

*

*

:					가
	: 1990	3			
	12		14		
				9	13
		. 1			
			가		
:		9		1.3(1~2)	,
		11.8(2~24)			(
50.8)		13		,
:					
				2	가
:					

(histiocyte),

(fibroblast)
 endoneural cell)가

(perineural or

⁶⁾.

Darrier

CD34 가

Ferrand 1924

1925 Hoffmann

⁶⁾.

¹⁷⁾

, Hashimoto ⁷⁾

:

215-4

Tel : 02) 974-2501, Fax : 02) 977-2954, E-mail : sylee@onnet.co.kr

(modified neural supporting cell)

1

가 1

(Table 1, 11) 1, 3 5 7

가 1 (Table 1, 12)
 CYVADIC(CTX;600mg/m², VCR;1mg/m², ADR;60
 mg/m², DTIC;300mg/m²) 14

50.8

1990 3

9 12 1.3(1~2)
 11.8(2~24)

14

11

2

, 2

12

1

3

가

가 9
 1

, 2

5 7

2

2

9

가

8

1

Table 1. Clinical information of 14 patients with dermatofibrosarcoma protuberans

No	Age/ Sex	Site	F/U period*	Recur rate	Disease free period*	Final Surgical margin	Recur/ Meta	Present Status	Other Remark
1	29/F	Axilla	12	2	18	W [†]	No	CDF [§]	
2	54/F	Calf	42.3	1	2	W	No	CDF	
3	30/F	Calf	33.6	-	-	W	No	CDF	
4	33/M	Subscapula	51.8	1	13	W	No	CDF	
5	50/M	Upperarm	14	1	14	W	No	CDF	
6	43/M	Back	104	-	-	W	No	CDF	
7	18/F	Axilla	72	-	-	W	No	CDF	
8	52/F	Shoulder	12	-	-	W	No	CDF	
9	16/F	Thigh	92	1	13	W	No	CDF	
10	50/F	Calf	54	-	-	W	No	CDF	
11	22/M	Forearm	43.3	1	2	W	No	CDF	Chemo [‡]
12	38/F	Groin	59.2	1	24	I [‡]	3/lung	DOD	Chemo**
13	57/F	Thigh	42.3	2	2	W	No	CDF	
14	21/M	Subscapula	79	2	18	W	No	CDF	

*period : months, [†]W: wide margin, [‡]I : intralesional margin, [§]CDF: continuously disease free,

DOD : died of disease, [‡]chemo : initial diagnosis was synovial sarcoma, **chemotherapy was done after metastasis.

9) 가
 9 5
 16) 가
 가 (plaquelike) 16
 가
 (Fig. 1). 가
 (storiform pattern) 가
 (Fig. 2).
 (nuclear pleomorphism)
 (mitosis)
 14) 가
 (malignant fibrous histiocyoma)
 1, 4, 6, 18)
 10%
 가 CD34

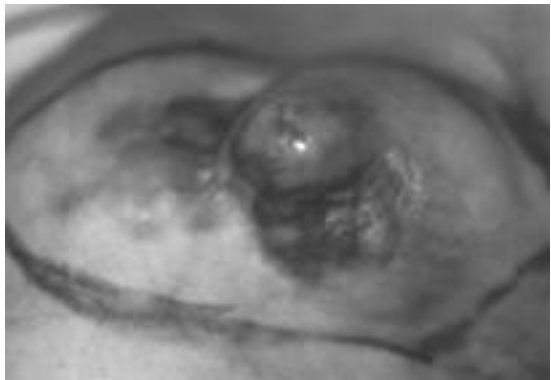


Fig. 1. Dermatofibrosarcoma protuberans from axilla shows nodular surfaced protruding mass with reddish color.

6) 가
 S100
 가
 (myxoid liposarcoma)
 (lipoblast) 가
 (,)
 2
 50%
 10, 13, 16)
 가
 Roses¹²⁾ 3cm
 , 2cm 41%
 , 4cm
 1 15
 96 5%
 가
 4, 14)

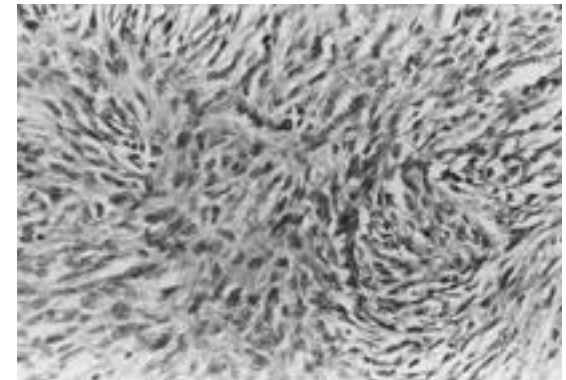


Fig. 2. Spindle cells arranged in a distinct storiform pattern characterize the majority of this tumor(H&E, × 200).

Abstract

Dermatofibrosarcoma Protuberans ; Treatment and Prognosis

**Soo-Yong Lee, M.D., Jong Hoon Park, M.D.*, Dae-Geun Jeon, M.D.,
Jong-Seok Lee, M.D., Sugjun Kim, M.D.**

*Department of Orthopaedic Surgery, Korea Cancer Center Hospital, Seoul, Korea
Department of Orthopaedic Surgery, Pundang Jesaeng General Hospital**

Purpose : Dermatofibrosarcoma protuberans(DFSP) is a rare tumor of the skin with a strong tendency of infiltration to surrounding tissues. Inadequate surgical intervention brings about frequent recurrence and poor prognosis. We attempted to find a guideline for adequate treatment for DFSP.

Materials and Methods : Fourteen cases who had been treated in our department since Mar. 1993 and followed up for more than 12 months postoperatively were reviewed. Including nine cases who were transferred from other hospital after recurrence, thirteen cases underwent wide resection. One case was treated by intralesional resection followed by chemotherapy (CYVADIC) due to neurovascular abutment to the mass in the inguinal area.

Results : The nine cases who were transferred due to recurrences experienced recurrence in average 1.3(1-2) times and the average period until first local recurrence from primary operation was 11.8(2-24) months. The thirteen cases with wide surgical margin showed no recurrence at the final follow up. One case treated by intralesional resection and chemotherapy showed multiple recurrence and died of the disease due to lung metastasis.

Conclusions : From these data, we could find that primary wide resection can be the way of reducing recurrence and metastasis, and the follow up period for the detection of recurrence should be at least two years.

Key Words : Dermatofibrosarcoma protuberans, Wide resection

Address reprint requests to

Soo-Yong Lee, M.D.

Department of Orthopaedic Surgery, Korea Cancer Center Hospital

#215-4 Gongneung-Dong, Nowon-Ku, Seoul, Korea

Tel : 82-2-974-2501, Fax : 82-2-977-2954, E-mail : sylee@onnet.co.kr