Surgical Management of Dermatofibrosarcoma Protuberans: A Siriraj Experience

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Background: *Dermatofibrosarcoma protuberans (DFSP) presents as an asymptomatic, firm, raised dermal nodule or plaque and interdigitating to surrounding tissue. That made DFSP easily inadequate resection and locally recurred. Wide excision with margins greater than 2 cm was recommended to reduce recurrence rates. Predictors for recurrence were tumor depth (Hazard ratio 3.14) and negative margin (Hazard ratio 22.43).*

Objective: *The aim of this study was to define proper margin width to achieve negative resection margin.*

Material and Method: *Patients with DFSP received wide excision at Siriraj Hospital were retrospectively reviewed between January 2008 and March 2014. Demographic data, operative records and pathologic reports were recorded, relation of resection margin and pathologic outcome were evaluated.*

Results: *Fifty-nine patients' data were collected. Patients' genders were equal. Tumors presented in head and neck region in 19 patients (32%), trunk in 26 patients (44%). The total free margin status was found in 46 patients (78%), positive margin status was found in 7 patients (12%). The median follow-up duration was 32.6 months. Two patients had recurrence. Significant difference occurs when comparing the group with margin at 1.5 cm or more and group with margin less than 1.5 cm.*

Conclusion: *The authors' recommended wide resection margins of 1.5 cm or more to achieve negative resection margin results.*

Keywords: *Dermatofibrosarcoma protuberans, Surgical management, Dermatofibrosarcoma protuberans, DFSP, Wide excision, Margin, Positive margin, Head and neck, Pregnancy*

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Dermatofibrosarcoma protuberans (DFSP) is a rare disease. The tumor is slow-growing and hard to be detected early, usually misdiagnosed and inadequately treated. With the characteristics of slender neoplastic cells interdigitating between preexisting collagen bundles, the tumor could easily be inadequately resected with only inspection or palpation. Many studies suggested wide excision with different margin width, at least more than 2 cm, which could reduce the recurrence rate from 26 to 60%[1,2] to 0 to 30%[1-7]. Increasing wider margins resulted in lower recurrence rates, but always resulted in larger defects which need complex reconstructive technique and may not be applicable in certain areas such as face, neck, or pediatric cases. Though recent studies introduced the Mohs Micrographic Surgery (MMS), with limiting resection, the defects were believed not to involve unnecessary resection. Recurrence rates were reduced to nearly 0%[2,4,5,7-11], but the cost was nearly triple of the time spent in the operating room[12] and may not be feasible in every center. Field et al suggested the predictors for recurrence were tumor depth (hazard ratio 3.14) and negative margins (R0) (hazard ratio 22.43)[13]. The aim of this study was to define proper margin width to achieve negative resection margins and report other informative demographic data of our experience in Siriraj Hospital.

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Material and Method

Approved by the Ethic Committee of Siriraj Hospital, patient records were obtained from Syria's database by coding for Dermatofibrosarcoma Protuberans (ICD-9 M8832/3). A retrospective chart review was performed between January 2008 and March

2014 for patients received wide excision of DFSP at Siriraj Hospital. Patient characteristics were collected as age, sex, site and size of tumor, pre-treatment duration and previous treatment. Operative reports were reviewed to determine margin of excision. If margin width was not documented, calculating from pathology report by subtracting the maximal length of gross tumor size and total specimen size divided by half was substituted. Patients documented as marginal excision or debulking were excluded from this study. After operation, pathological reports were reviewed for positive margin (presence of tumor at margin) or closed margin (presence of tumor at less than 0.5 cm from margin) either circumferential or deep surface. For the analysis, positive circumferential margin patients with or without positive deep margin were classified as positive margin group. No residual tumor or positive deep margin patients were classified as in the negative margin group. The immediate or delay closure was noted as well as the final reconstruction methods. The defect size was also collected. Postoperatively, patients were followed-up according to individual status and availability. The secondary procedure and adjuvant therapy were noted if performed. The duration of follow-up visit, documentation of recurrence and time to recur were collected. If the patient underwent re-wide excision at the surgical site for any reasons, the pathology report was reviewed for any residual tumor presentation.

Using SPSS v.18 program for calculation, the statistical analysis in this study used descriptive analysis for clinical factors and Fishers exact test for analysis of the relation of resection margin and pathologic outcome, with statistical significance at $p<0.05$.

Results

From January 2008 to March 2014, a total of 63 patients' charts were obtained from our database by coding ICD-9 for M8832/3. Three patients were excluded from the study because resection methods were marginal or debulking resections without further operation. These patients were diagnosed as myxoidliposarcoma, peripheral malignant nerve sheath tumor (PMNST), post-partum DFSP with positive bone scan. Another patient was excluded from the analysis, as she presented with suspected pulmonary metastasis and brain metastasis from persisted recurrent DFSP at vagina and underwent left pneumonectomy. The remaining 59 patients were diagnosed DFSP and were treated by plastic surgeons, orthopedists and gynecologists at Siriraj Hospital. Patients' demographic data were shown in Table 1.

* Surgeons who treated DFSP in this study included plastic surgeons, orthopedists and gynecologists.

** Previous treatment to patients prior to wide excision in Siriraj hospital, primary meant patient did not undergo any operation or received only incisional biopsy; inadequate meant patient received excision and pathologic reports were DFSP, recurrent meant patient received wide excision before and tumor recurred upon following up period.

*** Duration from time that patients detected their mass to operation date Patients' genders were equal, 30 males and 29 females (51% and 49%, respectively). The mean age was 41 years (18 to 74

Table 1. Patients' demographic data

Parameters	n (%)
Sex	
Male	30 (51)
Female	29 (49)
Age (years)	
Min	18
Max	74
Mean	41.05
Median	37
Age (intervals)	
≤20	1 (2)
21 to 40	34 (58)
41 to 60	17 (28)
>60	7 (12)
Department*	
Plastic	51 (86)
Other	8 (14)
Previous treatment**	
Primary	21 (36)
Inadequate	29 (49)
Recurrent	9 (15)
Time to treatment (months)***	
Min	2
Max	300
Median	36
Site	
Head & neck	19 (32)
Trunk	26 (44)
Upper extremity	5 (9%)
Lower extremity	9 (15%)
Size (cm)	
Min	1.5
Max	30
Median	5

years). Subgroup classification of age was as shown in Table 1. Of the total, 51 patients (86%) were treated by plastic surgeons. For the tumor characteristics, 21 patients (36%) were treated with wide excision primarily after either physical examination or incisional biopsy. There were 29 patients (49%) who were treated by excision and further wide excision was performed after pathology report indicated DFSP. The other 9 patients (15%), who were diagnosed DFSP and underwent wide excision from other hospitals had recurrence and were treated in our hospital. The median duration from patient detection to treatment was 36 months (2 to 300 months). Tumors presented in head and neck region in 19 patients (32%), trunk in 26 patients (44%), upper extremity in 5 patients (9%) and lower extremity in 9 patients (15%). The median size at operation was 5 cm (1.5 to 30 cm). The median margin width was 2.5 cm (0.4 to 5 cm). Margin width and pathology outcome is presented in Table 2.

* Margin width obtained from operative note or by subtracting the maximal length of gross tumor size and total specimen size divided by half.

** Positive margin meant presence of tumor at circumferential margin regardless to presence at deep margin.

*** Negative margin meant no tumor presence at circumferential margin regardless to presence at deep margin or no residual tumor detected.

**** Positive rates were calculated from positive group population compared to the same margin width group.

The totally free margin status was found in 46 patients (78%) while positive deep margin was found in only 3 patients (5%); no tumor was reported in 3 patients (5%). These patients were classified as negative margin group. Six patients in this group had report free margins, but with closed margin ranging from <0.1 to 4 mm. The positive margin group was divided into circumferential positive margin which were found only in 2 patients (3%) and both circumferential and deep positive margins, which were found in 5 patients (8%). Four patients in this group had re-wide excision and pathology reports were of free margin. Of those 7 patients whose margin status was found positive, their margin widths were 0.5, 1, 2, 3 cm. The tumor size in these patients ranged from 3.5 to 17 cm.

Four patients (7%) had myxoid DFSP while eight patients (14%) had DFSP with fibrosarcomatous transformation, and one patient (2%) had myxoid DFSP with fibrosarcomatous transformation.

Of the total defects, 43 patients (73%) had immediate closure and 16 patients (27%) had been delayed for pathology report. The closure techniques were primary closure in 23 patients (39%), grafting in 20 patients (34%), local flap in 2 patients (3%) and free flap coverage in 14 patients (24%).

Twenty patients (34%) received postoperative radiotherapy, 2 patients were from positive deep margin group and the other 2 patients were from positive circumferential and deep margin group. One patient received preoperative chemotherapy (Paclitaxel and Cisplatin) and postoperative chemotherapy (Cisplatin), which she had recurrence within 7 months postoperative and underwent re-wide excision followed by postoperative chemotherapy (Cisplatin, Ifos, Mesna).

The median follow-up duration was 32.6 months (2 to 113 months). Eleven patients (19%) had lost to follow-up (3.5 to 55 months). Only two of all patients in this study (3%) had recurrent disease. One was a 49-year-old woman with 7 cm DFSP at vulva for which she had chemotherapy courses as mentioned above and had 1.5 cm wide margin resection. Pathology report was DFSP with fibrosarcomatous transformation, free margin but closed deep and circumferential margin. Seven months later, she had recurrent tumor with 2 cm margin re-wide excision and the result was positive deep margin. Consequently, she received another chemotherapy course. The other recurrent patient was 33-year-old man with 5 cm DFSP at his right cheek and had 2 cm wide margin resection. The pathology report was positive deep margin, then anterior maxillectomy was done and defect was closed by radial forearm free flap. He was lost to follow-up appointments but was detected recurrent at 55 months postoperative. Incisional biopsy confirmed recurrent DFSP and he was scheduled for re-wide excision after the end of this study period.

In addition, we also found that in 5 women in

Table 2. Margin width and pathology outcome

Margin width (cm)*	Population	Positive margin**	Negative margin***	Positive rate****
≤1	7 (12%)	3 (5%)	4 (7%)	42.9%
1.5	5 (8%)	0	5 (8%)	-
2	16 (27%)	2 (3%)	14 (24%)	12.5%
2.5	3 (5%)	0	3 (5%)	-
3	19 (32%)	2 (3%)	17 (29%)	10.5%
4	5 (8%)	0	5 (8%)	-
5	4 (7%)	0	4 (7%)	-
Total	59 (100%)	7 (12%)	52 (88%)	12%

the reviewed charts were detected with tumor enlargement during pregnancy or postpartum period, with their age ranged from 26 to 35 years. This number accounted for 16% of all women and 24% of reproductive age women.

For the correlation between various factors and positive margin, we found a significant correlation when comparing the groups with margin at 1.5 cm or more and groups with margin less than the 1.5 cm cut point. Other cut point comparisons between margin width such as 2 cm and more margin width did not have significant results. The comparison between each margin width is presented in Table 3. All patients with positive margin were male. Of 19 patients with tumor in head and neck region, 1 patient (5.2%) had positive circumferential margin, 4 patients (21%) had positive circumferential and deep margin, 3 patients (15.8%) had positive deep margin. The overall rate of inadequate resection was 42% for tumor at head and neck region.

Discussion

As generally found, Dermatofibrosarcoma Protuberans is a rare disease, with annual incidence of about 4.2 to 4.5 cases per million in the US[14,15]. Rouhani et al analyzed 2,229 cases of DFSP from the Surveillance, Epidemiology, and End Results (SEER) program database from 1992 to 2004 and found DFSP accounted for 18% of sarcomas, occurring predominantly on the trunk (42%), extremities (40%), head and neck (16%). Incidence rates peaked in the 40s. The survival rates were high, up to 99%[15]. The other large series reported by Cai et al found overall 5-year survival rate was 98% and 10-year survival rate was 95.7%[16].

The tumor had fibroblast origin and was characterized by a plaque-like or nodular growth pattern involving dermis and subcutaneous tissue. There was a diffuse proliferation of uniform spindle cells arranged in a compact storiform pattern. The peripheral margins were ill defined, with slender neoplastic cells interdigitating between preexisting collagen bundles, along connective tissue septa, and between lobules of subcutaneous adipose tissue. That made DFSP easily inadequate resection and often left the tumor at the margin and locally recurred[17].

Clinically, DFSP usually presents as an asymptomatic, firm, raised dermal nodule or plaque, and diagnosis is frequently delayed because of the large differential diagnosis and the typically low clinical suspicion of malignancy in this slowly growing lesion[18].

Historically, DFSP had high recurrence rates as high as 26 to 60% for conservative surgical margins[1,2]. The recurrence rates decreased with wide local excision margins >2 cm which varied from 0% to 30%[1-7], increasingly wider margins resulted in lower recurrence rates. However, the wider resection margins always resulted in larger defect which need complex closure technique and may not be applicable in certain areas such as face or neck or pediatric cases.

Mohs Micrographic Surgery (MMS) uses microscope to trace out the tentacle-like projections and a map to guide residual tumor excision. With limiting resection, the defects were believed not to involve unnecessary resection. Recurrence rates were reduced to nearly 0%[2,4,5,7-11]. Meguerditchian studied the effectiveness between 2 cm margin wide excision, and MMS resulted in higher rate of positive margins. Nevertheless, MMS spent nearly triple of the time spent in the operating room[12] and may not be feasible in every center.

The population in previous studies was almost inadequate for comparative analysis. Only a few large series reported safe margins for wide excision and recurrence rates came from Cai, Field and Farma[13,16,18] as well as the other studies as summarized in Table 4.

Fields et al reviewed 244 patients treated at Memorial Sloan-Kettering Cancer Center (MSKCC) and calculated that the predictors for recurrence were tumor depth (HR 3.14) and negative margin (R0) (HR 22.43). The overall local recurrence rate was 4% at 10 years[6]. Between the positive and negative margins groups, the 5-year local recurrence rates were 8% vs. 2.3%, respectively, and the rates of distant metastasis at 10 years were 3.6% vs. 1.4%, respectively[6]. The other studies reported distant metastasis rates of about 2 to 3.5%[6,16]. This emphasized the importance of achieving

Table 3. Comparison of margin widths

Calculation of each cut point			*p*-value
Narrow margin (cm)	Wide margin (cm)	Positive rate (%)*	
<1.5	≥1.5	42.9	0.030
≤1.5	>1.5	25	0.141
<2.0	≥2.0	25	0.141
≤2.0	>2.0	17.9	0.240
<3.0	≥3.0	16.1	0.428
<4.0	≥4.0	14	0.581

* Positive rates were calculated from positive margin population compared to the same margin width group

Table 4. Summary of recent studies' results

Author	Year	Population	Margin (cm)	Recurrence rate (%)	Follow-up period (months)
Chuongsakul	2014	59	0.4 to 5	0	32.6
Goldberg[19]	2013	25	2.5	0	108
Hamid[20]	2013	30	2.5 to 3	20	68
Hersant[21]	2013	66	1.8	0	34
Cai[16]	2012	217	Overall	8.5	84
			≥3	5.7	
			1.5 to 2.5	13.6	
Fields[13]	2011	240	2 to 3	5.8	50
Farma[18]	2010	206	2	1	64
Meguerditchian[12]	2010	28	2	3.6	50
Heuvel[22]	2009	38	2 to 3	7	89
Yu[23]	2008	25	3	0	68
Lindner[24]	1999	35	2.1	8	58
Khatri[25]	2003	11	2.5	0	54
		13	3.3	0	
Monnier[26]	2006	4	<0.9	50	115
		31	1 to 2.9	46	
		31	≥3	7	
Chang[26]	2004	60	≥3	16	59
Dubay[5,10]	2004	43	1 to 2	0	48
Gloster[8]	1996	39	2.5 to 5	12.8	36
Ratner[9]	1997	58	2	41	58
			2.5	24	
			3	11 to 20	

negative margin. Fewer studies mentioned correlations between resection margin width and pathologic outcomes, as shown in Table 5.

According to our experience at Siriraj Hospital, with 75 months period of study, we had 63 patients, which averaged at 10 new patients per year. For the 59 patients enrolled, we found no sexual predominance (51: 49) as same as other reports[14,15]. However, statistical significance of correlation with positive margin occurred to male group as all cases were male. More than half of the patients age was within 21 to 40 years, with mean and median age of 41 and 37 years, respectively, which are correlated with previous studies[14,15]. The interval between the time that the mass was first detected and the treatment was about 3 years, with median size at presentation of 5 cm reflecting the asymptomatic and slow growing of disease. Half of the patients were treated inadequately before wide excision, which pointed out the unawareness, misdiagnosis, and difficulty to distinguish from other diseases. Fifteen percent of the patients had recurrence after wide excision from other centers. From the median 32.6 months of observation period, we had only 2 patients with recurrence (3.4%) at 7 and 55 months. Most of the tumor occurred in trunk, followed by head and neck region as reported by the others[15]. The tumor in head and neck region has statistically significant correlation with positive margin which is consistent with previous studies[12,25]. No metastasis was observed in this study.

The margin width and positive margin correlation was statistically significant when calculated using cut point at the widths of less than 1.5 cm and 1.5 cm or more. The narrow margin group had positive margin rate of 42.9%. This rate tended to be lower with the wider margin width groups, 12.5% at 2 cm and 10.5% at 3 cm, same as previous reports by Farma and Stojadinovic[18,28]. No positive margin was found in the group with margin width of 4 and 5 cm. However, the small population in each group and the lack of positive margin patients in every group were the limitations in this study. The exact optimal margin width might not be concluded but the very high rate in the group of less than 1.5 cm could be the reference for further study or clinical practice.

Tumor at head and neck region, in this study,

Table 5. Correlations between Resection Margin Width and Pathologic Outcomes

Author	Year	Populations	Margin (cm)	Positive rate (%)	Recurrence rate (%)
Chuongsakul	2014	59	≤1	42.9	0
			2	12.5	
			3	10.5	
			4 to 5	0	
Hersant[21]	2013	66	1.8	21.2	0
Goldberg[19]	2013	25	2.5	48	0
Fields[13]	2011	240	2 to 3	14.5	5.8
Farma[18]	2010	74	≤1	26	1
		113	1 to 2	16	
		16	2 to 3	19	
Meguerditchan[12]	2010	28	2	21.4	3.6
Heuvel[22]	2009	38	2 to 3	5	7
Dubay[5]	2004	43	1 to 2	5	0
Stojadinovic[28]	2000	16	<2	69	9
		17	≥2	0	
Gloster[8]	1996	39	2.5 to 5	15	12.8

had the high rate of inadequate resection either circumferential margin or deep margin or both. The overall rate of inadequate resection was 42% for tumor at head and neck region. Despite of the principle of resection of one investment layer of tumor, the positive deep margin was found in 8 patients (13.5%). Five of these patients (55%) had tumors on their cheeks while the others had tumors on their nose, scalp, shoulder and foot. The location in face might cause the hesitation of complete resection as planned thus resulted inadequate.

The pathological transformation had no correlation with positive margin rate, but was found in one of the recurrent patients whose report was free margin. According to the report by Goldblum[29], it suggested no increased risk of local recurrence or distant metastasis in sarcomas arising in DFSP patients. Twenty patients (34%) had postoperative radiation therapy and no recurrence was observed in these patients.

Five pregnancy related women were observed in this study, representing 24% of reproductive age female patients. Previous studies reported observation of rapid growing of tumors aggravated by pregnancy[30-35] but could not reach the conclusion. By the report of Parlette et al[30], the suspected accelerator was an increased expression of PDGF receptors in DFSP patients as found in other neuroectodermal tumors (Chromosomes 17 and 22 cytogenetic abnormalities) and further increases in PDGF during pregnancy. Cytogenetic studies have demonstrated the chromosomal translocation of 17q22 and 22q13. This phenomenon has led to fusion of the platelet-derived growth factor (PDGF) gene and the collagen type 1 alpha 1 chain (COL1A1), resulting in activation of the PDGF receptor. The other suspect from Palette's report was increased Progesterone receptors (PR), which found weak to moderate staining, but also appeared similarly in men and non-pregnant patients. The following reports found no PR receptors staining in their studies[32-35].

Conclusion

It is controversial for oncological safety in dermatofibrosarcoma protuberans resection margin. Statistically significant correlation occurred to resection margin width less than 1.5 cm and positive tumor at circumferential resection margin, which was nearly 50% chance. Although limited by the small population, the authors suggested the resection margin of at least 1.5 cm or more. The study confirms better outcome for tumor control with minimal risks of recurrence.

What is already known from this topic?

Dermatofibrosarcoma protuberans (DFSP) presents as an asymptomatic, firm, raised dermal nodule or plaque and interdigitating to surrounding tissue. That made DFSP easily inadequate resection and locally recurred. Wide excision with margins greater than 2 cm was recommended to reduce recurrence rates. Predictors for recurrence were tumor depth (hazard ratio 3.14) and negative margin

What this study adds?

Wide resection margins of 1.5 cm or more to achieve negative resection margin resulted in a minimal risk of recurrence.

Potential conflicts of interest

None.

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แนวทางการผ่าตัดโรค dermatofibrosarcoma protuberans จากประสบการณ์ของศิริราช

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ภูมิหลัง: Dermatofibrosarcoma protuberans (DFSP) เป็นเนื้องอกที่ไม่มีอาการแข็งและนูนจากผิวหนังมักจะกลมกลืนไปกับเนื้อเยื่อโดยรอบ ทำให้ไม่สามารถตัดเนื้องอกออกอย่างเพียงพอทำให้มีโอกาสกลับเป็นซ้ำได้ การตัดแบบกว้างโดยมีขอบเขตห่างจากเนื้องอก 2 เซนติเมตร เป็นวิธีที่จะสามารถลดอัตราการกลับเป็นซ้ำ สิ่งที่ใช้สำหรับทำนายการกลับเป็นซ้ำ คือ ความลึกของเนื้องอก (hazard ratio 3.14) และผลการตัดไม่พบเนื้องอกที่ขอบ (hazard ratio 22.43)

วัตถุประสงค์: จุดมุ่งหมายเพื่อการศึกษานี้เพื่อกำหนดขอบเขตที่เหมาะสมในการผ่าตัดเนื้องอก DFSP เพื่อให้ตัดได้หมดสมบูรณ์

วัสดุและวิธีการ: เป็นการทบทวนผู้ป่วยที่มี DFSP ที่ได้รับการผ่าตัด ณ โรงพยาบาลศิริราช ตั้งแต่ มกราคม พ.ศ. 2551 ถึง 2557 โดยดูข้อมูลประชากร, บันทึกการผ่าตัด, และประเมินความสัมพันธ์ของขอบเขตที่ตัดและผลทางพยาธิวิทยา

ผลการศึกษา: พบข้อมูลของผู้ป่วย 59 รายที่ถูกบันทึก จำนวนเพศชายและหญิงเท่ากัน เนื้องอกพบในบริเวณศีรษะและคอ จำนวน 19 ราย (32%), ลำตัว 26 ราย (44%) จำนวนผู้ป่วยที่ทำผ่าตัดและไม่พบเนื้องอกที่ขอบ 44 ราย (78%), ผลขอบเนื้องอกเป็นบวกมี 7 ราย (12%) ค่าเฉลี่ยของระยะเวลาที่ผู้ป่วยมาติดตามการรักษาคือ 32.6 เดือน พบว่ามีผู้ป่วย 2 ราย ที่กลับเป็นซ้ำ พบความแตกต่างที่มีนัยสำคัญระหว่างกลุ่มที่ตัดเนื้อมากกว่าหรือเท่ากับ 1.5 เซนติเมตรจากขอบเมื่อเทียบกับกลุ่มที่ตัดน้อยกว่า 1.5 เซนติเมตรจากขอบ

สรุป: ในการผ่าตัดรักษา DFSP ผู้นิพนธ์แนะนำให้ตัดเนื้องอกห่างจากขอบอย่างน้อย 1.5 เซนติเมตร เพื่อให้ได้ผลการตรวจเนื้อทางพยาธิวิทยาที่ขอบเขตเป็นลบ