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"NEGLECTED GIANT DERMATOFIBROSARCOMA PROTUBERANS"

Surgery

Dr. Divij Goel PG Resident, General Surgery, Shri Guru Ram Rai Institute Of Medical & Health Sciences Patel Nagar, Dehradun.

Dr. S. C. Sharma Professor, General Surgery, Shri Guru Ram Rai Institute Of Medical & Health Sciences, Patel Nagar, Dehradun.

ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow growing, intermediate to low grade malignant soft tissue neoplasm originating from the dermal layer of the skin. We encountered a massive DFSP over the back which presented to us with acute bleeding and pus discharge. This was successfully managed by wide local excision of the tumor. This case is remarkable due to the large size of tumor presenting unusually as acute bleeding and its successful management with wide local excision as a lifesaving procedure.

KEYWORDS

Dermatofibrosarcoma protuberans, Mohs micrographic surgery, wide local excision

INTRODUCTION:

Dermatofibrosarcoma protuberans (DFSP) is a rare slow growing [1,2], intermediate to low-grade malignant soft-tissue neoplasm originating from the dermal layer of the skin [1-3]. It is most commonly seen on the trunk followed by proximal extremities and the head and neck region (1). It has an incidence of 0.8 to 4.5 cases per million persons per year. It is characterized by the proliferation of monomorphous spindle cells arranged in a distinctive storiform pattern.

Classical DFSP often presents with a typical protuberant appearance. Its variants include fibrosarcomatous, myxoid, pigmented, giant cell, giant cell fibroblastoma, sclerotic, granular cell, and atrophic type. [3,4]

Case Report:

43/female- low body built- Biopsy proven case of Dermato fibrosarcoma Proturberans (DFSP) presented to emergency with recurrent large irregular swelling over the lower back-midline x 6 months-(15 x 15 x 12cm) progressive in size, a/w on and off fever, local pain with pus and blood discharge from it.

There are h/o two previous surgeries for the same done outside- WLE under LA- 4yrs back, WLE with STSG under SA- 1.5yrs back with h/o recurrence after both surgeries. No h/o chemoradiotherapy. No comorbidities. she was referred from community health center of a nearby subdivisional town.

Her family gave h/o presence of mass lesion over the lower back for the last 4.5 years. It was insidious in onset and gradually progressive in nature. They also revealed quack treatment initially. There was no history of trauma but there was h/o on and off bleeding with pus discharge from the tumor.

On presentation, we could find a bulky cotton wool dressing over the back which was soaked in blood. patient was conscious, alert and ambulatory. There was evident pallor and sweat over her face although she was calm. Her Pulse was 88/min, blood pressure 100/70 mmHg on the right arm –left lateral position, afebrile and breathing normally. A quick survey of rest of general physical and systemic examination was essentially normal.

She was transfused with 4unit of PRBC (3 units pre op and 1-unit intra op), i/v/o her low haemoglobin value of 5.6gm%. A formal informed written consent was taken regarding the procedure and associated recurrence risk with prognosis of the situation in general. Under GA, wide local excision was done, ensuring a margin of 3cm. Tumor was found to evade the para spinal muscles in the right lumbar region; hence part of the muscles was excised out.

Vertebrae, Ribs and left para spinal muscles were inspected thoroughly and was found to be free of any tumor deposits. Split-thickness skin graft was placed over the exposed defect so as to close the wound.

The excised specimen measured approximately 13.5 × 13.5 × 8 cm in the greatest dimensions, that was sent for HPE that confirmed the diagnosis of DFSP with negative surgical margins.

The postoperative period had been uneventful. She has been in our follow-up for 8 months now. She has maintained stable health till now with no evidence of tumor recurrence at the local site.

DISCUSSION:

DFSP is a low grade, uncommon soft-tissue sarcoma that was originally described in 1924 by Darier and Ferrand [4]. The term DFSP was coined by Hoffman in 1925[4]. It is commonly seen in persons in the third or fourth decades of life with a slight male predominance.

Congenital DFSP has been reported in literature although it is unusual in children. In one large case series, the average tumor size has been reported to be fewer than 5 cm, and there is handful of cases of tumor size more than 10 cm [6-8]. Cases of patient's neglect resulting in giant basal cell carcinoma have been reported earlier, and patient's relatives play an imperative role in the treatment process.

Regional lymph nodal and distant metastases are infrequent. A high local recurrence rate has been reported. It may be as high as 18% when wide excision margins (more than 2 cm) have been taken [7,8].

Mohs micrographic surgery has been recommended as the method to minimize local recurrence [6-8]. Biopsy of the lesion is typical. The lesion is typically located in the dermis with irregular infiltration of the subcutaneous fat in lace-like pattern. The cellular arrangement in a storiform or mat-like arrangement is typical.

The most frequent and difficult differential diagnosis is with the giant dermatofibromas (DF). Immunohistochemical analysis can be done to differentiate DFSP from DF. DFSP are CD34 positive and factor XIIIa, CD117 negative whereas DF is CD34 negative and factor XIIIa positive [9].

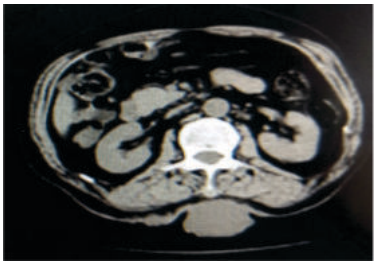
Postoperative adjuvant therapy includes Imatinib, a tyrosine kinase inhibitor, has been approved to treat adult patients with unresectable, recurrent, and/or metastatic disease. The role of radiotherapy in adjuvant settings is limited [10].

It may be of some value when the resection margins come out to be positive. We did not give any adjuvant chemotherapy and radiotherapy to the patient as the excised margins were free of tumor deposits.

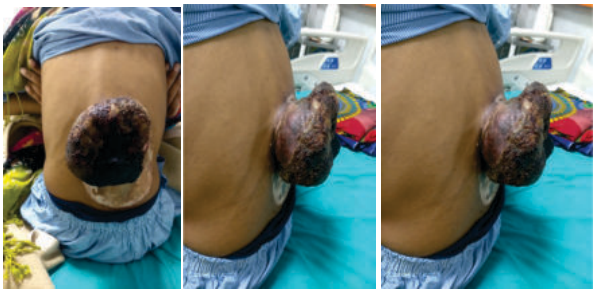
Long-term follow-up is essential as majority of the local recurrences occur within 3 years after the surgery. We intend to further follow-up, the patient for any local recurrences or distant metastasis due to our concerns of the massive size of the tumor.

CONCLUSION:

This case has been extraordinary due to several aspects. The size of the tumor was 15 x 15 x 12cm (Giant DFSP). In spite of its giant nature, there was no evidence of any distant metastasis. It presented to us with pus discharge and bleeding from the tumor. This again is an unusual presentation of DFSP. The tumor was efficiently managed with wide local excision.



MRI:



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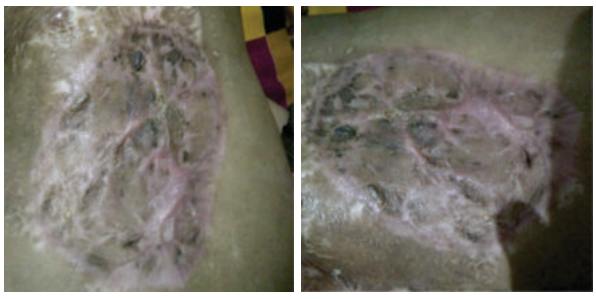
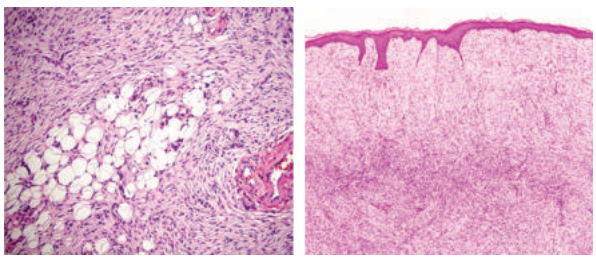


Surgical Site (after 8 Months):



HPE:

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