Dermatofibrosarcoma Protuberans of the Forehead

B Sayanthan[1], S Lavanya[2], R Ramprasad[2] and M Udukala[2]

Batticaloa Medical Journal 2016: 7: 32 – 33.

Introduction

Dermatofibrosarcoma Protuberans (DFSP) is an uncommon fibrohistiocytic tumour. It is a slow to intermediate growing tumour and commonly affects middle aged people. DFSP commonly occurs in the trunk and then the limbs. The potential for distant metastasis is low, but in DFSP local recurrences are common (1).

Case report

A 33 year old male had observed a small lump in the forehead region 8 months ago. He had been symptomless and had had no history of weight loss, night sweats, fevers or chills. The lymph nodes in the head and neck region and the axillae had been non-palpable. The lump had gradually increased in size. It had been clinically diagnosed as a sebaceous cyst and surgical excision had been done, but he had not received the histological report.

A couple of months later he had developed a lump at same site of the forehead and this had gradually increased in size. The history was uneventful and the examination findings were similar to the first episode.

Thereafter, he underwent surgery again with the histology indicating that it was a Dermatofibrosarcoma Protuberans(DFSP) with margin involvement.

Therefore, he underwent a wide local excision with full thickness skin grafting of the defect.

Histology and immuno-histochemistry confirmed the diagnosis.

Discussion

Dermatofibrosarcoma Protuberans (DFSP) is a very rare type of a soft tissue sarcoma, which originates in the deep layer of skin.

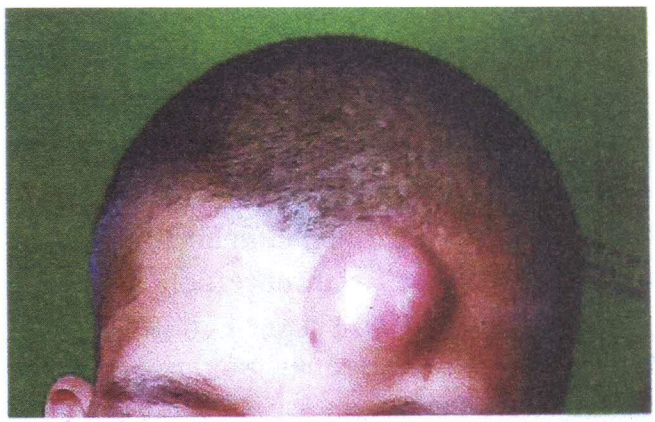

Fig. 1 Photo taken before surgery for the recurrence

DFSP had been first discussed in literature in the 1890s, but Darier and Ferrand first documented it as a distinct cutaneous disease in 1924. In 1925 Hoffman used the term of "dermatofibrosarcoma protuberans" (2).

DFSP accounts for less than 0.1% of all malignancies as well as 1% of all soft tissue sarcomata. DFSP is the most common type of skin sarcoma. The incidence of DFSP is very low and in United Status the incidence of DFSP has been estimated to be 0.85 cases per million population per year (2). It shows a male predominance.

DFSP usually starts as a small, thickening of the skin and ultimately the size varies between one to five cms. The colour can be variable, including the colour of normal skin.

The lump gradually increases in size over months to years (3).

In the early stages, the lesions may clinically resemble benign skin lesions and lead to confusion in diagnosis and delay in treatment. But in advanced stages lesions are ulcerated. Occasionally these tumours contain gelatinous material or altered blood in degenerated cystic areas. Redness and pain occurs in only 15% of cases (3,4).

[1]Department of Surgery, Faculty of Medicine, University of Jaffna. [2]National Cancer Institute, Maharagama. Corrspondence should be addressed to BS(email – srisayan@yahoo.com).

DFSP frequently affects trunk (40 – 60%), followed by the limbs(20 –30%) and the head and neck (10% – 16%) (1).

It has a low incidence of metastasis, either to regional nodes or distant sites. Recurrence occurs in up to 60% of the patients.

Genetic studies in DFSP shows translocation between chromosomes 17 and 22 (1). Microscopic studies show various types such as Bednar tumours, myxoid DFSP, Giant cell fibroblastoma, fibrosarcomatous type.

Molecular techniques like multiplex reverse transcription polymerase chain reaction (RT-PCR) and fluorescence in situ hybridization (FISH) are useful in investigations (4).

Surgery is the definitive treatment and the Mohs micrographic surgery is the treatment of choice for DFSP (1).

Inoperable, recurrent DFSP can be treated with Imatinib, a tyrosine kinase inhibitor and radiation therapy.

Local recurrence of DFSP commonly occurs in first few years, therefore continuous follow up is necessary (1).

References

1. Sanjay BD, Avani DM, James B *et al*. Dermato fibrosarcoma Protuberans, *J lin Aesthet Dermatol* 2008 May;1(1): 34 – 36.
2. Raman KM. Dermatofibrosarcoma Protuberans Presentation, medscape 2016 (cited 2016 April 15). *http://emedicine.medscape.com/*article/ 1100203- clinical.
3. Gloster H. Dermatofibrosarcoma Protuberans. *J Am Acad Dermatol* 1990; 35: 355 – 74.
4. Repertinger S, Teruya B, Sarma D. Common spindle cell malignant neoplasms of the skin; Differential diagnosis and review of the literature. *The internet journal of dermatology* 2008; 7(2).