Malignant fibrous histiocytoma of the breast

A case report

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Summary
Primary malignant fibrous histiocytoma of the breast was diagnosed in a 45-year-old Indian woman. She presented with a large ulcerated lesion and had lung metastases. Treatment was by radiotherapy, toilet mastectomy and chemotherapy. The patient died of rapid extension of lung metastases 15 months after first being seen. Although this tumour has previously been considered to have a relatively good prognosis, this report emphasises its fully malignant metastatic potential.

Malignant fibrous histiocytoma (MFH) is one of the more unusual malignant breast tumours. It is regarded as a common soft-tissue sarcoma of late adult life but few cases of primary breast involvement have been published. Although characterised by bizarre morphology, it is considered a potentially curable condition if local excision is carried out early.

A case of primary breast MFH illustrating the tumour's potential to metastasise is described.

Case report
A 45-year-old Indian woman was admitted to Coronation Hospital on 31 January 1984 with an ulcerated, bleeding tumour of the left breast. Eight months previously she had noticed a breast lump which had grown steadily and had begun to ulcerate 4 months before. Although bleeding from the lesion had prompted her to seek medical attention, she also complained of dizziness, loss of weight and a cough. She was only mildly pyrexial.

Examination revealed a hard lobular mass invading the whole upper outer quadrant of the left breast but not fixed to the pectoralis major. The overlying skin was warm, inflamed, and ulcerated with a serosanguineous discharge. There was no nipple retraction or discharge nor was there palpable axillary or supraclavicular lymphadenopathy. The right breast showed no abnormalities. The patient had 5 children and was receiving contraceptive injections. A familial history of breast disease could not be elicited.

Histopathological examination of a Tru-cut needle biopsy specimen confirmed the malignant nature of the tumour. A biopsy specimen was also submitted for oestrogen and progesterone receptor determinations; both were negative. The haemoglobin level was 13.6 g/dl, leucocyte count 9 300 x 10^9/l and alkaline phosphatase level 68 U/l. Staphylococcus epidermidis, Escherichia coli and Bacteroides vulgarus were cultured from the ulcerated surface of the tumour. Liver, bone and brain scans showed no evidence of metastasis. Radiotherapy to the left breast and draining lymph node areas was given (3 500 rad over a 6-week period), but despite this the ulceration increased. An offensive discharge, bleeding and disabling pain led to a toilet mastectomy on 5 April 1984, 1 week after completion of radiotherapy. The wound was left open to granulate and was successfully covered 2 weeks later by a simple split-thickness skin graft from the thigh.

Histopathological examination of the mastectomy specimen revealed an MFH (Fig. 1 (a) and (b)) characterised by a cartwheel (storiform) pattern of variably shaped spindle cells, numerous atypical mitoses and scattered giant cells containing several hyperchromatic irregular nuclei. Necrosis was very prominent.

At this stage, a coin lesion (Fig. 2) became apparent in the left lower lung lobe. Although radiologically visible at the time of admission, it had been largely masked by the heavy left breast shadow. Chemotherapy was started on 7 May 1984 with adriamycin, cyclophosphamide, methotrexate and vincristine. There was initial improvement, the mastectomy wound healed completely and the patient gained weight. A chest radiograph on 15 July 1984 showed no progression of the pulmonary metastasis, but 3 months later the patient complained of worsening cough and a second lesion was found to have developed at the right lung apex. In early May 1985 she was again admitted to hospital complaining of a disabling cough. There was no evidence of local recurrence or lymph node involvement, but she had become cachectic and a solid nodule had developed in the left lobe of the thyroid gland. Chest radiographs showed diffuse infiltration of both lung fields by metastatic tumour. She was discharged on symptomatic treatment and died at home on 16 May 1985. Permission for autopsy was refused.

Discussion
Sarcoma of the breast is uncommon (less than 1% of malignant mammary lesions) and MFH exceptional. The term MFH was first introduced in 1963 by Ozzello et al. to describe a malignant soft-tissue tumour morphologically characterised by a storiform or cartwheel-like growth pattern. Although the
histogenesis is still the subject of some debate, there is now a clearer understanding of the behaviour of this tumour. Enzinger and Weiss regard it as arising from primitive mesenchymal cells showing partial histiocytic and fibroblastic differentiation. Essentially a tumour found in skeletal muscle, retropertioneum and dermis, it has also been described in the larynx, conjunctiva, vulva, bone and breast. Its malignant character has often been underrated, although more recently several authors have documented cases with distant metastases and fatal outcome. Metastases occur in almost every organ and have been described in thyroid, lungs, diaphragm, heart, pericardium, spine, kidneys, adrenal glands, stomach, large and small bowel, liver, pancreas, spleen, bone, subcutaneous tissue and regional lymph nodes. The incidence may be greater than previously thought. In 1978, Weiss and Enzinger emphasised that MFH was 'the most common sarcoma of late adult life', a fully malignant lesion with a 2-year survival rate of 60%, a recurrence rate of 44%, and a metastatic rate of 42%. Because of a poor sensitivity to radio- and chemotherapy, it has been recommended that this tumour be treated by 'prompt radical surgery'. Local spread beyond the gross tumour mass is common, and must therefore be taken into account when determining the necessary extent of excision. The incidence of metastasis to the regional lymph nodes is believed to be only 12%, and routine dissection of clinically negative lymph nodes does not seem indicated. Tumours of the distal extremities, the most common, have a better prognosis than those of the proximal extremities or the retroperitoneum.

In a review of MFH of the breast, Langham et al. noted 7 previously reported cases and added 1 of their own. In 1984 another 2 cases were reported by Vera-Sempere and Llombart-Bosch and 4 cases were reported from the Memorial Sloan Kettering Cancer Center by Callery et al. in 1985.

The differential diagnosis of malignant histiocytic lesions involving the breast includes dermatofibrosarcoma protuberos (DFP). This primary dermal lesion can invade the breast tissue and be morphologically quite similar to the storiform type of MFH. But for 7 exceptional reported cases, DFP is regarded as an indolent, essentially locally malignant, non-metastasising tumour.

Radiation-induced MFH has been described after irradiation of breast carcinoma, retinoblastoma, Hodgkin's disease and multiple myeloma. Airing in the irradiated field several years after treatment, these lesions are highly malignant with rapid lethal evolution. The prognosis of primary MFH of the breast is not as well documented. Considering it as fairly good, Langham et al. discussed 4 patients whose lesions were treated by simple or radical mastectomy. There was local recurrence but no metastases or deaths (follow-up periods from 11 to 54 months).

The 1 patient of Vera-Sempere and Llombart-Bosch was apparently free of disease 5 years after radical mastectomy. A less favourable prognosis was found among the 5 patients with MFH diagnosed at the Sloan Kettering between 1949 and 1982; Callery et al. reported that they all presented with breast parenchymal lesions and none had previously received irradiation. Two patients died of distant metastases 13 and 22 months after mastectomy (and radiotherapy after local recurrence); 1 patient died of an unrelated cause 23 months after partial mastectomy, and 2 patients were apparently disease-free after 108 and 25 months respectively.

Our patient also illustrates the rapid evolution this tumour may have. Only 24 months elapsed between the awareness of a breast lump and the death of the patient from disseminated disease despite radiotherapy, surgery and chemotherapy. The local control obtained by simple mastectomy with skin graft (after 3500 rad without clinical response) was surprisingly good, but the distal dissemination (in lungs and thyroid) proved lethal. Chemotherapy was unable to control further spread. Although acceleration of the growth rate of MFH has been observed during pregnancy suggesting hormonal dependency, oestrogen and progesterone receptors could not be demonstrated in this tumour. It is therefore very unlikely that hormonal manipulation would have helped.

In summary, it would appear that primary MFH of the breast may not be less malignant than MFH in any other site. In this patient with rapidly evolving disease, pulmonary metastasis was present at the time of diagnosis and, despite a combined approach of radiotherapy, surgery and chemotherapy, the patient died 15 months after first admission to hospital.

Radiotherapy was administered by the Radiotherapy Department, Hillbrow Hospital and the University of the Witwatersrand, Johannesburg. Chemotherapy was administered at Coronation Hospital Chemotherapy Clinic by Mr R. White.

REFERENCES