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A Case of Recurrent Scrotal Dermatofibrosarcoma and Review of Literature

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Abstract

60-year-old presented with swelling on the right side of scrotum extending to opposite side and to the upper thigh. It was excised in total, and the skin graft was applied to the exposed area. The biopsy of the specimen revealed it as recurrent dermatofibrosarcoma. Dermatofibrosarcoma of the scrotum is a very rare disease.

Keywords: ***Recurrent; Dermatofibrosarcoma, Excision, Skin grafting, Spindle cells***

1. Introduction

Dermatofibrosarcoma in the scrotum is a rare disease. Incomplete removal causes recurrence of the disease. We are reporting a case of recurrent scrotal dermatofibrosarcoma who underwent wide excision and split skin grafting.

2. Material and Methods

60-year-old male was admitted with history of swelling over both sides of scrotum for 8 months. Seven years back, he had similar swelling over right upper thigh, which was excised, and skin grafting was done. At that time biopsy report came as Dermatofibrosarcoma. On examination multiple swellings in the scrotum of size 14 cm × 12 cm extending from the anterior, posterior, and lateral aspect of the scrotum and involving the cord structures. Ultrasonography showed multiple hetero-echoic mass seen in both sides of scrotum. The largest measuring 30 mm × 33 mm. Total excision of scrotum with bilateral orchidectomy with split skin grafting was done (FIG. 1-3).

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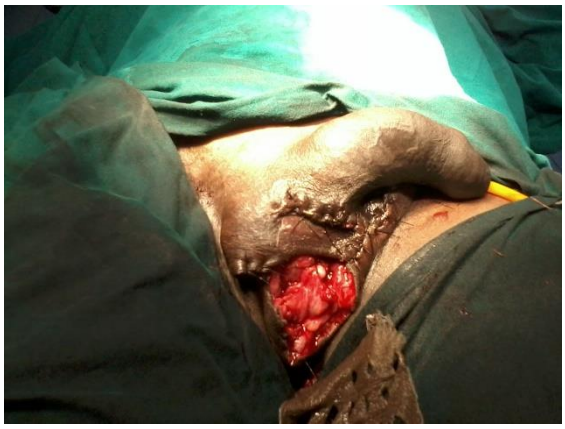

FIG. 1. **Per operative picture showing after excision of tumor.**

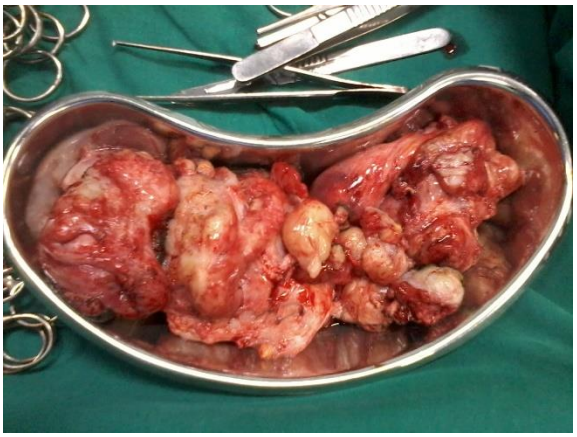

FIG. 2. **The excised specimen.**

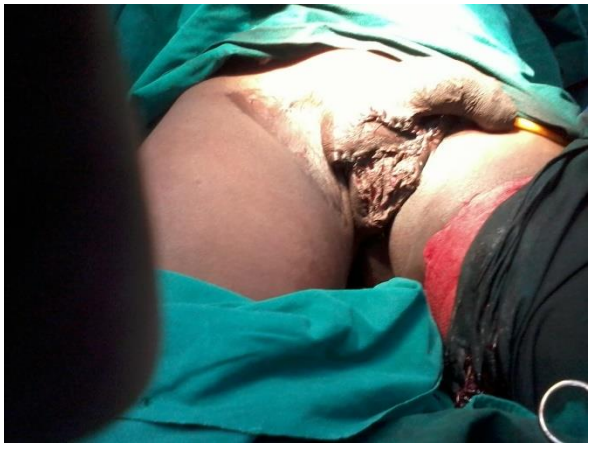

FIG. 3. **Per operative picture showing raw area covered by skin graft.**

2.1 Histopathology

The tumor is highly cellular and composed of spindle shaped cells arranged in vague storiform pattern. There are occasional normal mitoses. There are no areas of necrosis or hemorrhage or cystic change. Cellular atypia is minimal. Intermediate grade dermatofibrosarcoma protuberance (FIG. 4-6).

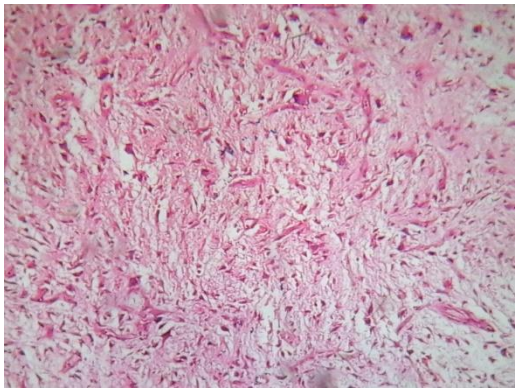

FIG. 4. **Microphotograph 1 × 40 magnification O&H stain.**

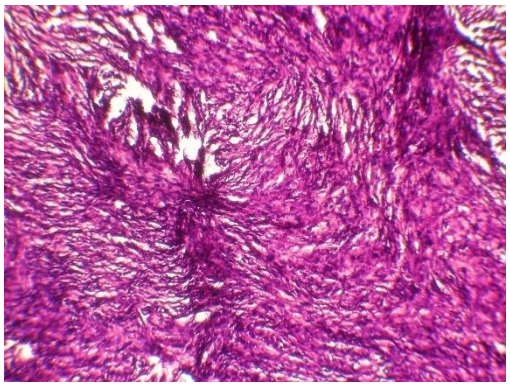

FIG. 5. **Microphotograph 1 × 40 magnification O&H stain.**

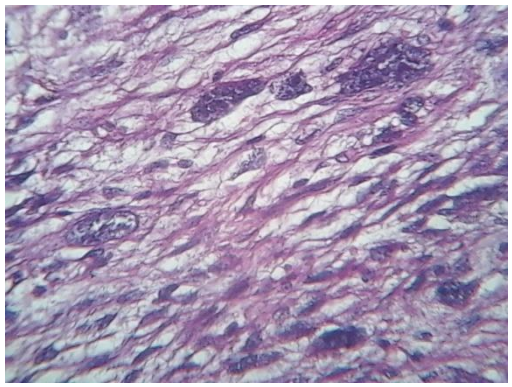

FIG. 6. **Microphotograph 1 × 100 magnification O&H stain.**

3. Discussion

DFSP is a rare malignant tumor of skin and accounts for 2 to 6% of all soft tissue sarcomas [1]. The estimated incidence of Dermatofibrosarcoma protuberans (DFSP) is 0.8 to five cases per 1 million people per year. Male: female ratio is 1.2:1. The frequent locations are trunk (45%) followed by the proximal extremities (38%) and head and neck (10%-16%) especially scalp. The occurrence of this tumor on the face is extremely rare [2]. The median age was 37 years (range, 14 to 77) [3]. Other sites are ankle, lower extremity, hip and buttocks region, trunk, retroperitoneum [4]. A DFSP lesion involving ventral aspect of glans and distal shaft of penis vulva [5,6]. Survival rates range from 91% to 100%, and local recurrence rates of 20% to 49% have been reported [7]. Dermatofibrosarcoma protuberans involving subcutaneous tissue of the breast, treated by wide excision of the tumor was reported. Ultrasound played an important role in histological diagnosis before definitive surgery and might be helpful in follow-up of patients with DFSP of the breast [8]. The expression of CD34 is a consistent finding in DFSP and sensitivity of ranges from 84% to 100% and it is extremely useful in differentiation of DFSP from benign fibrous histiocytoma [9]. Cryptogenic abnormalities of DFSP is supernumerary ring chromosomes or a unique translocation involving chromosomes 17 and 22 as in (17; 22) (22; q13) in more than 90% of cases. This abnormality results in a constitutive stimulation of the platelet-derived growth factor receptor (PDGFR) with subsequent enhancement of the tumor cell growth. Furthermore, among all the family of PDGF receptors, this translocation is responsible for the activation of PDGFR-beta [10]. Staining for factor XIIIa is positive in dermatofibroma and tends to be negative in dermatofibrosarcoma protuberans. Staining for smooth muscle actin is often focally positive in cellular dermatofibroma. By contrast, this stain is usually negative in cases of dermatofibrosarcoma protuberans [11]. The fibrosarcoma change in DFSP is associated with p53 mutations and increased proliferative activity [12]. With the standard treatment of wide local excision with 3 cm margin the local recurrence rate of 47% vs. only 7% for margins ranging from 3 cm to 5 cm. The mean time to a first local recurrence was 2.65 years. The overall recurrence rate was 16.7%. The mean time to recurrence was 38+/-12 months (range 1-100 months). In 30% of those patients with recurrences, the local regional recurrence was after 5 years. Close surveillance is necessary even beyond 5 years because late recurrences occur [13]. Girl aged 13 years, had a "dermatofibroma" excised from her left breast developed local recurrence after twenty-six years. Even after the radical operations, the patient presented a local recurrence 16 years later and patients with DFSP needs a life-long follow-up.[14] Wide resection of DFSP (whether recurrent or primary) with negative histological margins predicts a superior local recurrence-free survival [15]. Farma et al found a very low recurrence rate (1%) was achieved with relatively narrow margins (median 2 cm), allowing primary closure in 69% of patients. This approach spares the additional morbidity associated with wider resection margins and in their experience represents the treatment of choice for DFSP occurring on the trunk and extremities [16]. The risk of recurrence was 41% when the excision margin was less than 2 cm and 24% when it was equal to or higher than 2 cm [17]. DFSP is a radiosensitive tumors. Additional postoperative RT reduces the risk of local recurrence in patients with questionable or positive surgical margins. When complete surgical resection is unachievable, RT is indicated [18]. A local control probability of 82% with adjuvant or curative radiotherapy in the local management of DFSP. Radiotherapy in DFSP patients is advisable where repeated surgery may cause mutilation or functional impairment. In children, DFSP is less common and after wide excision the reconstruction with skin grafting and adjuvant postoperative radiotherapy complement each other. The former minimizes disfigurement while the latter minimizes recurrences [19]. Percutaneous cryoablation is a relatively safe and efficient technique for the treatment of local recurrence of DFSPs [20]. The patients with sarcomas arising in DFSP do not have an increased risk of distant metastasis within a 5-year follow-up period, provided they

are treated by wide local excision with negative margins [21]. Adjuvant therapy is not an alternative to wide and deep excision with adequate tumor-free margins to prevent local recurrence [22]. DFSP can transform, especially in the recurring forms, into fibro sarcomatous DFSP (FS-DFSP), a tumor with higher invasion and malignancy potential [23]. Salvage by further resection increases the risk of functional deficits and metastatic disease. As DFSP activates PDGFR targeted chemotherapy is evaluated by inhibiting PDGFR protein-tyrosine kinase. Imatinib is a selective PDGFR tyrosine kinase-inhibitor that is approved for the treatment of DFSP at a dose of 800 mg daily. Sorafenib is a small molecule B-raf and vascular endothelial growth factor (VEGF) receptor inhibitor. Sorafenib could be used for recurrence or metastasis [24]. Metastases of DFSP is seen in more than 6% of all cases, are almost invariably preceded by two or more local recurrences and occur primarily through the bloodstream to the lungs and bones and less often to regional lymph nodes [25].

4. Conclusions

DFSP is a rare tumor found commonly over the truck and extremities. But DFSP can occur in other sites also like breast, vulva and we are reporting DFSP in the scrotum for the first time involving the spermatic cord requiring bilateral orchidectomy. The recurrence is usually within 2.65 years but as in our case late recurrence can occur after 5 years. So long term follow-up is necessary. Wide margin more than 5 cm the recurrence rate is less. DFSP can turn into fibrosarcoma with early recurrence. Multimodality of treatment wide excision, post-operative radiotherapy when margin is not free. Imatinib or Sorafenib can be given for recurrence or distant metastases.

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