

Dermatofibrosarcoma Protuberans: A Deceptive Neoplasm.

K Vagholkar, I Gopinathan, S Nair, S Nachane, O Joglekar, A Vaishampayan

Citation

K Vagholkar, I Gopinathan, S Nair, S Nachane, O Joglekar, A Vaishampayan. *Dermatofibrosarcoma Protuberans: A Deceptive Neoplasm.* The Internet Journal of Oncology. 2012 Volume 8 Number 2.

Abstract

Dermatofibrosarcoma protuberans is a slow growing deceptive tumor. It is a low grade malignancy with potential to develop local recurrence as well as metastases especially to the lungs. Surgery is the mainstay of treatment. Neoadjuvant therapy is a useful adjunct in large and recurrent tumors. Molecular targeting with imatinib mesylate is a promising option. Optimum neoadjuvant therapy followed by Moh's micrographic surgery is the best therapeutic modality with least morbidity and mortality. A case of recurrent dermatofibrosarcoma protuberans is presented along with a review of literature.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare slow growing neoplasm of the skin, usually arising from the dermis. Darier and Ferrand first described this tumour in 1924 as a distinct entity and named it as progressive and recurring dermatofibroma. Hoffman was responsible for naming the tumour as dermatofibrosarcoma protuberans in 1925. [1] It accounts for less than 0.1% of all malignant neoplasms and approximately 1% of all soft tissue sarcomas. [1,2] The annual incidence of DFSP as reported in literature based on population based cancer registries of various countries is less than 3 cases per million populations making it a rare tumor.[3] The tumor is slow growing with a potential for metastasis in a select few cases. Recent advances in molecular cytogenetics have led to evolution of therapies which are partially useful as an adjuvant to surgery which still remains the mainstay of treatment for this tumour. We report here a case of recurrent DFSP along with a review of literature.

CASE REPORT

A 45 year old male patient presented with history of multiple exophytic growths overlying the right infraclavicular region. The patient had already undergone surgery for a similar growth 5 years back at the same site. 3 years after the surgery the patient redeveloped exophytic growths which have grown over a period of time. (Figure 1) There was no regional lymphadenopathy. Chest x ray was normal. Pre-operative diagnosis by FNAC revealed a spindle cell tumour. The patient underwent wide local excision taking utmost care to achieve a 2.5 cm resection margin all around the

previous scar. (Figure 2) The entire lesion was excised right up to the pectoral fascia (Figure 3). The defect was closed primarily. Post-operative recovery was uneventful. Histopathological examination of the tumour revealed dermatofibrosarcoma protuberans with clear resection margins (fig.3). Patient has been started on imatinib mesylate. The patient has been following up for the last 2 months with no evidence of recurrence.

Figure 1

Figure 1: Multiple exophytic growths seen along with the scar of previous surgery.



Figure 2

Figure 2: Resected specimen



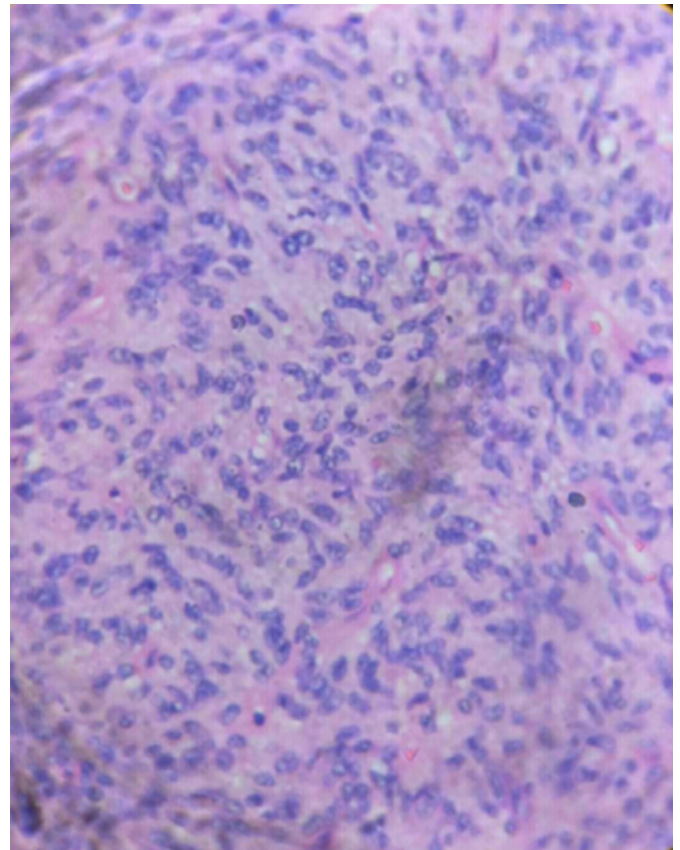
Figure 3

Figure 3: Resected specimen including some part of the pectoral fascia



Figure 4

Figure 4: Ilassical storiform pattern with monotonous presence of spindle shaped cells. (Magnification 100X)



DISCUSSION

DFSP is a locally aggressive tumour with high recurrence rate. It has no racial predilection as per previous studies. An uncommon pigmented variant of DFSP accounted for 1% of DFSP called Bednar tumour has been described to have a high incidence in Blacks.[4] The tumor has a slight male predilection usually in the age group of 20-50 years. A multicentric variant of DFSP is seen to occur in children suffering from Adenosine deaminase – Deficient severe combined insufficiency (ADA-SCID). [5] Development of DFSP has been attributable to significant changes in the molecular cyto-genetics of the patient. This usually involves chromosomes 17, 22 and in other rare cases even t (5; 8). There is usually a chromosomal translocation involving COL1A1 gene on chromosome 17 and the gene platelet derived growth factor B (PDGF B) on chromosome 22. The resultant deregulated expression of PDGF B leads to continuous activation of PDGF receptor β protein tyrosine kinase and promotes DFSP tumour cell growth. [6] Understanding this mechanism has helped in revolutionizing neo-adjuvant treatment for this tumour. [7]

Diagnosis of this tumour can be done by FNAC. However, exacting details are best revealed by histopathological examination of the specimen. Metastatic workup in cases of recurrent tumours involves evaluation of lungs followed by lymph nodes. Magnetic resonance is the investigation of choice for evaluating the extent of local spread. [8] However, lymph node involvement is extremely rare. Surgery is the treatment of choice for fresh cases. The size of the tumour dictates the extent of resection and the need for reconstruction thereafter. Pre-operative neo-adjuvant therapy is helpful in reducing the size of the tumour, thereby, rendering it more amenable to resection without the need of sophisticated reconstruction. Neo-adjuvant treatment comprises of a tyrosine kinase inhibitor namely, Imatinib mesylate along with radiotherapy. [9, 10] If the tumours are resectable without the need of advanced reconstruction, surgery remains the mainstay of treatment. The traditional surgical treatment for this tumour is its resection with a margin of at least 2.5cm to prevent local recurrence. Since, this may necessitate further reconstruction in view of the wide defect created, Moh's micrographic surgery (MMS) helps in resolving this issue. MMS helps in reducing the extent of resection, thereby conserving significant amount of normal tissue. Therapeutic efficacy in the form of cure rates by MMS is superior to wide local excision.[11] In view of high potential for recurrence, continued follow up is mandatory. Recurrent tumours are usually more aggressive. Surgery should be preceded by molecular targeted therapy to improve the outcome in such patients. Radiotherapy is also a viable option, but its efficacy is inferior to that of imatinib mesylate. The dose of radiotherapy varies from 50 to 70 Gy. Local recurrence rate with just radiotherapy is just 24-90%. [12] Imatinib mesylate is administered orally in the dose 400-800mg daily. [7] Literature pertaining to dosage scheduling is insufficient to detect the response to imatinib therapy. Side effects of imatinib are fluid retention, nausea oedema, anaemia, skin rash, fatigue, thrombocytopenia, vomiting, neutropenia and diarrhea. Imatinib mesylate treatment is specifically indicated in large tumours, recurrent tumours and in cases with positive resection margins. [10]

CONCLUSION

In conclusion, DFSP is a surgical disease with a molecular

savior. Neo-adjuvant treatment in the form of tyrosine kinase inhibitor and radiotherapy leads to tumour down staging, less cosmetic disfigurement and functional impairment with improved surgical morbidity.

ACKNOWLEDGEMENTS

We would like to thank the Medical Superintendent of Rajawadi Municipal General Hospital, Mumbai -77, India for allowing us to publish this case report.

We would also like to thank Parth K. Vagholkar for his help in typesetting the manuscript.

References

1. Lemm D, Mugge LO, Mentzel T, Hoffken K. Current treatment options in dermatofibrosarcoma protuberans. *J Cancer Res Clin Oncol.* May 2009; 135(5):653-65.
2. Dimitropoulos VA. Dermatofibrosarcoma protuberans. *Dermatol Ther.* Nov-Dec 2008; 21(6):428-32.
3. Hussain SK, Sundquist J, Hemminki K. incidence trends of squamous cell and rare skin cancers in the Swedish national cancer registry point to calendar year and age dependent increases. *J Invest Dermatol.* May 2010; 130(5): 1323-8.
4. Quigley EA, Marghoob AA, Busam KJ, Chen CS. A firm red brown plaque on the arm. Dermatofibrosarcoma protuberance (DFSP), pigmented variant (Bednar tumor). *Arch Dermatol.* May 2009; 15(5): 589-94.
5. Kesserwan C, Sokolic R, Cowen EW et al. Multicentric dermatofibrosarcoma protuberans in patients with adenosine deaminase deficient severe combined deficiency. *J allergy Clin Immunol* Dec 2011 (Epub ahead of print)
6. Abott JJ, Oliveira AM, Nascimento AG. The prognostic significance of fibrosarcomatous transformation in dermatofibrosarcoma protuberans. *Am J Surg Pathol.* Apr 2006; 30(4): 436-43.
7. McArthur G. Molecularly targeted treatment for dermatofibrosarcoma protuberans. *Semin Oncol* April 2004; 31(2 suppl6): 30-6.
8. Riggs K, McGugan KL, Morrison WB, Samie FH, Humphreys T. Role of magnetic resonance imaging in perioperative assessment of dermatofibrosarcoma protuberans. *Dermatol Surg.* Dec 2009; 35(12):2036-41.
9. McArthur G. Dermatofibrosarcoma protuberans: recent clinical progress. *Ann Surg Oncol* Oct 2007; 14(10):2876-86.
10. Han a, Chen EH, Niedt G, Sherman W, Ratner D. Neoadjuvant imatinib therapy for dermatofibrosarcoma protuberans. *Arch Dermatol.* Jul 2009; 145(7):792-6.
11. Gloster HM Jr, Harris KR, Roenigk RK. A comparison between Moh's micrographic surgery and wide surgical excision for the treatment of dermatofibrosarcoma protuberans. *J Am Acad Dermatol.* Jul 1996; 35(1): 8207.
12. Dagan R, Morris CG, Ziotecki RA, Scarborough MT, Mendenhall WM. Radiotherapy in the treatment of dermatofibrosarcoma protuberans. *Am J Clin Oncol.* Dec 2005; 28(6): 537-9.

Author Information

Ketan Vagholkar, MS, DNB, MRCS, FACS.

Professor, Department of Surgery, Dr. D. Y. Patil Medical College

Indumati Gopinathan, MD

Consultant Pathologist, Clinicopathological Laboratories

Shalini Nair, MS

Associate Professor, Department of Surgery, Dr. D. Y. Patil Medical College

Siddhartha Nachane, MBBS

Resident, Department of Surgery, Dr. D. Y. Patil Medical College

Omkar Joglekar, MBBS

Resident, Department of Surgery, Dr. D. Y. Patil Medical College

Amogh Vaishampayan, MBBS

Resident, Department of Surgery, Dr. D. Y. Patil Medical College