
Dermatofibrosarcoma Protuberans: "Epidemiological and Clinical Study"

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Abstract

Background: Dermatofibrosarcoma protuberans is a locally aggressive invasive dermal and subcutaneous mesenchymal neoplasm. Males are affected more often than females. It typically occurs during the third and fourth decades of life.

Objective: To determine the clinical, histopathological and epidemiological aspects of dermatofibrosarcoma protuberans in Iraqi patients.

Patients & Methods: This case series study was carried out in the Department of Dermatology & Venereology-Baghdad Teaching Hospital during the period from June 2000 to December 2006. Twenty patients with dermatofibrosarcoma protuberans were included in this work. Socio-demographic information, full clinical, dermatological examination and biopsy were done for each patient.

Results: Twenty patients were involved in the present work, all showed typical clinical features of the disease. Their ages ranged from 7-75 (mean \pm SD) (38.55 ± 21.33) years, while the duration of the disease ranged from 3-24 (mean \pm SD) (23.28 ± 10.53) months. There were 14(70%) males and 6(30%) females with a male to female ratio 2.3:1.

The geographical distributions of these cases were: 9(45%) cases from South of Iraq, 11(55%) patients from Baghdad. The main sites involved were the extremities 10(50%) and 5(25%) for each one of the trunk and head. The clinical features of the diseases were not much different from what have been published in medical literatures. Histopathological study of lesions revealed the typical histological features of dermatofibrosarcoma protuberans in all cases.

Conclusion: The present study confirmed that dermatofibrosarcoma protuberans is disease of male and it is the first report from Iraq recording cases from Middle and South regions only.

Keywords: dermatofibrosarcoma protuberans, malignancy, clinical, epidemiological.

Introduction

Dermatofibrosarcoma protuberans (DFSP) is an uncommon malignant tumor arising de novo in the dermis and subcutaneous tissues, characterized by local aggressive infiltration and indolent progression. While rarely metastasizing, this mesenchymal neoplasm is notorious for its propensity to recur even after wide excision [1-3].

Males are affected more often than females. It typically occurs during the third and fourth decades of life. There are rare examples of this condition occurring in infancy [1, 2, 4-7].

The tumor is more often situated on the front of the trunk and proximal extremities of the patients. It often grows to several centimeters in diameter and may form large tumors [1, 2, 8].

The clinical morphology is variable, either it may begin in early adult life with one or more small, firm, painless, flesh colored or red dermal nodules. These nodules grow slowly, coalesce and extend, becoming a reddish or bluish as they enlarge [1-4, 8-10].

The surface is raised by irregular, protuberant swellings, and a hard, indurated plaque of irregular outline forms of the base. In later stages, a proportion of lesions become painful and there may be rapid growth, ulceration and discharge [1-4, 8-11].

It can also present as a non protuberant, atrophic, violaceous lesions simulating sclerosing basal cell carcinoma, anetoderma, or scar [2, 4, 7, 11].

Histologically, the tumor shows a subepidermal fibrotic plaque with uniform spindle cells and variable vascular spaces. In many

instances, there is a cartwheel pattern of spindle cell arrangement surrounding a central area of collagen. Giant cells and histiocytes are also present but only in small numbers [8-10, 12, 13].

The objective of the present work is to describe the epidemiological and clinical aspects of this disease among Iraqi patients.

Patients & Methods:

This case series study was conducted as outpatient hospital based clinical, epidemiological and histopathological study in the Department of Dermatology & Venereology-Baghdad Teaching Hospital during the period from June 2000 to Dec. 2006.

All patients enrolled in this work were interrogated and evaluated regarding the following points:

- 1- History was taken from each patient including: age, sex, address, duration of the diseases signs and symptoms and previous diagnosis.
- 2- Clinical examination was carried out to describe: location, size, consistency clinical characters, tenderness, and the presence of enlarged regional or distant lymph nodes.
- 3- Incisional biopsies were performed for all patients to confirm the diagnosis.

Results:

Twenty patients with the clinical diagnosis of DFSP were assessed. There were 14(70%) males and 6(30%) females with a male to female ratio 2.3:1.

Their ages ranged from 7-75 (mean \pm SD) (38.55 ± 21.33) years, while the duration of the disease

ranged from 3-84 (mean ±SD) (23.28 ± 10.53) months.

The location of the tumor was in the following regions: extremities 10(50%) and 5(25%) for each one the trunk and head (Table 1).

Table (1): Showing the locations of the DFSP in Iraqi patients.

Locations	No	%
<u>Extremities:</u>	10	50
Forearms	4	20
Arms	3	15
Thighs	3	15
<u>Head:</u>	5	25
Scalp	2	10
Forehead	2	10
Nose	1	5
<u>Trunk:</u>	5	25
Upper back	2	10
Chest	1	5
Abdomen	2	10
Total	20	100

The geographical distributions of these cases were: 9(45%) cases from South of Iraq (Basra 7, Babylon one & Najf one), 11(55%) patients from Baghdad and no cases were recorded from North of Iraq.

Regarding the clinical features of the disease, all patients had history of non painful, small papules

and nodules that gradually overtime enlarged in size and coalesced together to form large, hard masses. While examination, in all patients showed masses with irregular surface & edge, elevated, deep seated, adherent, subcutaneous tender masses, fleshy red in color with hard stony indurations (Fig.1).



Figure (1) Showing 38 years old male patient with dermatofibrosarcoma protuberans on the chest.

The size of lesions ranged from 3-6 (mean \pm SD) (4.2 ± 1.1) cm². In 6(30%) cases finally, the lesions consisted from multiple, protuberant, small, firm, and fleshy to deep red stony nodules (bosselated). The lesions were tender in 6(30%) patients only.

No single case was associated with regional lymph nodes enlargement, while systemic examination and investigations showed no abnormal findings.

Histopathological readings revealed all the features DFSP in all patients.

Discussion

There has been obvious increase in many types of tumors in Iraq like Kaposi's sarcoma, leukemia, and mycosis fungoides^(14, 15).

This was observed during the last 17 years that consisted of first Gulf war followed by blockade for 13 years then invasion and occupation of Iraq for the last 4 years. This increase might be attributed to exposure to the depleted uranium in the weapons that have been used during wars in Iraq^(14, 15).

Kaposi's sarcoma was reported mainly from Middle and South regions of Iraq where depleted uranium (DU) had been used⁽¹⁴⁾.

The present study is the first report recording DFSP in Iraqi population, and all cases were originated from the Middle and South areas of Iraq. So accordingly, we can speculate that the etiopathogenesis of this tumor could be also related to the depleted uranium exposure in concordance with the Kaposi's sarcoma⁽¹⁴⁻¹⁷⁾.

The present work also showed that males were more affected than females and this is comparable with what had been published in the literatures^(1, 2, 4-7, 13). The ages of patients that commonly affected were the 2nd and 3rd decades of life and this is similar to what had been mentioned in medical reports^(1, 2, 4-7).

The commonest sites affected in this work were extremities followed by head & trunk which is also consistent with what has been noticed in the literatures^(1, 2, 8).

The clinical and histopathological features of the diseases were not much different from those cases that have been published in medical records^(1-4, 8-13).

Nodal and systemic metastasis may occur although such biological behavior is rare. The preset report found no metastasis in any of these cases^(1, 2, 4, 11).

In conclusion, this is the first report recording cases of dermatofibrosarcoma protuberans from Middle and South of Iraq and this increase in the frequency of this tumor have been seen following Iraqi wars during which depleted uranium has been used.

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