

# A Retrospective Study of Dermatofibrosarcoma Protuberans Cases in a Nigerian Tertiary Hospital

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## ABSTRACT

**Introduction:** Dermatofibrosarcoma protuberans (DFSP) is usually regarded as a superficial low-grade sarcoma with a potential to transform into a high grade lesion. A dearth of studies solely on this tumour had been observed. It is imperative that periodic studies should be done to gather and analyse data in order to ascertain the tumour's current trajectory and, on the other hand, compare it with the available baseline data from earlier studies. To this end, this study aimed to determine the frequency, age and sex distribution of DFSP in the University of Benin Teaching Hospital, Benin City, Edo State, in the Southern part of Nigeria.

**Methods:** This was an eighteen-year retrospective descriptive study carried out to investigate the epidemiological and pathological characteristics of DFSP in this region. The data for the study was gotten from the surgical pathology register, histology request forms, duplicate copies of the histology reports and patient case notes. Histology slides were retrieved, reviewed under the light microscope and the diagnosis recorded against the corresponding patient's name on a data spread sheet. The data obtained was analysed using the Statistical Package for Social Sciences, version 16 (SPSS 16, SPSS Inc. Chicago, Illinois, United States of America).

**Results:** DFSP accounted for 5.15% of malignant skin tumours during the period under review. Compared to females, there were more male diagnosed with DFSP. It peaked in the fourth decade with an interquartile range of 26.00 to 40.75years.

**Conclusion:** This study showed that DFSP is an uncommon malignant skin tumour that is predominantly seen in males, and occurs more recurrently in the young age groups (20-39 years)

## INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a mesenchymal neoplasm of the dermis and subcutis, generally regarded as a superficial low-grade sarcoma.<sup>1</sup> It is an uncommon locally invasive fibroblastic tumour.<sup>2,3</sup>

This tumour infiltrates around the skin adnexal structures, and extends into the subcutaneous areas in a lace-like pattern.<sup>2</sup> This is associated with immense difficulty in the surgical resection of the tumour with attendant high rate of local recurrence.<sup>2,3</sup> This tumour has the potential of transforming from a low grade sarcoma to higher grade sarcoma.<sup>2</sup> The latter lesions have a potential to metastasize in 10-15 % of cases.<sup>1</sup> These features (i.e. high rate of local recurrence, transformation into a higher grade sarcoma and potential to metastasize) can contribute to the morbidity and mortality of this lesion and by extension add to ever growing huge cost of running the health care sector. Furthermore, there is a dearth of reported studies on skin tumours solely dedicated to DFSP in the South-south of Nigeria in particular and the rest of the country in general. Most of these studies were malignant skin tumours some of which are focused on specific tumours other than DFSP.<sup>4-25</sup> This study therefore, aimed to determine the frequency, age and sex distribution of DFSP in the University of Benin Teaching Hospital, Benin City, Edo State, in the Southern part of Nigeria. It is anticipated that the information gathered from this study will be useful for documentation, research, and eventually local and national planning.

## MATERIALS AND METHODS

This was a descriptive cross-sectional retrospective study. The targets of this study were all cases of dermatofibrosarcoma protuberans diagnosed histologically over a 15-year period between January 2004 and December 2018 in the Department of Anatomic Pathology, University of Benin Teaching Hospital. The Department of Anatomic Pathology receives skin biopsies from Dermatology and General Surgery Departments in University of Benin Teaching Hospital, Secondary and Primary Health Care Centres within the Benin City metropolis, neighbouring towns and villages in Edo State, as well as neighbouring states. The surgical pathology register, histology request form and duplicate copies of the histology report were useful in providing information on the age, sex, nature of specimen, hospital number, histology laboratory number, clinical presentation and clinical diagnosis of each patient/case. Histology slides were retrieved, reviewed under the light microscope and the diagnosis recorded against the corresponding patient's name on a data spread sheet. Newly prepared sections were made from the paraffin-embedded tissue blocks that were retrieved from the archives of the Department of Anatomic Pathology, University of Benin Teaching Hospital and stained with haematoxylin and eosin in cases where slides were missing or the quality of the slide had greatly diminished. The data obtained from this study were analysed using descriptive statistics. For categorical variables (sex and histologic diagnosis of dermatofibrosarcoma protuberance), the measures of frequency (i.e. count, percent and frequency) while for

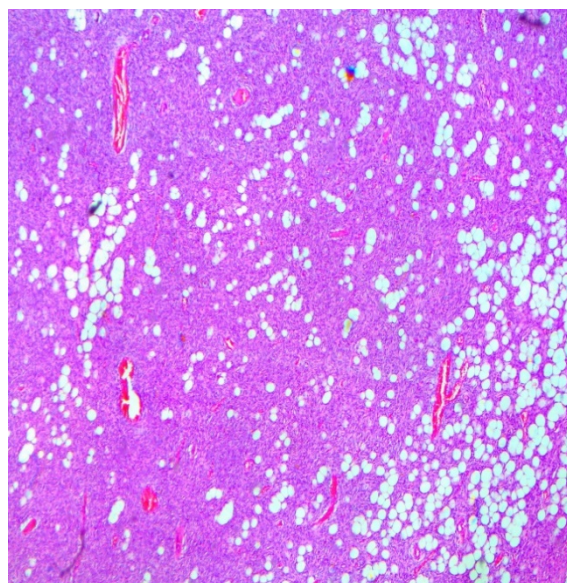
continuous variables, the measures of central tendency (i.e. median) and measures of dispersion (interquartile range) were analysed.

**RESULTS**

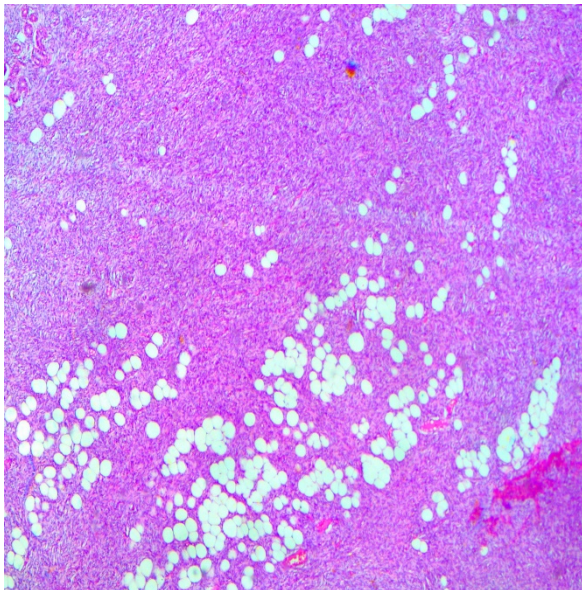
Of the 233 cases of malignant skin tumours seen during the 15 years study period, 233 cases were malignant in their course. There were 12 cases of Dermatofibrosarcoma protuberans seen during the period under review, and this represented 5.15% of malignant skin tumours. Of these, 8 cases occurred in the males while 4 cases occurred in females giving a male to female ratio of 2:1. The median age for DFSP was 34.50 years with an interquartile range of 26 to 40.75 years, and a peak in the 3<sup>rd</sup> decade. There was a bimodal peak in the 3<sup>rd</sup> and 4<sup>th</sup> decades in male while the 4<sup>th</sup> decade accounted for the peak age in the study population. Table 1 shows other age groups and their respective sex distribution of DFSP. Figures 1a-c are photomicrographs of DFSP showing spindle shaped cells with cartwheel pattern at scanning magnification, also seen are matured adipocytes encircled by the tumour cells. The lower limb was the most common site.

Table 1 Age and sex distribution of dermatofibrosarcoma protuberans

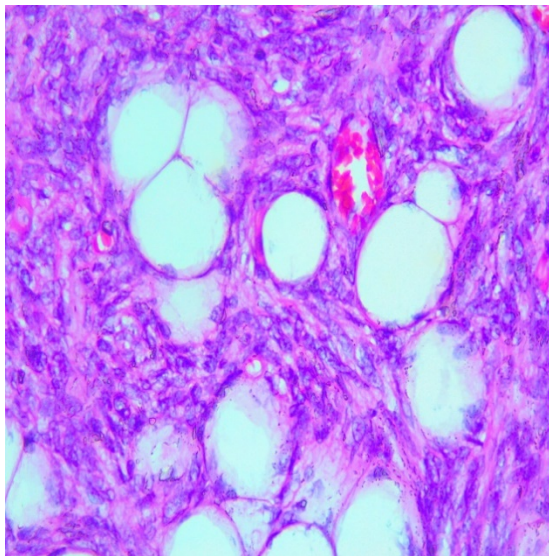
Age group	Male	Female	Total
20 – 29	3	1	4
30 – 39	3	2	5
40 – 49	1	1	2
50 – 59	1	0	1
<b>Total</b>	<b>8</b>	<b>4</b>	<b>12</b>



1a



1b



1c

**Figures 1a-c:** Dermatofibromasarcoma protuberance showing characteristically storiform pattern with deeply infiltrative proliferation of spindle shaped cells (arrow heads) entangling subcutaneous fat (squares) in a lace-like pattern. **Figures 1a-c:** (1a: H&E X 40, 1b: X 100, and 1c: X 400).

## DISCUSSION

There were 233 cases of malignant skin tumours seen during the period under review. Of these, 5.15% were DFSP. This finding is comparatively similar to the findings of previous studies done in Benin City by Forae *et al* and Azeke *et al*, where DFSP accounted for 4 % and 4.81% respectively of malignant skin tumours.<sup>4, 13</sup> This finding (5.15 %) also falls within the reference range for DFSP as reported by previous studies done in Nigeria.<sup>4, 5, 9, 13, 14, 26</sup> These studies have shown that this tumour accounts for 4 to 10.2% of all malignant skin cancers in Nigeria.<sup>4, 5, 9, 13, 14, 26</sup> Of these, the highest and lowest percentages of 10.2 and 4 percent were reported by Nggada *et al*<sup>26</sup> (Maiduguri) and Forae *et al*<sup>13</sup> (Benin City) respectively. In between these extremes, studies done by Azeke *et al*<sup>4</sup> (Benin), Gana *et al*<sup>14</sup> (Ibadan), Adeyi *et al*<sup>5</sup> (Lagos), and Asuquo *et al*<sup>9</sup> (Calabar) reported 4.81, 9.5, 8.3 and 5.8 percent respectively. Studies done in Tunisia<sup>27</sup> (Southern Tunisia) and Saudi arabia<sup>28</sup> (Quassim region) have reported lower figures of 2 % and 4.3 % respectively. All these comparative studied dermatofibrosarcoma protuberance as part of their respective studies on malignant skin tumours. However Ngadda *et al*<sup>26</sup> and the present investigation exclusively studied DFSP.

The World Health Organization (WHO)<sup>1, 29</sup> documented that dermatofibrosarcoma protuberans predominantly affects males. The finding of this study is consistent with this documentation by WHO. It is also consistent with the reports of previous studies done in Benin City,<sup>13</sup> and Maiduguri.<sup>26</sup>

In this study the peak age for the development

of dermatofibrosarcoma protuberans was seen in the third decade. Nggada *et al*<sup>26</sup> reported a bimodal peak incidence in the 4<sup>th</sup> and 6<sup>th</sup> decade, while Forae *et al*<sup>34</sup> and Adeyi *et al*<sup>32</sup> documented a peak incidence in the 3<sup>rd</sup> and 4<sup>th</sup> decades respectively. It therefore follows that this tumour was observed to peak between the third and sixth decades from studies done in Nigeria.

From the foregoing, it is concluded that DFSP accounts for approximately one-twentieth of malignant skin tumours in our environment. It is more common in males, and the young age groups (20-39 years).

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