Cutaneous Malignant Fibrous Histiocytoma

A Rare But Serious Malignancy

JOHN BERTH-JONES,[1] ALAN FLETCHER[2] and ROBIN GRAHAM-BROWN[1]

[1]*Department of Dermatology and* [2]*Department of Histopathology, Leicester Royal Infirmary, Leicester, England*

We report 3 cases of malignant fibrous histiocytoma occurring as primary neoplasms of the skin. The first case developed in a leg ulcer of traumatic origin. The second developed on the lower lip at the site of a squamous cell carcinoma which had been treated by radiotherapy. The third arose on a calf at a site of previous surgery. The literature on this malignancy is reviewed, with emphasis on cutaneous involvement. ***Key words: Sarcoma; Trauma; Radiotherapy.***

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J. Berth-Jones, Department of Dermatology, Leicester Royal Infirmary, Leicester LE1 5WW, England.

Malignant fibrous histiocytoma (MFH) is the commonest soft tissue sarcoma of later life (1). It is a highly malignant neoplasm with a marked tendency to metastasize and also to recur locally following excision. MFH arises in a wide range of tissues but relatively few cases have been reported arising in the skin.

The importance of this tumour lies in the fact that it may be curable if treated early and correctly. Cases seen by dermatologists will often present early. Furthermore these cases will often fall into a group arising superficially and on distal parts of the limbs, which carry the best prognosis.

We report here 3 cases of cutaneous MFH which illustrate predisposing factors of which dermatologists should be aware.

CASE REPORTS

Case 1

A 69-year-old male patient presented in August 1985, had developed an ulcer on the front of the left lower leg. It had been growing slowly for 8 months following a laceration to this area.

Examination revealed an ulcer on the front of the left leg with lumpy irregular margins. The histology of the ulcer margin is described below. Wide excision and skin grafting were performed, followed by radiotherapy. Although the grafted area broke down after radiotherapy and remained ulcerated, there was no evidence of tumour recurrence 4 years later.

Case 2

A 83-year-old male patient, presented in April 1986 complained of a swelling on the lower lip. He had received radiotherapy to this area for a squamous cell carcinoma in 1976.

Examination revealed an ulcerated mass on the lower lip. A biopsy performed on presentation showed only inflamed scar tissue. The ulcer persisted, however, and was excised in August 1986. Histology then showed radionecrosis. By March 1987 there was again a large indurated area of ulceration on the lower lip (Fig. 1). This was excised, allowing a wide margin. The tissue was found to contain MFH as described below. Six months later, biopsy of an enlarged submandibular lymph node revealed metastatic MFH. This area was treated with radiotherapy and he remained alive and well after a further 2 years of follow-up.

Case 3

A 66-year-old female patient presented in April 1987, with a 3-month history of redness and tenderness at the back of her left calf. In 1982 she had undergone excision of a small cutaneous lump from the same site. This had been diagnosed clinically as a sclerosing haemangioma, but the excised tumour had been lost prior to pathological examination.

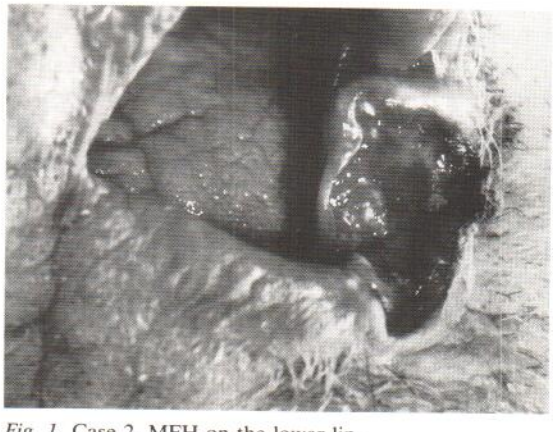

Fig. 1. Case 2. MFH on the lower lip.

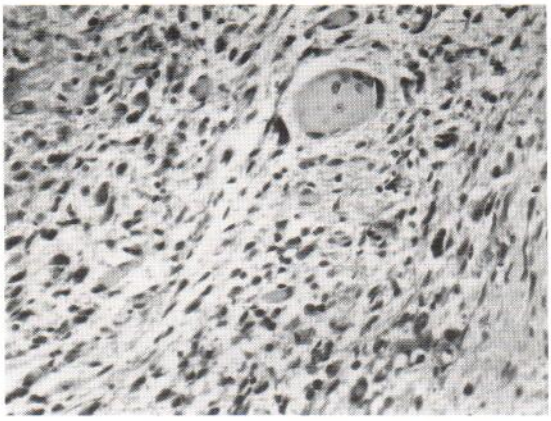

Fig. 2. Case 2. H/E, ×400. Pleomorphic and multinucleate cells of malignant fibrous histiocytoma surrounding an adnexal remnant.

Examination revealed marked induration around the scar from the previous excision over an area measuring 6×4 cm. No lymphadenopathy was present. A biopsy revealed MFH and the histology is discussed below. Investigations revealed no evidence of metastasis. The indurated area was excised, allowing a wide margin and there was no evidence of tumour recurrence 2 years later.

Histology

The three tumours were similar in appearance. Each consisted of pleomorphic spindle cells arranged in a markedly storiform pattern in some areas. Occasional multinucleate cells were present (Fig. 2). Mitotic activity ranged from one to four mitoses per high-power field. In all cases a diagnosis of MFH was made morphologically and with the help of immunocytochemistry; epithelial markers CK1 and CAM 5.2 were negative, all the tumours stained positively for vimentin but not for desmin, S100 or leukocyte common antigen. There was patchy labelling of tumour cells for α_1-antitrypsin and α_1-antichymotrypsin.

The important differential diagnoses of spindle cell squamous cell carcinoma and melanoma were excluded in each case, both morphologically and by immunohistochemistry.

DISCUSSION

Malignant fibrous histiocytoma is a sinister neoplasm. It appears from major series (1, 2) to occur slightly more often in males than in females and most commonly in the 5th to 8th decades. Deep structures (usually muscle), are the commonest sites of origin. Most often it develops on the limbs. MFH may penetrate through to the skin from underlying tissues, but occasionally arises as a primary cutaneous tumour (3–12).

Histopathologically, there are five recognized patterns (13) and more than one may be found in the same tumour. The commonest, to which all our cases conform, is the pleomorphic variant in which plump spindle cells are seen together with giant cells and histiocytes. There may be a storiform pattern, and frequent mitoses are found. The other patterns are myxoid, giant cell, angiomatoid, and inflammatory. Immunohistochemistry demonstrates vimentin, (14) a marker of mesenchymal tumours.

Weiss & Enzinger (1) reported that following excision only 32% of patients appeared cured, 42% developed metastatic disease and 44% local recurrence. The majority of deaths occurred within 2 years. The most significant prognostic factor was the depth of the tumour. Those confined to the subcutis or invading no deeper than fascia were less likely to recur or metastasize than those in deeper structures. However, even tumours confined to the subcutis recurred locally in 30% of cases. In the series of Kearney & Soule (2) there was again an alarming recurrence rate, even in superficial tumours. This series also showed that tumours of the trunk and those proximal to the knees and elbows fared worse than distal lesions, perhaps partly due to the distal tumours being more superficial.

In the treatment of MFH it is therefore essential to perform wide excision or amputation as early as possible. Pre-operative, or post-operative radiotherapy may improve the prognosis (15).

Cutaneous MFH has been reported following trauma (3), and in scars from a vaccination (3) and a burn (7). Other cases appear to be associated with chronic inflammation and have arisen in a chronic ulcer of traumatic origin (8), chronic venous ulcers (12), discoid lupus erythematosus (11) and chronic lymphoedema (10). Radiotherapy is another predisposing factor (9). One of our patients had received radiotherapy and the others had a history of trauma. The malignancy in case 3 probably arose in the surgical scar following excision of the initial lesion 5 years earlier. Whilst it is also possible that this represented a recurrence of a primary tumour excised in the initial procedure, it would seem unlikely that this malignancy would not recur until 5 years following a simple local excision. Our cases therefore provide further evidence of the importance of trauma and radiotherapy as predisposing factors.

Tumours developing in the skin often fall into the distal and superficial groups which carry a relatively good prognosis, and, although the period of fol-

low-up has been short in some reports, a large proportion appear to be curable (3–6, 12). It is therefore important that dermatologists are familiar with this tumour.

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