Dermatofibrosarcoma protuberans.
A single center retrospective experience

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AIM: *Dermatofibrosarcoma protuberans (DFSP) is a rare, slowly growing, painless mesenchimal tumor particularly originating from cutaneous and subcutaneous tissues. This neoplasy mostly presents as protrude indurated plaque with brown-reddish color or same color of the skin. DFSP has a high rate of recurrence but a low rate of metastasis.*
METHODS: *We present a retrospective study of 23 patients who were diagnosed with DFSP and operated at our institution. We examined the clinicopathological parameters with clinical outcome and the follow-up.*
RESULTS: *We retrospectively analysed the data of 23 patients operated for DFSP in Ankara University Medical School Department of Surgical Oncology between 2006 and 2017. Out of these 23 patients, 14 of them were male and 9 of them were female. Dermatofibrosarcoma protuberans has been detected within body in 13 patients, extremities in 7 patients, chest in 2 patients, neck in one patient as well. 6 patients were opereted one times and 17 patients were operated twice due to getting tumor free margins. One patient devaloped local recurrence and reexcision was performed. All patients are still alive and follow up period varied from 12 up to 144 months with a median of 54.2.*
CONCLUSION: *In conclusion, DFSP is an uncommon, low-grade sarcoma of dermal fibroblast origin with a high local recurrence rate. Diagnosis is established by histology and immunohistochemistry. The greatest clinical challenge in the management of DFSP is achieving local control. Surgical excision is the treatment of choice.*

KEY WORDS: Dermatofibrosarcoma protuberans, Local Recurrence, Mesenchymal tumor

Introduction

Dermatofibrosarcoma protuberans (DFSP) was first described by Daier and Ferrand in 1924 and subsequently named by Hoffman in [1]. It accaounts for approximately %6 of soft tissue sarcomas and less than %0.01 of all [2]. DFSP is a rare, slowly growing, painless mesenchimal tumor particularly originating from cutaneous and subcutaneous tissues [3]. This neoplasy mostly presents as protrude indurated plaque with brown-reddish color or same color of the skin [2]. To great extent, these are low grade, slowly growing lesions with rare metastasis. However DFSP are associated with high local recurrence rates, especially if excised incompletely.
DFSP occurs as a result of a specific t(17;22) (q22;q13) translocation leading to the formation of COL1A1-PDGFB fusion transcripts [4]. DFSP is histopathologically divided into two types as normal (O- DFSP) and fibrosarcomatoid (F-DFSP). %90 of lesions are O-DFSP type. F-DFSP is a more aggressive tumor with high recurrence and metastasis rates [5,6] . CD 34 expression is strongly and commonly seen in DFSP but it is not specific or these tumors as it can also be seen in vascular and hematopoetic malignancies. CD99 and SMA expressions can be seen in DFSP but typically these tumors have negative S100, sitokeratin and desmine [4-7]. Apolipoprotein D expression is an important sign for DFSP as favoring the diagnosis. In addition, Apolipoprotein A provides the differantial diagnosis from fibrous histiocytoma [8,9].

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The main aim of the treatment is to fully remove the lesion within the healthy surgical margins. Wide surgical excision may be required to reach healthy surgical margins because of high local recurrence rates. There are existing studies which state that local excision is required from 2 cm to 5 cm surgical limits [10]. Subcutaneous tissue and facia shall be included in the excision area within lesion lower limit. There are existing studies which states that local recurrence grow between 26% and 60% within the cases on which local excision can not be implemented [11]. Nowadays the tumoral tissue, which is being removed using Moh's microscopic surgery, is being considered within the frozen and paraffine sections and surgical excision is being carried out until the healthy surgical margins have been reached. There are existing studies which indicate that lower local recurrence rates has been reached using Moh's mikroscopic surgery [12]. The defect occurring after applying wide surgical excision can be closed with local skin fleps or myocutaneous fleps [13].

In this study, we aimed to present DFSP cases which operated in our clinic with the literature. Age, gender, tumor localization, tumor size, recurrence and follow-up time of the patients were examined.

Methods

92 patients who underwent surgery in the department of surgical oncology, Faculty of Medicine, Ankara University between 2006 and 2017 were evaluted retrospective. The research was carried out in line with the decisions of the Helsinki Committee. The pathology results of 26 patients were DFSP. Three patients were

TABLE I - *Baseline characteristics of study population.*

Patients	Age/Sex	location	Number of Operations	Tumor Size	Follow up Period (Month)	Recurrence
1	50/Male	body	3	35x33*24 mm	28	+(After 13 months)
2	36/ Male	body	2	26x18*14 mm	130	-
3	43/Male	body	2	22*14*10 mm	40	-
4	38/Female	body	1	18*16*13 mm	36	-
5	42/Male	body	2	24*18*14 mm	20	-
6	56/Male	body	2	25x13*10 mm	26	-
7	42/ Male	body	2	24x18*11 mm	112	-
8	33/ Female	body	2	28x16*11 mm	28	-
9	41/Male	body	2	26*18*10mm	36	-
10	39/Female	body	1	15*15*9 mm	96	-
11	48/Male	body	2	28*20*10 mm	36	-
12	38/Female	body	1	18*16*11 mm	64	-
13	52/Male	body	2	28*20*16 mm	48	-
14	34/ Male	extremity	2	27x21*13 mm	48	-
15	34/ Male	extremity	2	34x28*22 mm	24	-
16	38/ Female	extremity	2	32x22*13 mm	60	-
17	44/ Male	extremity	1	31x27*21 mm	42	-
18	26/ Female	extremity	2	28x21*18 mm	48	-
19	40/ Female	extremity	2	36x27*21 mm	12	-
20	23/ Female	extremity	2	32x21*18 mm	18	-
21	43/ Male	chest	1	17x12*12 mm	144	-
22	46/ Male	chest	1	14x10*14 mm	120	-
23	26/ Female	neck	2	24x18* 12mm	30	-

TABLE II - *Demographic Distrubition of the patients.*

Gender: n (%)	
Male	14 (%60,9)
Female	9 (%39,1)
Age, year, mean±SD	39,65±8,1 (23-56)
Location	
Extremity	7 (%30,4)
Body	16 (%69,6)
Number of Operation, mean±SD, range, median	1,78±0,51 (1-3), 2
Tumor Size, cm^3,mean±SD, range, median	8,12 ±7 (2-28), 5,18
Follow-up time, month, mean±SD, range, median	54,17± (12-144), 40

excluded due to lack of regular follow-up. The remaining 23 patients were included in the study. Patients were examined age, sex, tumor localization, tumor size and pathological parameters.

Statistical Analysis

The statistical analyzes were performed using SPSS version 22.00. Pearson and chi-square test were used in nominal data analysis, Levene's Test was used in scale parametric data; and Mann-Whitney U was used in scale non parametric data. ROC curve analysis was used for finding cut-off value. P values <0.05 were considered as statistically significant.

Results

We retrospectively analysed the data of 23 patients operated for DFSP in Ankara University Medical School Department of Surgical Oncology between 2006 and 2017 (Table I). Out of these 23 patients, 14 of them were male and 9 of them were female with a mean age of 39,6 (23-56) (Table II). We found that 100% of female patients were younger than 40 years and 78.6% of male patients were 40 years and older. We found a significant relationship between age and gender (p=0.00) and there was no significant relationship between gender and other clinicopathological data (Table III). Dermatofibrosarcoma protuberans has been detected within body in 13 patients, extremities in 7 patients,

Table III - *Association between gender and other clinicopathological factors.*

Clinicopathological Factors	**No. of patients (%)**		**P value**
	Male (14)	**Female (9)**	
Age, yearP=0.00			
<40 years	3 (%21,4)	9 (%100)	
>40 years	11 (%78,6)	0 (%0)	
Number of operation, mean±SD, range, median	1,86±0,35(1-3), 2	1,67±0,5(1-2), 2	P=0.411
Tumor Size, cm³, mean±SD, range,median	8,24±8 (1,91-27,72), 5,17	7,91±5,84(2,02-20,41), 5,84	P=0.688
Follow-up time, month, mean±SD	61±44,27(20-144), 41	43,56±26,43(12-96), 36	P=0.468
Vimentin Status			
Negative	9(%64,3)	4 (%44,4)	P=0.417
Positive	5 (%35,7)	5 (%55,6)	
Location			
Extremity	3(%21,4)	4 (%44,4)	P=0.363
Body	11(%78,6)	5(%55,6)	
Continuity of surgicalmargin			
Negative	3 (%21,4)	3 (%33,3)	P=0.643
Positive	11(%78,6)	6 (%66,7)	

Table IV - *Association between location and other clinicopathological factors*

Clinicopathological Factors	**No. of Patients (%)**		**P value**
	Extremity (7)	**Body (16)**	
Age, year			
<40 years	6 (%85,7)	6 (%37,5)	P=0.069
>40 years	1 (%14,3)	10 (%62,5)	
Vimentin Status			
Negative	1(%14,3)	12(%75)	P=0.019
Positive	6(%85,7)	4(%25)	
Tumor Size, cm³, mean±SD, range, median	13,23±6,86 (1,91-20,94), 12	5,87±6,11 (1,96-27,72), 4,71	P=0.022
Mitosis, mean±SD,range, median	3,14±1,57 (1-5), 3	4,13±3,91 (1-15), 3	P=1.00

chest in 2 patients, neck in one patient as well. 85,7% of the DFSP patients in the extremity were 40 years or younger and 62,5% of the DFS patients in the body were older than 40 years (p=0.019). Tumor size was statistically significant in patients with DFSP in the extremity and the median value was 12cm³ (p=0.022) (Table IV). The cut-off value of age and tumor size data was evaluted by ROC curve method. The age data area under curve value was 0.772 and significant (p=0.042). Cut-off value is 38,5 years. The cut-off value of tumor size is 7,75 cm³. Although it was statistically significant, but the area under curve was 0,196 (Fig. 1). Five patients haver been operated within external centres and forwarded to our clinic. First diagnosis of the 18 patients were made in our clinic. While 2 patients had diagnosis after punch biopsy, 21 patients had diagnosis after excisional biopsy. 17 patients were operated twice due to getting tumor free margins. Fascia excision was performed in all patients. One patient devaloped local recurrence and reexcision was performed. All patients have been considered as normal DFSP. All patients are still alive and follow up period varied from 12 up to 144 months with a median of 54.2. Surgical excision has been applied to all patients to provide 3 cm negative surgical margin. Surgical defects have been closed primarily in 6 patients and with partially thick skin grefts in 17 patients. For all patients adequate surgical limit has been provided and chemotherapy or radiotheraphy was not required. In one patient local nux grew after 13 months follow up and the patient was reoperated. Patients are brought under control, with physical examination once in the first 3 months, once in the 6 months during the following 2 years and after 2 years annually and with BT visualisation annually.

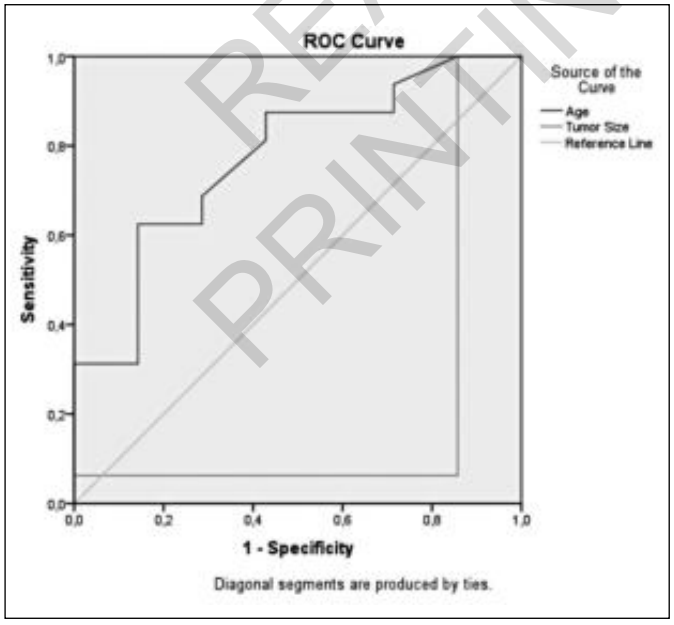

Fig. 1: - ROC, Curve analysis between location and tumor size, age.

Discussion

According to classification of World Health Organization, DFSP is evaluated as superfical, low grade, locally agressive fibroblastic neoplasm [14,15]. The most important findings of diagnosis are immunohistochemically positive expression of CD 34 and vimentin, and negative expression of s100, sitokeratins and desmin [4-7]. Annual incidence is 0.8-5/1.000.000 [16]. Despite that it has been seen among adults within the ages of 25 and 45, there are studies indicating that it has also been seen among children and adolescents [4,17]. In our study, age distribution of patients varied between 23 and 56 (median: 39.6). Unlike other studies we found a significant relationship between age and gender. In the studies male prodispodence exists and also in our study male patients was highly compatible with the literature [18,19]. Most seen localizations are body (42-72%), proximal extremities (20-30%) as well as head and neck (%10-16) [20]. In our study, DFSP is detected at body in 13 patients, extremities in 7 patients, chest in 2 patients, neck in one patient. Superficial USG has been applied preoperatively to 14 of the patients, however sufficient evaluation could not be established in definitive diagnosis.

For the definitive diagnosis of DFSP; lipomas, epidermal cysts, keloids, dermatofibromas, nodular faciit, pyogenic granuloma, kaposi's sarcoma and other soft tissue neoplasms shall be reviewed [21]. DFSP can only be diagnosed with the histological and immunohistochemical findings. Our patients are reported as normal type DFSP. Excision has been done to all patients including minimum 3 cm surgical margin with subcutaneous tissue and fascia. Only one patient had local recurrence and no metastasis was observed in any of our patients. To provide negative limits surgically, sufficiency of 3-5 cm local excision has been indicated [22]. Even in the patients where sufficient excision was applied, there are existing studies indicating local recurrence with rates of 0-21% in lesions at body, and 50-75% in lesions at location of head and neck [19]. This difference which is indicated locally can be considered as the result of inability of establishing sufficient and strong surgical limit by reason of defect and cosmetic results occured by wide excision in the head and neck location. There are studies which indicates that with moh's microscopic surgery, which is being used frequently in the last period, lower recurrence values can be established and cosmetically more positive results can be obtained than "wide local excision" [23]. All patients are still alive and lenght of follow-up varies between 12 month to 144 months (median 54.2). It is forecasted that the period which is required for local recurrence to grow is 32 months on average [18]. Only in one of our patients local relapse has been grown after 13 months, metastasis has not been seen in any of our patients. Patients has been classified within two groups as O-DFSP and FS-DFSP, during the studies which has been carried out, with 67 patients having DFSP [24]. 7 patients

has been evaluated as FS-DFSP and 60 patients has been evaluated as O-DFSP. Metastasis has been localized in 5 patients while 4 of these patients has been classified as FS-DFSP and one of these patients has been classified as O-DFSP. It was found out that metastasis ratio were meaningfully high within the FS-DFSP group. In the same study, significant connection has been indicated between lesion size and metastasis and 4 of the lesions which make metastasis are considered as bigger than 10 cm and only one of the lesions was considered as 5cm[24]. While metastasis has been most frequently seen in the lung, spreading to lymph nodes is rare, there are studies which indicates spreading to "literaturally brain, pelvis, costa and soft tissues" [19,25]. Even though DFSP is local aggressive tumor, after excision which is done with surgically negative limits, 2 and 5 year recovery rates are 97% and 92% [26].

During follow-up of patients, evaluation can be done with physical examination, mrg, thoraco-abdominal pelvic bt and usg. Patients are brought under control, with physical examination once in the first 3 months, once in the 6 months during following 2 years and after 2 years annually and with BT visualisation annually. We can not give a clear result with the statistical data of this study conducted with limited numbe rof cases. Even so, in accordance with our present findings, DFSP is seen at an earlier age in the extremity, whereas cases in the body are more commen in the older age group. In addition, cases seen in the extremity apper with larger tumor sizes. We attribute this to late hospital admissions of the patients under the conditions of our country, bacause of the similarity with lipoma. However, a multicentre study is needed for disease that occurs in less than 0.01%.

Conclusion

DFSP is a local aggressive mesenchymal tumor which can be brought under control with wide surgical excision. Diagnosis of the disease can be made with histopathologic and immunohistochemical evaluation. For patients where sufficient surgical excision can not be provided or having terminal tumor, radiotherapy and chemotherapy are being used alternatively. Patients shall be under life long follow-up due to high local recurrence values. In our study, sufficient surgical limits has been provided for 23 patients with DFSP and value of disease-free recovery has been reached without the need of radiotherapy and chemotherapy. Follow-up of our patients are ongoing.

Riassunto

Il dermatofibrosarcoma protuberans (DFSP) è un tumore mesenchimale raro, a crescita lenta e indolore, originato in particolare dai tessuti cutanei e sottocutanei. Questa neoplasia si presenta principalmente come una placca indurita sporgente di colore bruno-rossastro o dello stesso colore della pelle, ha un alto tasso di recidiva ma un basso tasso di metastatizzazione.

Presentiamo qui uno studio retrospettivo su 23 pazienti a cui è stata diagnosticata la DFSP e sono stati operati presso il Dipartimento di Oncologia Chirurgica dell'Università di Ankara Medical School tra il 2006 e il 2017. Abbiamo esaminato i parametri clinicopatologici l'esito clinico e il follow-up.

Di questi 23 pazienti, 14 erano maschi e 9 erano femmine. Il dermatofibrosarcoma protuberans è stato rilevato al livello del corpo in 13 pazienti, alle estremità in 7 pazienti, nel torace in 2 pazienti, sul collo in un paziente. 6 pazienti sono stati operati una volta e 17 pazienti sono stati operati due volte per poter ottenere margini liberi dal tumore. Un paziente ha sviluppato una recidiva locale ed è stata eseguita una nuova escissione. Tutti i pazienti sono ancora vivi e il periodo di follow-up varia da 12 a 144 mesi con una mediana di 54,2.

Conclusione: In conclusione, DFSP è un sarcoma raro di basso grado di origine fibroblastica dermica con un alto tasso di recidiva locale. La diagnosi è stabilita dall'istologia e dall'immunoistochimica. La più grande sfida clinica nella gestione della DFSP è ottenere il controllo locale. L'escissione chirurgica è il trattamento di scelta.

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