Scrotal Dermatofibrosarcoma Protuberance: A Case Review at Mankweng Academic Hospital

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ABSTRACT

Background: Dermatofibrosarcoma Protuberance (DFSP) is an uncommon slow-growing soft tissue tumour. The overall incidence of DFSP in the population ranges from 0.3–5 per million and is more common in blacks than whites, males and females are affected equally. The main objective of this case report is to share our experience with Dermatofibrosarcoma Protuberance and give a literature review on the standard of care.

Case Presentation: A 28-year-old male presented to the outpatient department with a five-year history of a painless right inguinoscrotal mass. On physical examination the mass was lobulated, fixed to the underlying tissue, it was non-tender measured 8 × 8 cm and there were no inguinal lymph nodes palpable. Ultrasound showed a hypervascular soft tissue tumour not involving the testicle. Wide Local Excision was performed, and histology showed a protuberant ulcerated nodule. A staging CT scan of the chest and abdomen was normal with no features of any metastatic lesions.

Conclusion: DFSP is a slow-growing and locally aggressive tumor. Therefore, early diagnosis and complete surgical resection can result in patients being cured. Mohs micrographic surgery is becoming the treatment of choice for DFSP as it results in complete surgical clearance. Imatinib is a novel treatment option for DFSP and is increasingly being used.

Keywords: Dermatofibrosarcoma, Imatinib, Mohs micrographic surgery, Scrotum.

1. Introduction

Dermatofibrosarcoma Protuberance (DFSP) is characterized as a keloid-like sarcoma an uncommon slow-growing soft tissue tumour [1]. The overall incidence of DFSP in the population ranges from 0.3–5 per million and is more common in blacks than whites, males and females are affected equally [1], [2]. The age of diagnosis is between 30- and 50 years with a peak incidence in the fourth decade of life [3]. In the literature, only 1% of DFSP cases have been reported to occur in the genital area [4]. DFSP is well documented to be locally aggressive with high rates of recurrence following resection with traditionally wide local excision. However, at present with Mohs Micrographic surgery recurrent rates are reported to be less than 5% [4].

We report a case of a 28-year-old male patient with a scrotal Dermatofibrosarcoma Protuberance presented at Mankweng Academic Hospital, Limpopo. The main objective of this case report is to share our experience with Dermatofibrosarcoma Protuberance and give a literature review on the standard of care.

2. Material & Method

Description of rare clinical cases of Scrotal Dermatofibrosarcoma Protuberance patient who presented in Mankweng Academic Hospital, Limpopo, South Africa. A written informed consent was obtained from the patient to publish this case report.

3. Case Presentation

A 28-year-old male patient with no comorbidities presented to our outpatient department with a five-year history of a painless right inguinoscrotal mass. Patient reports that the mass increased in size rapidly and became ulcerated three months prior to presentation, as illustrated in Fig. 1. On physical examination the mass was, lobulated,
and fixed to the underlying tissue, it was non-tender measured $8 \times 8$ cm and there were no inguinal lymph nodes palpable. Ultrasound showed a hypervascular soft tissue tumour not involving the testicle Figs. 2 and 3.

The patient gave the consent for Wide Local Excision and routine preoperative blood test results were unremarkable. Resected specimen (Fig. 4) was sent for histological analysis. Histology showed a protuberant ulcerated multinodular tumour. A multinodular neoplastic infiltrate comprising spindled cells was seen extending into the underlying subcutaneous fatty tissue. The cells exhibited mild to moderate nuclear pleomorphism with indistinct cytoplasmic border. The nuclei were elongated containing finely dispersed chromatin and distinct nucleoli. An average of three mitotic figures was seen per ten high-power fields which was all consistent with an Ulcerated Dermatofibrosarcoma Protuberance.

Postoperatively, the condition of the patient was uneventful. The patient was followed up with a staging CT scan. A staging CT scan of the chest and abdomen was normal with no features of metastatic lesions. Fig. 5 shows a CT scan of the chest with no metastatic lesions.

4. Discussion

DFSP occurs equally in males and females, however, some case series have found an increased incidence in males compared to females [5], [6]. The tumors occur in all ages,
but it is most frequently diagnosed between the third to fifth decade of life [3]. In our case, patient is male and detected in the third decade of life. DFSP is a slow-growing tumour that involves the dermis and subcutaneous adipose tissue. It is frequently not attached to deeper structures; however, chronic long-standing tumors may be adhered to bone and fascia [1]. In our case, tumour was extending into the underlying subcutaneous fatty tissue and no distant metastasis.

There have also been some congenital DFSP cases reported [7]. DFSP often occurs on the trunk, this accounts for 50% of all cases. The second most common sites include the upper and lower limbs accounting for 20% to 35% of the case. The head and neck only account for 10% to 15% of cases. In our case, the tumor was in the inguinoscrotal region which to the best of our knowledge is the first case in our hospital. Doufekas et al. have reported a case of DFSP involving the vulva [8]. DFSP is usually asymptomatic and a definitive diagnosis is rarely made on presentation [9] as seen in our case, the patient had this mass for 5 years which was slowly growing. The list of differential diagnoses is very broad depending on the size and site of the tumor. The differentials include leiomyoma, neurofibroma, basal cell carcinoma, keloid, desmoid tumor, Kaposi’s sarcoma, fibrosarcoma fasciitis, sarcoidosis dermatofibroma, epidermal cyst, malignant melanoma, Morphoea, and keloids [9].

The classic histological appearance of DFSP consists of dermal spindle cells that infiltrate into the subcutaneous fat tissue. The cells are monotonous, with little pleomorphism and a low mitotic count. The spindle cells form a cartwheel/storiform pattern and the infiltrating portion forms tentacle/honeycomb-like projections into the fat tissue [10]. The other types of DFSP include myxoid DFSP and Bender tumor. Myxoid DFSP is delineated by prominent myxoid stromal changes and bender tumour by the presence of dendritic cells which produce melanin [4]. The Classic Immunohistochemistry profile of DFSP consists of positivity for CD34: the tumour may be positive for EMA and negative for S100, actin and desmin [4]. In our case, the tumour was positive for both CD34 and EMA. DFSP has non-specific features in Imaging [11]. Liang Zhang et al. reported that DFSP is characterized by a subcutaneous well-defined soft tissue nodule or mass on plain CT/MR scans and shows intermediate-to-marked enhancement on contrast-enhanced CT/MR scans [12]. In our case, the patient had an Ultrasound which showed a hypervascular soft tissue tumour and a CT scan of the chest and abdomen with no metastatic lesions.

Disease recurrence after resection of DFSP remains a significant issue [13]. According to the National Comprehensive Cancer Network (NCCN) guidelines, Treatment of recommendation wide local excision with margins of between 2 to 4 cm [11]. Currently, Mohs micrographic surgery (MMS) is emerging as the treatment of choice for DFSP. MMS allows for precise tumour mapping as it involves immediate histological examination of tumour margins after excision.

DFSP is radiosensitive, most commonly radiotherapy is used in combination with surgical resection [1]. There is little role for traditional chemotherapy in DFSP [1]. Imatinib is a novel treatment for DFSP. Anne Han et al. reported that patients treated with neoadjuvant imatinib before Mohs micrographic surgery had a tumour size reduction of 36.9% and 100% local control with no recurrence at four-year follow-up [14]. Imatinib is approved in Europe for the treatment of inoperable primary tumors, locally inoperable recurrent disease, and metastatic DFSP. Imatinib has also been given to patients with extensive, difficult-to-operate tumours for preoperative reduction of tumour size [15]. The NCCN also recommends Imatinib for recurrence after resection and for cases where the tumour is nonresectable [1]. DFSP rarely metastasises, however when DFSP metastasises the common site of spread is the lungs. It has a high local recurrence rate, with 50% of cases presenting within the first year postoperatively [4].

5. Conclusions

DFSP is slow-growing and locally aggressive tumor. Therefore, early diagnosis and complete surgical resection can result in patients being cured. Diagnosis is often made histologically with the assistance of immunohistochemistry. These tumours have a high rate of recurrence which necessitates close follow-up post-excision. Mohs micrographic surgery is becoming the treatment of choice for DFSP as it results in complete surgical clearance. Imatinib is a novel treatment option for DFSP and is increasingly being used.

Author Contributions

Authors equally contributed to concept, acquisition of data, analysis of data, drafting of the manuscript and critical revision of important intellectual content.

Conflict of Interest

Authors declare that they do not have any conflict of interest.

References


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