

Case Series

The varied presentation of dermatofibrosarcoma protuberans: a case series of 9 patients treated in single institute

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Received: 28 May 2021

Revised: 01 July 2021

Accepted: 03 July 2021

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare skin tumor with an estimated incidence of 0.8 to five cases per 1 million people per year. To our surprise, we'd encountered total 9 cases of DFSP with different presentation in our hospital from November 2009-July 2019. One patient with underlying DFSP presented with intraperitoneal bleeding tumor while another 8 patients were newly diagnosed. The mean patient age at the time of diagnosis were 38.4 years. Sex distribution were male in predominance. The anatomical location of tumor varied. Majority presented clinically with slow growing mass on skin/subcutaneous tissue, one tumor grew on a preceding scar, and 2 patients came with bleeding tumors. The median follow-up duration in this series was 23.6 months. Wide local excision was performed in 6 patients, with half of them reported to have surgical margins involved. Reconstruction of the surgical defect took place primarily in 4 patients and split skin graft in 2 patients. Adjuvant radiotherapy was offered to 3 patients with surgical margin involved and disease recurrence. Imatinib was given to one patient with metastatic DFSP prior definitive surgery which was successful. There were no local recurrence or death due to the DFSP course at the end of the follow-up. Final outcome was favorable. Hereby we present the 9 interesting case series.

Keywords: Dermatofibrosarcoma protuberans, Wide local excision, Mohs micrographic surgery, Recurrence

INTRODUCTION

DFSP is a slow growing but locally aggressive tumor with varied presentation. Often described as skin colored, erythematous, mildly elevated plaque, studded with solitary or multiple nodules. The overlying skin can become stretched, shiny, or ulcerated.¹

DFSP usually located on the trunk or proximal extremities. Clinical finding, biopsy and histopathology are standards for diagnosis. Microscopically DFSP give rise to spindle-shaped, monomorphic tumor cells which are found in the dermis and/or subcutaneous.² Immunohistochemical marker CD34 is useful for diagnosis. The mainstay of treatment for DFSP is surgery. Local recurrence of DFSP after incomplete resection is common, but distant metastasis is rare and probably closer to 1%.^{3,4}

CASE SERIES

Case 1

60 years old Malay female presented with right sub-mammary swelling for 3 months in 2009. Clinical examination showed a 4×5 cm swelling below right breast. FNAC done showed atypical cells suspicious of malignancy. Core biopsy suggestive of DFSP.

HPE of WLE reported as DFSP. Tumor size 50×45×40 mm. All surgical margins are free of tumor but very close to deep surgical margin (<1 mm). Histological section showed cellular spindle cells arranged in storiform and exhibited moderately enlarged nuclei with occasional mitosis. Immunohistochemical showed positive CD34 and vimentin but negative to desmin and actin.

USG HBS excluded liver metastasis and MRI of chest showed no evidence of residual disease. In view of long delay after surgery, she was treated conservatively. She has been put up for yearly surveillance with clinical examination and Mammogram whereby local recurrence was excluded. She was last seen in outpatient clinic on December 2020.

Case 2

16 years old malay male has been diagnosed with DFSP of left thigh in 2011. Surgical excision and skin flap done in October 2011 and he had completed adjuvant radiotherapy in January 2012. He presented to us on March 2013 with complaint of worsening right iliac fossa pain for 2 days.

Per abdomen examination revealed peritonitis. With the impression of perforated appendicitis, case posted for exploratory laparotomy. To our surprise, intraoperative findings revealed a pelvic tumor sitting below pelvic brim, bleeding anterior to the bladder dome mainly on the right side. Bladder was pushed up anteriorly. Appendix and all intraperitoneal organs appeared normal. Exploratory laparotomy, tumor debulking, ligation of bilateral internal iliac arteries, appendectomy done.

HPE reported pelvic tumor tissue as DFSP. Mild nuclear pleomorphism were seen with mitosis 7 per 10 hpf. Myxoid change was seen in focal areas. Its immunohistochemical profile showed positive reaction to CD34 and vimentin but negative to SMA, desmin, and CD117.

CECT TAP showed an enhancing cystic mass at the right pelvic measured 5.3×2.8×6.0 cm. The mass displaced urinary bladder anteriorly. No focal lung/liver lesion. No significant lymphadenopathy. Post operation, he was complicated with disseminated intravascular coagulopathy. He had a turbulent recovery and discharged on day 8 post operation. He was then referred back to orthopedic and oncology team in view of metastatic DFSP for further management.

Case 3

33 years old malay male who presented with scalp swelling for 1 year, gradually increasing in size. He first sought for treatment in 2012 whereby he went to our clinic and investigated with MRI brain which showed no intracranial extension. He underwent WLE in 2012 and referred for radiotherapy. However, patient defaulted RT and he had recurrence the next year and WLE was done.

HPE of the scalp swelling showed DFSP. Scattered capillaries noted within the tumor. It's immunohistochemical showed positive to CD34, Vimentin, and focal weakly positive to S100. Patient was referred for adjuvant RT but he was not keen for the treatment. Currently he is well with no local recurrence clinically.

Case 4

22 years-old Chinese male presented with right inguinal swelling for 2 years. Clinical examination showed right inguinal swelling measured 6×3 cm, well-defined, firm mass, associated with erythematous skin. USG inguinoscrotal showed a well-defined hypoechoic lesion over subcutaneous tissue of right inguinal area, likely represent inguinal lymph node. However, unable to rule out soft tissue tumor in view of increased vascularity. FNAC result consistent with soft tissue lesion. In view of inconclusive imaging and FNAC result, excision biopsy over right inguinal soft tissue was performed.

HPE reported as DFSP. Tumor size measured 45×40×30 mm. Mitosis was 3 to 5 per hpf. It's immunohistochemical profile showed positive reaction to CD34.

Patient defaulted follow up and was unable to be contacted for inquiry regarding current performance status and clinical evidence of local recurrence.

Case 5

21 years-old malay female presented with right shoulder swelling for months. Clinically there was a pedunculated mass over anterior right shoulder measuring around 6x4 cm. Excision biopsy over right shoulder was performed. HPE reported as DFSP, specimen measured around 40x30x10 mm.

Microscopically there were unencapsulated nodule composed of spindle cells arranged in storiform pattern. The cells exhibited enlarged nuclei with mitosis about 2 per 12 hpf. Abundant of hemosiderin laden macrophages seen. A few Touton's type of giant cells are present. The resection margins involved. It's immunohistochemical profile showed positive to CD34, Vimentin, and S 100, but negative to HMB 45. Patient defaulted follow up and was unable to be contacted for patient for inquiry regarding current performance status and clinical evidence of local recurrence.

Case 6

12 years-old Chinese male presented with swelling over abdominal wall for 1 year. The swelling gradually increased in size and associated with pain occasionally. USG abdomen done in April 2017 suggestive of complex cyst at the epigastric/left hypochondriac region. Differential diagnosis included infected sebaceous cyst and dermoid cyst. In view of presence of peripheral vascularity in USG, proceeded with CECT abdomen.

It showed left hypochondriac enhancing lobulated hypodense lesion arising from cutaneous/subcutaneous tissue abutting the abdominal wall. No intramuscular or intraperitoneal extension. Differential diagnosis includes soft tissue hemangioma and infected sebaceous cyst.

Excision biopsy done in June 2017. It was reported as incompletely excised DFSP. Patient underwent WLE and Split skin graft over abdominal wall in July 2017.

HPE reported as DFSP with incomplete deep margin resection (nodule to surgical margin measured around 3 mm). Nodular lesion measured 20×10×10 mm. The tumor cells were extended from dermis into subcutaneous fat. Mitosis was rarely seen.

Patient was referred for adjuvant radiotherapy and he completed #30 cycles radiotherapy in November 2017. CECT Abdomen post radiotherapy showed left anterior upper abdomen subcutaneous thickening was more compared to previous scan. It might represent post radiation changes/residual tumor. There was no distant metastasis. Patient was reviewed by oncologist and no further adjuvant therapy needed.

USG abdomen was done one month later and there was homogenous thickening of skin at the upper abdominal wall at the skin flap site likely represent post skin flap/radiotherapy changes. No definite abnormal nodule along or beneath the skin thickening.

During his follow up in August 2018, clinical examination reviewed scar over SSG area in thickness and hard in consistency. Suggested for wedge biopsy and his parents wished to continue further management in other hospital. Patient has been under 6 monthly follow up and no recurrence detected so far.

Case 7

31 years old malay female presented with swelling over right infraumbilical region for 8 years. She went to seek treatment only when the swelling started to bleed and became painful. USG Abdomen performed in August 2017 showed a suprapubic subcutaneous hypoechoic lesion, differential diagnosis included abscess, infected sebaceous cyst, or epidermoid cyst. Initially she was treated as abdominal wall soft tissue tumor and excision biopsy done.

HPE reported as DFSP. Tumor size measured 40×35×30 mm. The cells exhibited mildly pleomorphic elongated to oval nuclei with vesicular chromatin, small nucleoli and moderate eosinophilic cytoplasm with indistinct cell borders. Few accompanying elongated vasculatures were noted. Mitoses were 2 per 10 hpf. The lesion was incompletely excised. The cells were diffusely positive for CD34 and Vimentin with weak and focal positive Actin. Ki 67 proliferative index was 25%. They are negative for Desmin, S100, HMB 45, bcl2.

CECT TAP done noted small nodule at anterior segment of right upper lobe of lung with sclerotic lesion at vertebral body of T8 and left pedicle of L2, might represent metastasis. Otherwise, no enhancing subcutaneous lesion/nodule. Bone scan reported no finding suggestive of

bone metastasis. Oncologist planned for imatinib and after 2 cycles of tyrosine kinase inhibitor, patient was posted for WLE. HPE reported no evidence of residual neoplastic lesion. USG abdomen surveillance after 1 year showed no recurrent lesion at the subcutaneous suprapubic region. Patient is on yearly clinical surveillance and last seen in our clinic on November 2020.

Case 8

78 years-old Chinese male with underlying CCF, COAD, and hypertension. Patient had history of excision biopsy of left upper back lipoma in 2010. Swelling appeared over the previous excision scar 4 years later. He noted the swelling was gradually increased in size and finally he came to seek treatment in November 2018.

Clinical examination revealed irregular multinodular swelling, firm to hard consistency, measured around 8×5 cm. FNAC showed spindle cell lesion with mild atypia. CECT Thorax showed left posterior shoulder subcutaneous lesion with no muscle/bony involvement. Thereby proceeded with WLE+SSG in March 2019. Mass excised with 1cm circumferential margin.

HPE reported as DFSP. Tumor size measured around 40×70×15 mm and 10×7×7 mm. However, tumor tissue appeared to involve the inferior margin, which was only 1mm away from smaller tumor. The spindle cells showed strong immunopositivity for CD34 and weak positivity for Actin. The cells were negative for S100.

Post-operation, patient's wound was complicated with surgical site infection. Case referred to oncology team and suggested for re-excision in view of wound not well-healed. Patient not keen for surgery and defaulted follow up after last reviewed in June 2019.

Patient was contacted and claimed his wound was healed and no recurrence. He refused for radiotherapy and further intervention in view of old age and logistic issue.

Case 9

73 years-old, malay male presented with left inguinal swelling which was gradually increased in size over 1 year. Patient came to E on July 2019 due to bleeding from the left inguinal mass. On examination, the huge lesion measured around 15×10 cm over left inguinal region with central ulceration.

With the impression of bleeding dermatofibrosarcoma protuberans, case posted for wide local excision. Elliptical incision surrounding the lesion with the margins of more than 5 cm on each quadrant. No deep muscular involvement and successful primary closure achieved. The left inguinal swelling HPE reported as DFSP with all surgical margins clear. Tumor size measured 110×90×77 mm as shown in (Figure 1). Patient defaulted follow up subsequently. He was contacted and enquired regarding

his condition. Patient is doing well and clinically no recurrence.



Figure 1: Case 9 (A) DFSP on left inguinal region; (B) WLE specimen.

DISCUSSION

Surgical excision of primary tumor with tumor-free margin remains the mainstay of treatment for DFSP. WLE has been used conventionally as the treatment of choice for DFSP. However, MMS has emerged as a better treatment option with increasing number of supporting data. Precise and complete evaluation of surgical margin during MMS lower the recurrence rate compared to surgical excision. In view of no MOHS Surgeon in our center, WLE is the treatment available for DFSP patients and we tried to achieve tumor free margins.

We found that there was little agreement among authors regarding the most appropriate surgical margin to take during wide local excision for DFSP. Most authors concurred on surgical margin range from 2-4 cm as per National Comprehensive Cancer Network (NCCN) guideline recommendation for standard surgical margin for excision of DSFP. European Dermatology Forum (EDF) recommended 3 cm surgical margin with deep fascia as deep margins. Whereas Denmark guideline recommended 2-3 cm with deep fascia as deep margins.⁵ Some authors agreed on the idea of wide margins resulted in lower recurrence rate like Roses et al as they observed margins of 2 cm or less associated with recurrence rate of

41%, whereas margins of more than 2 cm resulted in 24% recurrence rate, and margins of at least 3 cm including the underlying fascia resulted in the lowest rate of 20%.⁶ On the other hand, Farma et al suggested smaller surgical margins could also yield satisfactory result.⁷

Their studies with 206 patients had shown recurrence rate of less than 1% with narrow margins of 0.5-3 cm. However, the technique used in their studies were similar to slow Mohs procedures to which multiple excisions to achieve clear margin and delay primary closure till negative histological margins obtained. So, the result was more representative of MMS than WLE. In our series, only Case 9 had documented 5 cm surgical margins taken and repaired with primary intention. Otherwise, surgical margins not specified in other cases.

DFSP poses a challenge in its treatment due to its aggressive nature for local invasion. It has 'tentacle-like' projections of neoplastic cells that extend deep into fascia and muscle and these extensions cannot be appreciated clinically. Failure to excise these extensions are held responsible for high local recurrence rate of DFSP after conventional surgery.⁸ Ratner et al had provided a reason behind the difficulty to achieve clear margins.⁹ With their experience treating 58 patients with DFSP using MMS, they found that 70% of tumor extended at least 1 cm microscopically beyond grossly visible lesion, followed by 15% of tumor extended 3 cm microscopically and 5% at least 5 cm extension microscopically.

Parker et al also conducted study to determine appropriate surgical margin by tracking subclinical extension with histopathological measurement using MMS.¹⁰ Their study showed a stunning 80% of tumor with complete excision with 1.5 cm margins. Thus, their opinion of surgical margin for DFSP was inconsistent with the recommended margins of more than 3 cm as it risked unnecessary removal of normal tissues. Parker and colleagues recommended surgical margins of 1.5 cm for tumor less than 2 cm and 2.5 cm margins needed for larger tumor. Same margins applied for lesions on difficult anatomical site like head and neck. Nevertheless, they realized that the clearance percentage for this margin would be too narrow for standard excision.

In our current series, 5 patients who underwent WLE and 2 of them had involved margins. followed up with mean duration of 23.6 months showed no local recurrence. 2 of them were offered radiotherapy but they refused. One female patient (Case 7) presented with anterior abdominal wall DFSP and lungs metastasis. She was offered Imatinib followed by WLE and she was successfully treated. Bowne et al study had told us that the prognosis after surgical resection with negative and sometimes positive microscopic margins for patients with DFSP was very good.¹¹ However, increase age, high mitotic index, and increased cellularity were predictors of poor clinical outcome.¹² So far, no mortality recorded in our series.

Table 1: Summary of clinical data.

Case	Age (years)	Sex	Area	Treatment	Diameter (mm)	Margins involved	H/o previous operation	F/up (months)	Recurrence
1	60	F	Submammary R	WLE (2010)	50×45×40	No, but very close to deep margins (<1 mm)	No	136	No
2	16	M	Pelvic	Exploratory laparotomy, tumor debulking, ligation of bilateral internal iliac arteries, appendectomy	**	**	WLE + vascularized skin flap (2011) + RT (2012)	**	CECT TAP suggestive of metastasis/recurrence
3	33	M	Scalp	WLE (2013) + RT (patient defaulted RT)	18×10×2	Yes	WLE (2012) + RT (patient defaulted)	12	No +
4	22	M	Inguinal R	Excision biopsy (2015)	45×40×30	No	No	**	**
5	21	F	Shoulder R	Excision biopsy (2016)	40×30×10	Yes	No	**	**
6	12	M	Anterior abdominal wall	WLE + SSG (2017) + RT	30×25×15	Yes, deep margin (3 mm)	No	16	CECT Abdomen post RT (2018) No
7	31	F	Anterior Abdominal wall	2 cycles of oral imatinib 400 mg OD WLE of anterior abdominal wall (2018)	No evidence of residual neoplastic lesion.	No	No	38	No. CECT TAP (2017). Suspicious lung and bone metastasis.
8	78	M	Upper back L	WLE+ SSG (2019) + RT (Patient not keen for RT)	A)40×70×15 B) 10×7×7	Yes, inferior margin (1 mm)	History excision biopsy for lipoma (2010)	8	No
9	73	M	Inguinal L	WLE (2019)	110×90×77	No	No	2	No

Note: F- Female; M- Male; L- Left; R- Right; WLE- Wide local excision; RT- Radiotherapy; **Not applicable; + No local recurrence after second WLE.

CONCLUSION

WLE with surgical margins at least 3 cm can be used to achieve lower recurrence rate as possible in center where MMS not available. Our patients' outcome was good though they didn't undergo MMS and this implied that DFSP can be co-managed by general surgeon and oncology team.

ACKNOWLEDGEMENTS

I wish to express my gratitude to my colleague, Dr. Nurul

Nadia Binti Mohammed Saffari who helped me in data collection during the writing of this case series.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Ping LS, Barathilingam KALN. The varied presentation of dermatofibrosarcoma protuberans: a case series of 9 patients treated in single institute. *Int J Res Dermatol* 2021;7:724-9.