

RECURRENT DERMATOFIBROSARCOMA PROTUBERANS OF SCALP

Muhammad Azhar Qureshi

Military Hospital Rawalpindi

INTRODUCTION

Dermatofibrosarcoma protuberans is an uncommon cutaneous tumour arising from fibroblasts, histiocytes or neuroectodermal cells of dermis and invades fat, muscles and even bone. It is a slow growing locally aggressive fibrous tumour with high recurrence rate. Its relatively rare occurrence lessens clinical awareness and most of the lesions remain undiagnosed until biopsy is done. Less than 5% of these tumors are located on the scalp and the recurrence rate is very high in this area¹. Here it can invade skull or even brain². Very rarely it can metastasise to lymph nodes, orbit, lungs or even pancreas³. It commonly occurs in adults but has been also reported in children⁴ and old age group. Diagnosis is made by biopsy of the lesion. The mainstay of treatment is wide surgical excision that is not always easy in head and neck area and newer techniques, combined with radiotherapy and chemotherapy in recurrent and metastatic lesions, are evolving.

CASE REPORT

A 31 years old soldier reported in surgical outdoor of PAF Hospital Faisal Karachi with a recurrent nodular skin lesion, about 2 X 3 cm in size, in frontal region of scalp. He had already undergone excision of a similar lesion 2½ years ago from a peripheral military hospital. The operation details and biopsy report were not available with the patient. The recurrence took place just 6 months after the first surgery and the growth gradually increased to attain the present size (Fig.1). The growth was not attached with underlying bone and overlying skin was not ulcerated. Regional lymph nodes were not involved. X-Ray Chest, abdominal ultrasound, and routine laboratory investigations were within normal limits. Biopsy of the lesion revealed dermatofibrosarcoma protuberans. Wide local excision of tumour was carried out and the

defect was covered with local flap. The tumour recurred just 3 months after second surgery and patient was transferred to CMH Rawalpindi for multimodality approach. The recurrent tumour was not fixed to the skull and there was no lymph node or visceral involvement. CT scan of brain was normal. Wide local excision was done once again and scalp transposition flap was used to cover the area. Donor site was covered by skin grafting (Fig. 2). After wound healing patient was given radiotherapy. A total dose of 60 Gy external radiation was given in 30 fractions in 6 weeks. Short term follow up of one year after surgery revealed no recurrence.



Fig. 1: Dermatofibrosarcoma protuberans of scalp.

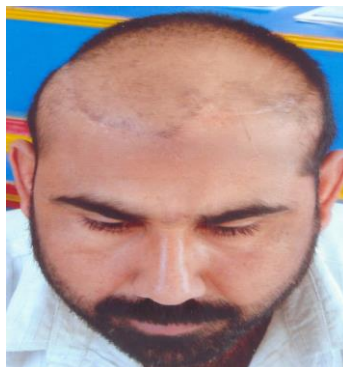


Fig. 2: Result after one year of surgery and radiotherapy

DISCUSSION

The case reported here shows a classical example of slow growth and recurrent nature of this tumour. Due to inadequate resection and no post operative radiotherapy the tumour recurred twice.

Cases of dermatofibrosarcoma protuberans were probably reported in literature for the first time in 1890. Later, in 1924, Ferrand and Darier, and in 1925 Hoffman, described this disease as

Correspondence: Lt Col Muhammad Azhar Qureshi
Classified Surgical Specialist, MH Rawalpindi
Email: surgazhar@yahoo.com

Received: 27 March 2009; Accepted: 13 Oct 2009

a distinct clinical entity⁵. It has high recurrence rate due to tentacles of tumour invading adjacent tissue. Due to this reason most of recurrent cases are local and very rarely it can metastasise to lymph nodes, orbit, lungs or even pancreas. In this case also the tumour did not metastasize to local lymph nodes despite local recurrence twice. This tumour has been described in various parts of the body including limbs, trunk, and head and neck. It has marked predilection for the trunk and head and neck regions are rarely involved. Deeper extension can involve skull or even brain.

Diagnosis can be made by careful history, and examination and should be confirmed by biopsy of the lesion. Dermatofibroma, keloid, melanoma, metastatic carcinoma of skin, and morphoea are skin lesions that need to be carefully differentiated. Imaging techniques are only helpful when there are metastases. The mainstay of treatment is wide surgical excision including 3 to 5 cm margins of healthy tissue. Some workers are of the opinion that a 2.5 cm surgical margin through the deep fascia (nonscalp) or periosteum (scalp) in lesions greater than 2 cm and 1.5 cm surgical margin in lesions smaller than 2 cm can clear the whole tumour⁸. If biopsy at the base of the lesion is positive and further excision is not possible due to its location the post surgical radiotherapy is mandatory. When local lymph nodes are involved wide local excision of lesion combined with block dissection is done. Reconstruction of resulting defect is done by various techniques using rotation flaps or free flaps. In scalp lesions where bone is involved wide local excision of scalp and underlying bone is followed by cranioplasty. In areas of head and neck wide local excision is not always technically feasible. Mohs Micrographic Surgery is a newer technique employed for certain carcinomas like BCC, melanoma and dermatofibrosarcoma protuberans in head and neck area⁶. It allows precise microscopic marginal control by using horizontal frozen sections. It gives superior cure rates, maximum tissue conservation, and ability to trace perineural infiltration. Postoperative RT may reduce the risk of local recurrence in patients with DFSP who have a high risk of residual

disease after surgery⁷⁻⁸. Chemotherapy has a limited role in the treatment of dermatofibrosarcoma protuberans. However, the tyrosine kinase inhibitor imatinib mesylate has been used as a chemotherapeutic agent in locally recurrent and metastatic lesions. Dermatofibrosarcoma protuberans is not a lethal tumour when localized and 5 years survival rate after surgery is 99.2 %. However, Long term follow up is necessary even when lymph nodes are not involved in order to detect any metastasis in time. Most lesions recur within three years after resection. During this time every 3 to 6 months follow up is necessary. Thereafter, annual follow up is sufficient.

The case presented here is a soldier and due to exigencies of military service he had to move from one city to another. There are many pitfalls in the initial management of the case presented here. The growth was initially excised in CMH Kohat with no record of treatment or biopsy report available with the patient. The tumour excision was probably inadequate in the second surgery. Moreover, it was not followed by radiotherapy resulting in treatment failure. This highlights the significance of multi-disciplinary approach in treating dermatofibrosarcoma protuberans especially in head and neck area where wide local excision is technically more demanding and not always possible and in such cases surgery should be followed by radiotherapy.

REFERENCES

1. Loss L, Zeitouni NC. Management of scalp dermatofibrosarcoma protuberans. *Dermatol Surg*. 2005 Nov; 31(11 Pt 1): 1428-33
2. Marakovic J, Vilendecic M, Marinovic T, Lambasa S, Grahovac G. Intracranial recurrence and distant metastasis of scalp dermatofibrosarcoma protuberans. *J Neurooncol*. 2008 Mar 15; [Epub ahead of print]
3. Yokoyama Y, Murakami Y, Sasaki M, Morifuji M, Hayashidani Y, Kobayashi T, Sudo T, Sueda T. Pancreatic metastasis of dermatofibrosarcoma protuberans. *J gastroenterol* 2004 Aug; 39(8): 798-800.
4. Keshen TH, Cederna PS, Savell VH Jr, Platz CE, Chang P, Ricciardelli EJ. Clinical and pathological features of pediatric dermatofibrosarcoma protuberans. *Ann Plast Surg*. 1995; 35: 6: 633-7.
5. McPeak C J, Cruz T, Nicastrì A D. Dermatofibrosarcoma protuberans: an analysis of 86 cases--five with metastasis. *Ann Surg*. 1967 November; 166; 5: 803-16.
6. Salomon D, Adatto M, Skaria AM. [Mohs micrographic surgery: concept, technic and indications] *Rev Med Suisse*. 2006 Apr 26; 2(63): 1094-6, 1098-9. French.
7. Tariq Mufti, M Saeed Khan, Muzher U Dua, Farah Waqar. Dermatofibrosarcoma Protuberans - A Rare Variant of Soft Tissue Sarcoma of Skin. An Experience of Two Cases at Ayub Teaching Hospital Complex (ATHC), Abbottabad. *J Ayub Med Coll Abbottabad* Dec 2001; 13: 4: 32-4
8. Dagan R, Morris CG, Zlotecki RA, Scarborough MT, Mendenhall WM. Radiotherapy in the treatment of dermatofibrosarcoma protuberans. *Am J Clin Oncol*. 2005; 28: 6: 537-9.