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Dermatofibrosarcoma Protuberans: Experience with Management of Eighteen Cases

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is a cutaneous malignancy that is a variant of Soft Tissue Sarcoma (STS).

In a Nigerian tertiary health institution, 232 patients with soft tissue sarcoma were seen over a period of 22 years (1985-2006). Eighteen of these patients had DFSP. The youngest patient was 15 years and the oldest was 65 years. Majority of the patients were in the 4th decade. DFSP was found more common in the trunk followed by the upper limb.

Delayed clinical presentation is common because of the indolent nature of the tumour. Incisional biopsy is done to confirm the diagnosis.

Wide excision was found useful in most cases.

Keywords: Dermatofibrosarcoma protuberans, Nigerians, Wide excision.

Introduction

Soft Tissue Sarcoma (STS) are a heterogeneous group of rare tumours that arise predominantly from the embryonic mesoderm. They account for about 0.7% of all adult malignancies but up to 15% of childhood malignancies[1]. In Nigeria it is said to constitute about 1.3% of solid malignancies[2]. Previous reports from this hospital showed that the commonest form of STS was fibrosarcoma[3, 4].

Dermatofibrosacoma protruberans (DFSP) is a variant of STS. DFSP is a cutaneous malignancy that arises from the dermis and invades deeper subcutaneous tissue (e.g. fat, fascia, muscle, bone). The

cellular origin of DFSP is not clear at this time. Evidence exists that supports the cellular origin being fibroblastic histiocytic or neuroectodermal. It is known for its locally aggressive growth and high rate of local recurrence.

Our study investigates the age and sex distribution, clinical presentation and site at presentation in a Nigerian Teaching Hospital.

Methods

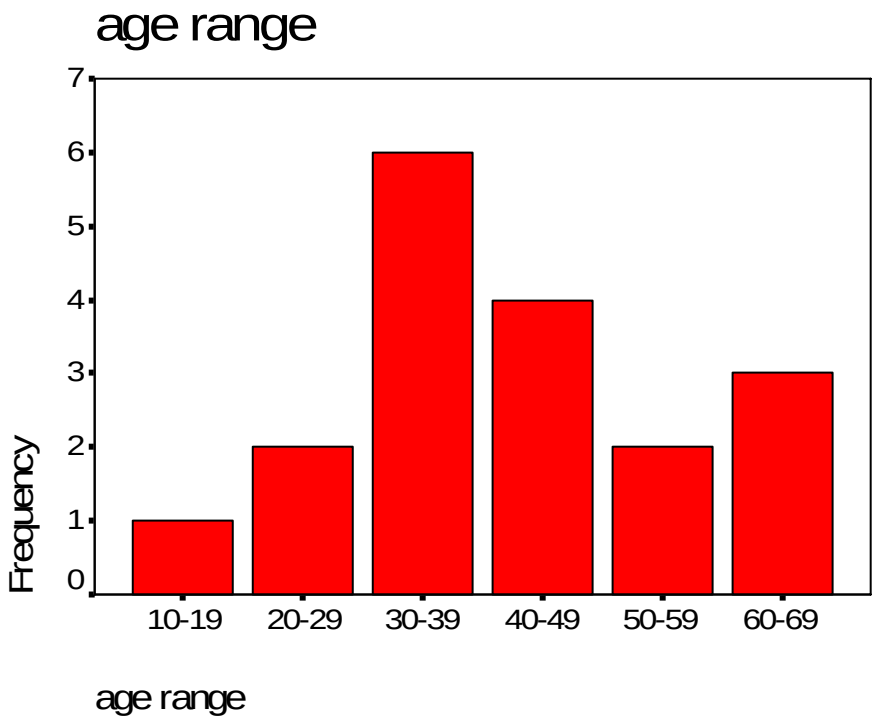
We reviewed patients with DFSP seen in a Nigerian Teaching Hospital over a period of 22 years. The hospital has 450-bed space and serves an estimated population of 10.5 million people. Information was obtained from the patients' records, operation notes and histopathology reports. The variables extracted included: age, sex, and site of Soft Tissue Sarcoma, histological variant of STS, clinical presentation and treatment given. These variables were analysed using SPSS Version 11.0.

Result

There were 232 patients with STS treated over a 22 year (1985-2006) period. Fibrosarcoma was the commonest STS constituting 36.6% of the STS seen during the period.

There were 18 patients with Dermatofibrosarcoma protuberans. The detailed analysis of these patients showed that the youngest patient at presentation was 15 years and the oldest was 65 years. The highest incidence was in the 4th decade of life (Fig.1). The male to female ratio was 3:2.

Figure 1: Age Distribution of Patients With Dermatofibrosarcoma Protuberans

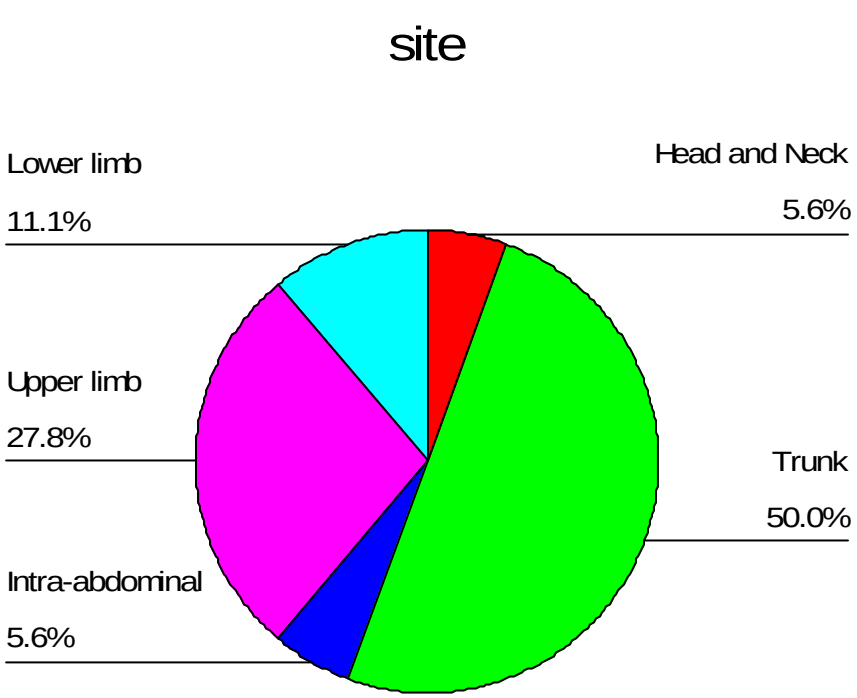


Fifty percent of the dermatofibrosarcoma protuberans seen were in the trunk, followed by upper limb in 27.8%. (Fig.2). There was only one patient with DFSP in the head and neck region and was aged 65 years. The two patients with DFSP in the lower limbs were in the age range 30-39 years. For DFSP of the upper limb in this study the male to female ratio was 2:3 (Table 1).

Table 1: Age Range, Gender And Site of Dermatofibrosarcoma Protuberans

Site			Gender		Total
			Male	Female	
Head and Neck	age	60-69			
	range		1		1
	Total		1		1
Trunk	age	10-19	1		1
	range	20-29	1	1	1
		30-39	1	1	2
		40-49	1	1	2
		50-59	1		1
		60-69	1	1	2
	Total		5	4	9
Intra-abdominal	age	50-59			
	range		1		1
	Total		1		1
Upper limb	age	20-29	2	1	1
	range	30-39	1	1	2
		40-49	1	1	2
	Total		2	3	5
Lower limb	age	30-39			
	range		2		2
	Total		2		2

Figure 2: Site of Dermatofibrosarcoma Protuberans



Most patients presented with masses ranging from 10 to 50 cm in diameter and sometimes ulcerated. The diagnosis was usually confirmed by incisional biopsy. The treatment of choice was wide local excision with appropriate skin cover. Some of the patients had chemo-radiotherapy. There was none with metastases.

Discussion

Some uncertainty exists as to when the first case of Dermatofibrosarcoma protuberans (DFSP) was reported, it may have been as early as 1890[5]. It is, however, generally acknowledged that Darrier and Ferrand established these interesting lesions as a distinct clinical entity[6]. Hoffman officially coined the term dermatofibrosarcoma protuberans in 1925[7].

DFSP accounts for less than 0.1% of all malignant neoplasms and approximately 1% of all soft tissue sarcoma The incidence has been estimated to be 0.8 – 5 cases per million population per year in 2 separate studies. In a most recent study based on data from 9 cancer registries from 1973 – 2002, the annual incidence of DFSP in the United States is 4.2 per million cases per year[8].

The annual incidence of DFSP is reported as 3 cases per million population from a population-based cancer registry from 1982 – 2002 in France[9].

During the period January, 1985 to December 2006, 232 patients with Soft Tissue Sarcoma were seen at the University of Ilorin Teaching Hospital. Eighteen (7%) patient had dermatofibrosarcoma protuberans (DFSP). This is in keeping with previous studies[10]. DFSP in this study had the highest incidence in the 4th decade, this is similar to the findings of Szollosi and Nemes[11] and Koh et al[12].

The slight male preponderance in this study is similar to previous work[13]. The sex ratio of four males to every female in Taylor and Helwig's[14] report must be, as suggested, a reflection of the Armed Forces Institute of Pathology patient source. In this study the male to female ratio in patients with DFSP of the upper limb was 2:3. Further study may be required to explain this variation.

The cause of DFSP is unknown. Laboratory studies have shown the chromosomal aberrations may contribute to the pathogenesis of DFSP; however, no evidence of hereditary or familial predisposition exists. In 10 – 20% of patients with this tumour, trauma at the site seems to be incriminated. Surgical and old burns scars and site of vaccinations have all be reported as sites of DFSP.

DFSP is seen mainly on the trunk and the proximal extremities. This may bring to fore the Binkley's[15] theory relating to the mammary ridge which used to be of historical interest.

Most patients had a history of a superficial slowly growing, painless mass. A major problem in our patients is delayed presentation. Large cutaneous mass in the rule in our patients. This delay in presentation may be because of the indolent nature of the tumour and the remarkable paucity of symptom. As a result of the enormous size at presentation, most of the patients had incisional biopsy to confirm the diagnosis.

There are lesions that can mimic DFSP; these should be considered in the differential diagnosis. They are: Dermatofibroma, Epidermal inclusion Cyst, Keloid and hypertrophic scar, malignant melanoma and metastatic carcinoma of the skin.

Imaging studies is mainly useful in metastic disease. Recent studies seem to support a role of MRI for preoperative assessment in larger or a typical lesions and recurrent disease. MRI may be helpful to define the approximate tumour border and depth of invasion[16, 17].

The investigations available to and affordable by our patients are plain x-rays, abdominal/ pelvic ultrasound and Computed Tomography (CT) when possible. It is known that CT of the chest is the best for patients with tumours greater than 5cm but is not readily available and affordable. This has always been a problem in developing country like ours[3].

Although the American Joint Committee on cancer has not set forth a system specific for staging of DFSP, it is currently staged in accordance with the American Musculoskeletal Tumour Society Staging System which takes into account tumour grade and compartmentalization[18, 19].

All the patients benefited from wide local excision. There was no case of metastases. This is not unusual. Most cases do not metastasise. Incidence of mestastasis is considered a deviation from the standard behaviour of these tumours[13].

Though the optimal treatment of DFSP is resection, the optimal mode of resection is still unclear. Patients may benefit from wide surgical excision or Mohs micrographic surgery. The

characteristic of finger-like extensions of DFSP that lies beneath clinically appearing normal skin makes complete removal difficult.

Radiation therapy has had a limited role in the past, but, recently, it has been used as an adjunct to surgery. Radiation therapy may be recommended for patients if the margins of resection are positive or for situations where adequate wide excision along may result in major cosmetic or functional deficits. Close follow-up care after radiation therapy is warranted because some of the tumours may become more aggressive[20, 21, 22].

Imatinib mesylate a molecularly targeted therapy holds promise as an additional treatment option. Imatinib is a potent and specific inhibitor of several protein-tyrosine kinases, including the PDGF receptors[20, 23].

Imatinbi mesylate is indicated for treatment of adult patients with unresectable, recurrent and/or metastatic DFSP[20].

DFSP is a soft tissue tumour that is often not recognized clinically due to it rarity and variable appearance. It is more common in the trunk and upper extremities. DFSP remains a distinct entity marked by triad of characteristic: its rarity, its slow growth and infiltrative nature. Wide local excision is still the treatment of choice.

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