete removal of these neoplastic cells results in a high local recurrence rate. Despite the local invasiveness, it rarely metastasizes. A wide surgical excision with adequate margins is treatment of choice. Postoperative radiotherapy can be considered as an adjuvant therapy, if the margins are not clear. Recently, Mohs micrographic Surgery is considered better surgical option. Although medical therapy is not a first line treatment for localised DFSP, newly approved molecular targeted drug, imatinib mesylate, is an effective oral medication for unresectable, recurrent and metastatic DFSP.

Conclusion

Dermato-fibrosarcoma protuberance is a rare soft tissue tumour arising from skin which is locally invasive but rarely metastasizes with a high rate of recurrence. A wide surgical excision is a must to avoid local recurrence.
Diagnosis is by histopathology of the biopsy (wedge or excisional) and immuno-histochemistry. Facial DFSP does occur very rarely.

References

RISK OF COMBINING PPIS WITH THIENOPYRIDINES: FACT OR FICTION?
During the past decade, the thienopyridine clopidogrel plus aspirin has become a cornerstone of medical treatment for patients with coronary artery disease who present with acute coronary syndromes or are having a percutaneous coronary intervention.
Prasugrel, a new thienopyridine agent that has recently been licensed, is more rapidly converted into its active compound and has significantly greater antiplatelet action than does clopidogrel. The need for in-vivo bioactivation, however, makes thienopyridines vulnerable to drug-drug interactions with other commonly coadministered drugs, such as proton-pump inhibitors (PPIs), statins, or calcium-channel antagonists, which also use the hepatic CYP system for metabolism.
Patients receiving PPIs are more likely to discontinue antiplatelet treatment because of recurring symptoms that could be linked to such antiplatelet treatment.
So is the interaction of PPIs with thienopyridines fact or fiction? This interaction is a fact in terms of pharmacodynamics. If absolutely needed, specific PPIs that are less likely to interfere with the platelet response to thienopyridines could be given to these patients. In all cases, careful monitoring of patients’ compliance with thienopyridine drugs is mandatory.