

## A CASE OF INTERDIGITAL DERMATOFIBROSARCOMA PROTUBERANS

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### ABSTRACT

A 61 year old male reported to the surgery OPD with a slow growing tumour between the third and the fourth fingers of the left hand. The tumour was excised and on histopathological examination, the diagnosis was that of dermatofibrosarcoma protuberans.

*Key Words: Dermatofibrosarcoma Protuberans, Interdigital, Hand.*

### INTRODUCTION

Dermatofibrosarcoma Protuberans (DFSP) is an intermediate grade sarcoma. It arises from the fibroblast cells of the dermis. It is an uncommon tumour with a minimal male predilection. Early to middle aged population is more commonly involved. It is rare in children. A history of trauma preceding the tumour may be present in a few cases.

the left hand for thirty five years. There was no history of antecedent trauma.

### CASE REPORT

A 61 year old farmer presented to the surgery outpatient department with a slow-growing painless lump between the third and the fourth fingers of

Fig. 1

*Fig. 1: Preoperative picture of the interdigital tumour.*

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Fig. 2

**Fig. 2: Picture after removal of the tumour. A corrugated rubber sheet drain had been placed in situ.**

On examination, the patient had a nontender, firm, round swelling about 5.5 cm in diameter between the third and the fourth fingers of the left hand. The surface was smooth and edges distinct. The swelling was nonreducible, noncompressible and nonpulsatile. There was no sign of inflammation over or near the swelling. Transillumination test was negative. The skin above the swelling was normal but the swelling was fixed to the overlying skin. The swelling was not fixed to the deeper structures. The adjacent bones and joints were normal. Regional lymph nodes were normal. A diagnosis of implantation dermoid was made.

Apart from this swelling, the patient had a small tumour of 1 cm diameter over the left cheek. Clinically, the tumour was diagnosed to be a lipoma.

Routine investigations and chest X-Ray were within normal limits. FNAC was inconclusive.

Excision of the swellings were done under local anaesthesia. Multiple H & E stained sections of the swelling from the left hand revealed that the tumour comprised cell population predominantly of spindle cells with eosinophilic cytoplasm arranged in thick, short cellular bundles mostly towards the periphery of the tumour. Central areas showed less cellularity and bundles with abundant collagenisation. Characteristic

Fig. 3

**Fig. 3: Photomicrograph showing spindle cell fascicles exhibiting storiform pattern (arrows). There is nuclear atypia, occasional mitotic figure (low power view).**

storiform cartwheel pattern was obvious and frequent in some parts. Nuclei were elongated mostly at periphery with atypia. Central areas showed nuclear polymorphism, nuclear hyperchromasia, focal atypia and mitotic figures. Focal herringbone pattern and nodular arrangement was also seen. Giant cells were quite a few throughout, more so central.

Focally also seen were epithelioid, predominant myxoid and osteoclast like round to oval cell areas. The impression was of Dermatofibrosarcome protuberans.

The tumour from the cheek was diagnosed to be Angiofibroma on histopathological examination.

## DISCUSSION

Dermatofibrosarcome protuberans (DFSP) initially presents as a solitary asymptomatic firm tumour. It is usually found on the trunk and proximal external extremities and occasionally only on the head and neck.<sup>1</sup> The tumour is slow growing and may develop small nodules over it. The tumour is usually fixed to the overlying skin but not to the underlying structures. The tumour can be confused with scar, lipoma, neurofibroma or cyst.

The tumour is locally aggressive but can rarely spread<sup>2</sup> to regional nodes and/or internal organs. A narrow margin of resection can lead to recurrences. After multiple recurrences, the underlying fascia, muscle, bones and even brain can get invaded. Patients with metastases usually die within 2 years.<sup>3</sup>

Primary or recurrent DFSP is best treated by surgical excision. The recommended margin of surrounding skin and underlying fascia is 3 cm. Radiotherapy has been used in a few cases.

Microscopically, the appearance of radial whorls of spindle cells producing the storiform or cartwheel appearance is characteristic but not pathognomonic. A monomorphic appearance is usual. High cellularity, moderate to light mitotic activity, paucity of haemosiderin loaded macrophages and entrapped fat cells may be found when the subcutis is invaded. Myxoid features may be found. Areas of atrophy and regression may also be found. Granular cell change may also be found.

Immunohistochemically, DFSP is positive for vimentin, actin (inconstantly and focally) and CD 34 (strongly and constantly),<sup>4</sup> thus raising the possibility of it being a peculiar nerve sheath tumour (but made of cells other than schwann cells or perineural cells.) They are negative for s-100 protein, HMB-45, keratin and FXIII antigen.<sup>6,7,8,9</sup>

Primary care doctors should suspect a dermatofibrosarcoma if they see a slow growing dermal swelling protruding like a cauliflower present for a long time. Such patients should be referred to a dermatologist or surgeon.

## REFERENCES

1. Beauchamp Evers Mattox . Sabiston Textbook of Surgery, Book 1. 2000: 505.
2. A dams JT, Saltzsteims L : Metastasizing dermatofibrosarcoma protuberans. A merican Journal of Dermatopathology 1993, 15: 429-434.
3. Rosenberg SA , Y ang JC, White DE, Steinberg SM : Durability of complete responses in patients with metastatic cancer, treated with high - dose interleukin-2 : I dentification of the antigens mediating response. A nn Surg 1998, 228 : 307-319.
4. Dominiques Malagon HR, Ordonez NG, Mackay B : Dermatofibrosarcoma protuberans. U ltrasound and I mmunocytochemical observations. U ltrastructural Pathol. 1995, 19 : 281 - 290.
5. Weiss S, Nickoloff B : CD 34 is expressed by a distinctive cell population in peripheral nerve, nerve sheath tumors and related lesions. A m J Surg. Pathol. 1995, 17 : 1039 - 1045.
6. A benaza P, L illemoe T : CD 34 and factor X I I I a in the differential diagnosis of dermatofibroma and dermatofibrosarcoma protuberans . A m J Dermatopathol. 1993, 15 : 429-434.
7. Kutzner H : Expression of the human progenitor cell antigen CD34 (HDL A -1) distinguishes dermatofibrosarcoma protuberans from fibrous histiocytoma in formalin- fixed, paraffin- embedded tissue. J A m A cad Dermatol. 1993, 28 : 613-617.
8. Leong A SV, L im MHT : I mmunohistochemical characteristics of dermatofibrosarcoma protuberans. A ppl. I mmunohistochem. 1994, 2 : 42 - 47.
9. Ma CK, Zarbo RJ, Gown AM; I mmunohistochemical characterization of atypical fibroxanthoma and dermatofibrosarcoma protuberans. A m J Clin Pathol. 1992, 97 : 478-483.



## Marjolin's Ulcer

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### ABSTRACT

Scar malignancies are generally known as Marjolin's ulcer and majority of them are epidermoid carcinoma. A 35 years old, lady who developed Marjolin's ulcer on three years old scar of burn on the dorsum of right foot is described here.

*Key Words: Marjolin's ulcer, Thermal burns scar of foot, Squamous cell carcinoma.*

### INTRODUCTION

Marjolin's ulcer is an ulcer that arises from previously traumatized, chronically inflamed, or scarred skin. Celsus described the association of abnormal tissue growth with burn scars as early as first century, but eponyms of Marjolin's ulcer is derived from Jean Nicolas Marjolin.<sup>1,2</sup> It is originally described as degeneration of cutaneous scars<sup>1</sup> (1928). Although most of the cases of Marjolin's ulcers are associated with old burn scars, but multiple sources have been reported including venous stasis ulcer,<sup>3</sup> frost bite,<sup>3</sup> decubitus ulcer,<sup>4</sup> vaccination site,<sup>5</sup> osteomyelitis,<sup>6,7</sup> urinary fistula,<sup>8</sup> hydradenitis suppurativa<sup>9</sup> and skin graft site.<sup>10</sup>

### CASE REPORT

A 35 years old, lady from low socio-economic strata came to Nepalgunj Medical College Teaching Hospital Kohalpur, Nepal, with the complaint of non healing ulcer for the last 2 years. It was painless and appeared on the healed scar of thermal burn which occurred 3 years back on the dorsum of the right foot. The healed scar of burn was healthy for one year then a nodule appeared which was painless and ulcerated that attained to the present size (Fig.1).

On local examination, a single large ulcer on the dorsum and medial aspect of the right foot

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Fig. 1

**Fig.1. Shows Majolin's Ulcer of right foot**

measuring 15 cm in the longest diameter. It was extending from medial malleolus to the medial two toes. Underlying bone was not exposed. The ulcer had everted edges and the base was indurated. The skin near by wound was erythematous with significant soft tissue oedema. A cheese like material was present on the floor of the ulcer. Pulses and the finding of a neurologic examination were with normal range. No lymphadenopathy was present. Radiographs of the foot and ankle showed osteolytic lesions in calcaneum, talus, metatarsals,

lower end of the tibia and fibula (Fig.2). Chest X-ray revealed no abnormality and result of liver function test was within normal limits. Patient was admitted with the diagnosis of chronic septic wound on the right foot. The culture of the ulcer grew staphylococcus aureus, staphylococcus epidermidis and peptostreptococcus anaerobius.

Treatment with intravenous ampicillin, cloxacillin and metronidazole was started. Biopsy of the lesion showed the features of well differentiated Squamous Cell Carcinoma. Below knee amputation was done. Post-operative period was uneventful.

## DISCUSSION

Although the incidence of Marjolin's ulcer is more common in males,<sup>3,11</sup> but this patient was a 35 years old female. The interval between the burn injury and appearance of ulcer was one year which is very much less than reported in previous studies.<sup>2,3,6,12</sup> The lesion was of the lower extremity which

Fig. 2

**Fig.2. X-ray of the right foot, Showing Osteolytic lesion in Calcaneum, Talus, Metatarsal and lower end of the Tibia and Fibula**

accounts the commonest site of the tumor.<sup>2,3,13</sup> Similar to other study, the tumor presented as an ulcer with everted edges, indurated base and covered with cheesy foul smelling slough.<sup>2</sup> There was no clinically palpable inguinal lymph nodes supporting the previous theories that the lymphatics and blood vessels were destroyed during the scarring of previous burn.<sup>2,11</sup> Patient was initially managed for infection followed by confirmation of diagnosis by histopathological examination. Later on, the definitive management of below knee amputation was done.<sup>1,3,11,13</sup>

## REFERENCE

1. Marjolin JN: Dictionnaire de Medicine. A delon NP (ed). Paris, Bechet, Vol 21, 1928, pp 31-50.
2. Traves N, Pack GT: The development of cancer in burn scars. A n analysis and report of 34 cases. Surg Gynecol Obstet 1930; 51: 749-782.
3. Fleming MD, Hunt JL, Pudue GF, et al: Marjolin's ulcer: a review and reevaluation of a difficult problem. J Burn Care Rehabil 1990; 11: 460-469.
4. Berkwits L, Yarkony GM, V: Marjolin's ulcer complicating a pressure ulcer: case report and literature review. Arch Phys Med Rehabil 1986; 67: 831-833.
5. Marmelatz WL: Malignant tumour in small pox vaccination scar: a report of 24 cases Arch Dermatol 1968; 97: 400-406.
6. Bowers RP, Young JM: Carcinoma arising in scars, osteomyelitis, and fistulae. Arch Surg 1960; 80: 564-570.
7. Johnson LL, Kempson RL: Epidermoid carcinoma in chronic osteomyelitis: diagnostic problems and management. Report of 10 cases. J Bone joint Surg Am 1965; 47: 133-145.
8. Hooper AA, Ford HT: Marjolin's ulcer complicating supra pubic cystostomy. Br J Urol 1973; 45: 534-535.
9. Alexander SJ: Squamous cell carcinoma in chronic hydradenitis suppurativa. A case report. Cancer 1979; 43: 745-748.
10. Hammond JS, Thompson S, and Ward CG: Scar carcinoma arising acutely in skin graft donor site. J Trauma 1979; 27: 681-683.
11. Barr LH, Menard JW: Marjolin's ulcer. The L SU experience. Cancer 1983; 52: 173-175.
12. Ames FC, Hickey RC: Squamous cell carcinoma of the skin of the extremities. Int Adv Surg Oncol 1980; 3: 179-199.
13. Clairmont AA, Hanna DC, and Gaisford JC et al: Marjolin's ulcer. Pa med 1979; 82: 31-33.

