DOI: 10.14260/jemds/2015/175

CASE REPORT

DERMATOFIBROSARCOMA PROTUBERANS IN UMBILICAL REGION: A CASE REPORT

S. Mythili Devi[1], K. Babji[2]

HOW TO CITE THIS ARTICLE:
S. Mythili Devi, K. Babji. "Dermatofibrosarcoma Protuberans in Umbilical Region: A Case Report". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 07, January 22; Page: 1266-1269,
DOI: 10.14260/jemds/2015/175

ABSTRACT: Dermatofibrosarcoma protuberans is a rare fibroblastic mesenchymal skin tumor with low-grade malignancy and occasional distant metastasis. The tumour accounts for less than 0.001% of all malignancies. Most cases involve the trunk, extremities, scalp, and neck. In rare cases, the genital urinary system may be affected. Diagnostic tools include computed tomography and magnetic resonance imaging. The standard treatment of wide-margin surgical excision with a clear margin of 3-4 cm may be problematic in large tumors or in some areas of the body, as a radical excision may produce unacceptable cosmetic, structural, or functional results. An un resected or positive margin should be treated with adjuvant radiotherapy to help prevent recurrence. Chemotherapy plays a limited role in treating this disease. This article describes a 55years male with mass in umbilical region who underwent wide local excision and rhombhoid flap transfer to cover the defect. Later patient sent for adjuvant radiotherapy, patient was tumour free at 6 months follow up.
KEYWORDS: Dermatofibrosarcoma protuberans, wide excision.

INTRODUCTION: Dermatofibrosarcoma protuberans (DFSP) is a rare fibroblastic mesenchymal skin tumor. The most commonly affected area is the trunk followed by the extremities, scalp, and neck. Although DFSP is a low grade malignancy of the skin and subcutaneous tissues with low potential for the developing distant metastases, treating DFSP is often challenging.(1) This article describes a case of umbilical dermatofibrosarcoma protuberans following a literature review, the clinical presentations, histopathological characteristics, treatment modalities, and outcomes of DFSP are reported.

CASE REPORT: A 55 year old male visited the out-patient department of government general hospital, Kakinada with chief complaint of ulcerative swelling in umbilical region for 5months. The patient is hypertensive on treatment since 10years. The patient was admitted for further evaluation and management. On admission, his blood pressure was 130/80mmHg, pulse rate was 88/min, respiratory rate was 16/min, body temperature was 98.4F.Physical examination revealed a 5x3cm ulcerated tumour in umbilical area, which is of firm in consistency, bleeds easily.

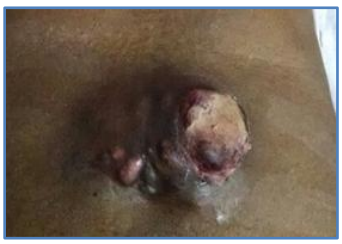

Fig. 1: Gross view of umbilical ulcerative tumour

DOI: 10.14260/jemds/2015/175

Laboratory results disclosed normal findings (haemoglobin:10.2g/dl, white blood cells:6200/cu.mm,platelets:150000/microlitre, random blood sugar: 136mg/dl, blood urea nitrogen: 26mg/dl, serum creatinine:1mg/dl. Chest radiograph was normal. There is no regional lymphadenopathy. MRI shows The mass lesion revealed a low signal intensity appearance with tiny tail like extensions superiorly on T1 weighted image and in fat saturated T2 weighted axial MR image with its high signal intensity pattern shows tail like deep extensions in to dermis in umbilical region. Fine needle aspiration failed due to copious vascularity.

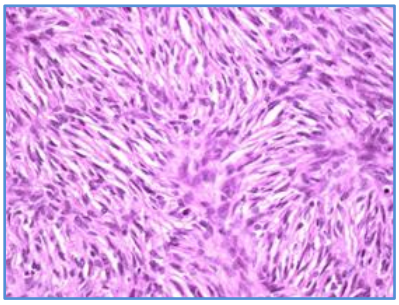

Fig. 2: Histopathology specimen showing proliferative neoplastic spindle cells arranged in a vague storiform pattern (hematoxylin & eosin staining;)

Assuming as a malignant tumour, wide local excision with 2cm of clearance margin was done. Due to the resulting large skin and soft tissue defect, rhombhoid flap was created by a plastic surgeon. The post-operative outcome was uneventful, and the patient was discharged on 7th day. Histologically, the tumour was composed of proliferative neoplastic spindle cells arranged in a vague storiform pattern. The immunohistochemical study revealed that the tumour cells were strongly positive for CD34 and negative for smooth muscle actin, CD117, and S100. The final pathology confirmed a diagnosis of DFSP with negative margins. Postoperatively patient sent for adjuvant radiotherapy. No tumour recurrence was noted in 6 months follow up.

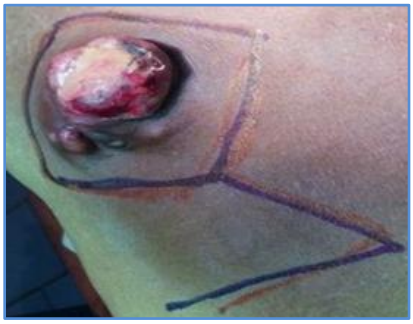

Fig. 3: wide local excision with rhombhoid flap transfer

DISCUSSION: DFSP is a rare, low-grade malignancy involving the skin and subcutaneous adipose tissue, and in rare cases, lung or lymph node metastasis. Most DFSP cases affect the trunk and proximal extremities (> 70%), although the most common single site is the back. No gender difference in DFSP has been reported. A prior trauma to the tumor site may contribute to tumor

DOI: 10.14260/jemds/2015/175

development.[2] Histological study usually reveals multiple spindle cells in a storiform pattern. As in the reported case, tumor cells usually stain positive for CD 34. Optimal management of DFSP cases is wide resection of the tumor with at least 3 cm of a tumor-clear margin or mohs micrographic surgery. However, resection might not be possible in large tumors or in special areas, because the radical excision may have unacceptable cosmetic, structural, or functional consequences. The local recurrence rate is 20%~50% in cases with incomplete resection.[2] An unresectable or positive margin should be treated with adjuvant RT. Adjuvant RT can reliably reduce the local recurrence rate and avoid the mutilation and functional deficit caused by repeated surgery.[3] In this case, the tumour had spread to surrounding skin and to deeper soft tissues. Due to the wide distribution of the tumor as well as cosmetic considerations in this case, the local tumor was excised with 2 cm of a grossly clear tumor margin. The pathology confirmed a diagnosis of DFSP. The patient sent for postoperative adjuvant RT to minimize the possibility of local recurrence.

In addition to indications during its typical clinical course, imaging studies are necessary to determine the extent of tumor invasion. Primary diagnostic tools are CT scans, MRI, and sometimes angiography. A CT scan allows the detection of the tumor anatomy and the invasion depth and usually shows a well-defined and moderately enhanced tumor with a distinct lobular or nodular architecture and tissue attenuation nearly equal to the muscle density.[4] Those tumors with a CT scan showing marginal enhancement and central low density caused by necrosis may have a poor prognosis.[5] Although MRI study is nonspecific, arteriograms can reveal mild to moderate hypervascularity.[4]

The earliest study on DFSP described 10 cases including 3 patients treated by RT alone and 7 patients treated by adjuvant RT with surgery. In the latter group, 6 achieved a tumour-free status after 16-105 months of follow-up.[6] Suit et al. described 18 cases of DFSP. Fifteen were treated by adjuvant RT plus surgery while 3 were treated by RT alone. The combined treatment group achieved a 10-year local control rate of 88%.[7] The largest published study of 38 DFSP cases by Haas et al. also revealed the local control rate achieved by combination therapy to be superior to that by surgery alone (82% vs. 7%, respectively).[8] Sun et al. reported similar results in a study of 35 cases of DFSP and also documented a higher control rate in the combined treatment group than in the group receiving surgery alone (80% vs. 28%).[3] In the latest report by Dagan et al. of 10 cases of DFSP (including 1 case of a DFSP-fibrosarcoma) treated with combination therapy, 9 achieved a tumor-free status, and only 1 died of the DFSP-fibrosarcoma.[9] Although DFSP is non-sensitive to chemotherapy, Hg et al. reported a case of pediatric DFSP with chemosensitivity. The patient was given a trial treatment with weekly vinblastine and oral methotrexate. A good partial response was achieved after 6 weeks, and complete remission had been achieved at the 5-month follow-up.[2]

CONCLUSION: DFSP is a skin malignancy with a high recurrence rate if resection is inadequate. Thus, adjuvant radiotherapy is effective in decreasing the recurrence rate and improving local control. Chemotherapy plays a limited role in treating this disease. Close surveillance is mandatory for 5 years or longer to identify late recurrence.

REFERENCES:

1. Mc Arthur GA. Dermatofibrosarcoma protuberans: a surgical disease with a molecular savior. Curr Opin Oncol 2006; 18: 341-6.

DOI: 10.14260/jemds/2015/175

2. Hg A, Nishikawa H, Lander A, et al. Chemosensitivity in pediatric dermatofibrosarcoma protuberans. J Pediatr Hematol Oncol 2005; 27: 100-2.
3. Sun LM, Wang CJ, Huang CC, et al. Dermatofibrosarcoma protuberans: treatment results of 35 cases. Radiother Oncol 2000; 57: 175-81.
4. Mark J. Kransdorf, Jeanne M. Meis-Kindblom. Dermatofibrosarcoma protuberans: Radiologic appearance. Am J Roentgen 1994; 163: 391-4.
5. E. Miyakawa, H. Fujimoto, K.Miyakawa et al. Dermatofibrosarcoma protuberans: CT findings wih pathologic correlation in 6 cases. Acta radiol 1996; 37: 362-5.
6. Lawrence B. Marks, Herma D.Suit, D.Phil et al. Dermatofibrosarcoma protuberans treated with radiation therapy. Int J Radiat Oncol Biol Phys 1989; 17: 379-84.
7. Suit H, Spiro I, Markin HJ, et al. Radiation in management of patients with dermatofibrosarcoma protuberans. J Clin Oncol 1996; 14: 2365-9.
8. Haas RLM, Keus RB, Loftus BM, et al. The role of radiotherapy in the local management of dermatofibrosarcoma protuberans. Eur J Cancer 1997; 33: 1055-60.
9. Dagan R, Morris CG, Zlotecki RA, et al. Radiotherapy in the treatment of dermatofibrosarcoma protuberans. Am J Clin Oncol 2005; 28: 537-9.

AUTHORS:

1. S. Mythili Devi
2. K. Babji

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Surgery, Rangaraya Medical College, Kakinada.
2. Professor, Department of General Surgery, Rangaraya medical College, Kakinada.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. S. Mythili Devi,
9-169/1, Sarpavaram Junction,
Kakinada-533005,
East Godavri District, Andhra Pradesh.
E-mail: mythilidevi64@gmail.com

Date of Submission: 12/01/2015.
Date of Peer Review: 13/01/2015.
Date of Acceptance: 16/01/2015.
Date of Publishing: 21/01/2015.