

## ANGIOMATOID MALIGNANT FIBROUS HISTIOCYTOMA

Z.B.M. NIAZI

*Department of Plastic Surgery,  
Addenbrooke's Hospital, Cambridge.*

### INTRODUCTION

Angiomatoid malignant fibrous histiocytoma is a rare soft tissue tumour affecting young individuals. It is often misdiagnosed even after biopsy. We report a case and present the clinical and morphological features of this peculiar and rare tumour, its treatment and prognosis are also discussed.

### CASE REPORT

A 17-year's women was referred from another hospital in September 1989 with a two and a half years history of a slow growing lump in the left suboccipital region. She had been asymptomatic apart from the lump at presentation in December 1988. Clinically the lump felt like a lipoma but on ultrasound scanning it was found cystic. Biopsy reported it to be an Angiomatoid malignant fibrous histiocytoma. She was also anaemic with a low serum iron, and her haemoglobin improved only slightly with iron therapy.

Computerised tomogram (Fig. 1) reported as "A large soft tissue mass in the posterior lateral aspect of the left side of the neck extending to the occipital bone with loculation within this well circumscribed and no bony erosion" (Fig. 2).

Removal of the 5-cm diameter suboccipital tumour through an S-shaped inci-

sion and excision of the previous biopsy scar was done. Some portion of the trapezius and the rectus capitus muscles were excised along with the tumour as were some of the nodes in the posterior triangle. The lesion was encapsulated and composed of two large cysts and had to be dissected off the occiput, the upper cervical vertebrae and the vertebral arteries. Closure of the wound was obtained without the need for flap reconstruction. She was transfused blood pre and postoperatively. Histological examination of the specimen confirmed the diagnosis of an angiomatoid malignant fibrous histiocytoma that had been completely excised and none of the lymph nodes excised were involved with the tumour.

In the immediate postoperative period the patient was given a soft cervical collar for neck support. She made a normal recovery and was allowed home a week post surgery. Patient remained asymptomatic with no evidence of recurrence on follow-up over the last three years.

### PATHOLOGY AND FEATURES

The histogenetic origin of fibrous histiocytomas is elusive but it is thought to arise from the primitive mesenchymal cells capable of multidirectional differentiation. The malignant fibrous histiocytomas represent 20-30% of soft tissue sarcomas, and

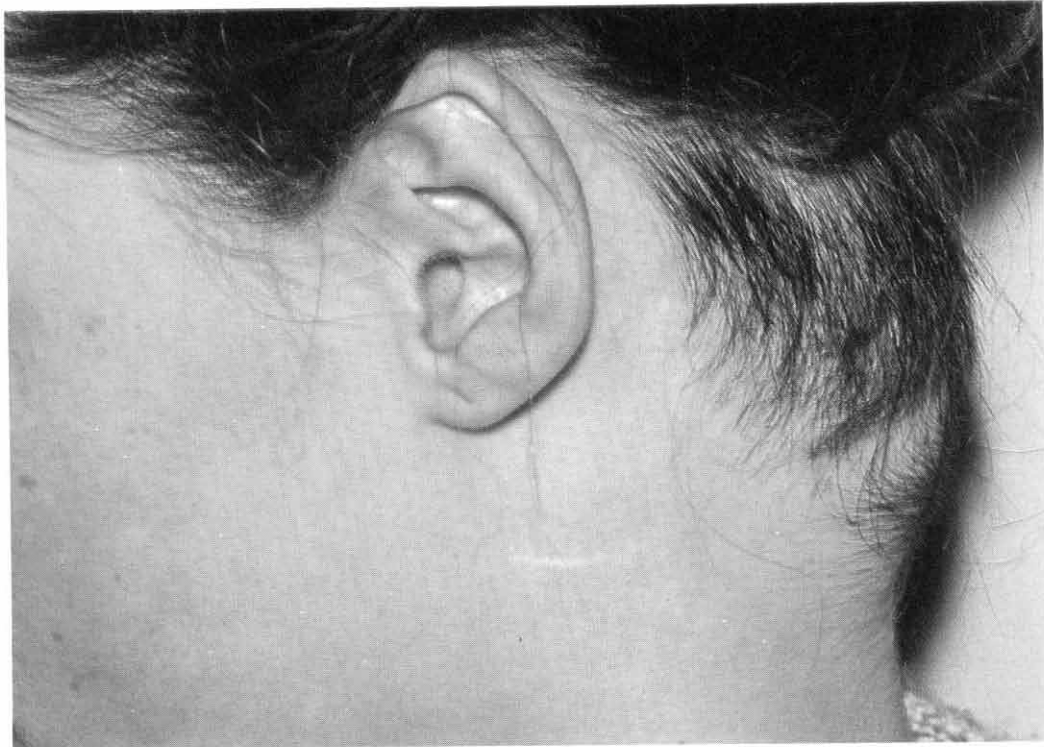


Fig. 1. A lateral view from the left showing the swelling with a biopsy scar in the left occipital region.

occur preponderantly in soft tissues, but also occasionally in bone. The sites of origin of these tumours are 50% in the lower extremities, 20% in the upper extremities, 20% in the abdominal cavity and retroperitoneum, 10% in the head and neck region and bones, lungs, ovaries etc. Soft tissue lesions are multilobulated, grey-white, fleshy, infiltrative, unencapsulated but deceptively circumscribed masses. Sometimes they are soft and translucent owing to an abundant myxoid stroma. Haemorrhages and areas of necrosis are common.

Five histologic variants of malignant fibrous histiocytoma have been described. The most common is called the Storiform-pleomorphic and the next most common is the Myxoid malignant fibrous histiocytoma. The three less common variants are the Giant cell, the Inflammatory and the

Angiomatoid malignant fibrous histiocytomas.

Angiomatoid malignant fibrous histiocytoma usually affects young individuals, mean age being 17, with females being affected slightly more than males;<sup>1</sup> In some of the series sex incidence was the reverse, males were also more often affected than females.<sup>2,3</sup> The site most commonly affected are the extremities (65%) followed by the trunk (28%) and the head and neck (77%). In general the tumours are 2–4 cm in diameter, are painless and located superficially. They grow slowly with an average time of one year before presentation. Most often the clinical diagnosis is that of a benign cyst. The angiomatoid lesions may have a bluish tint and clinically resemble a cutaneous haemangioma or a haematoma. The incidence of constitutional symptoms

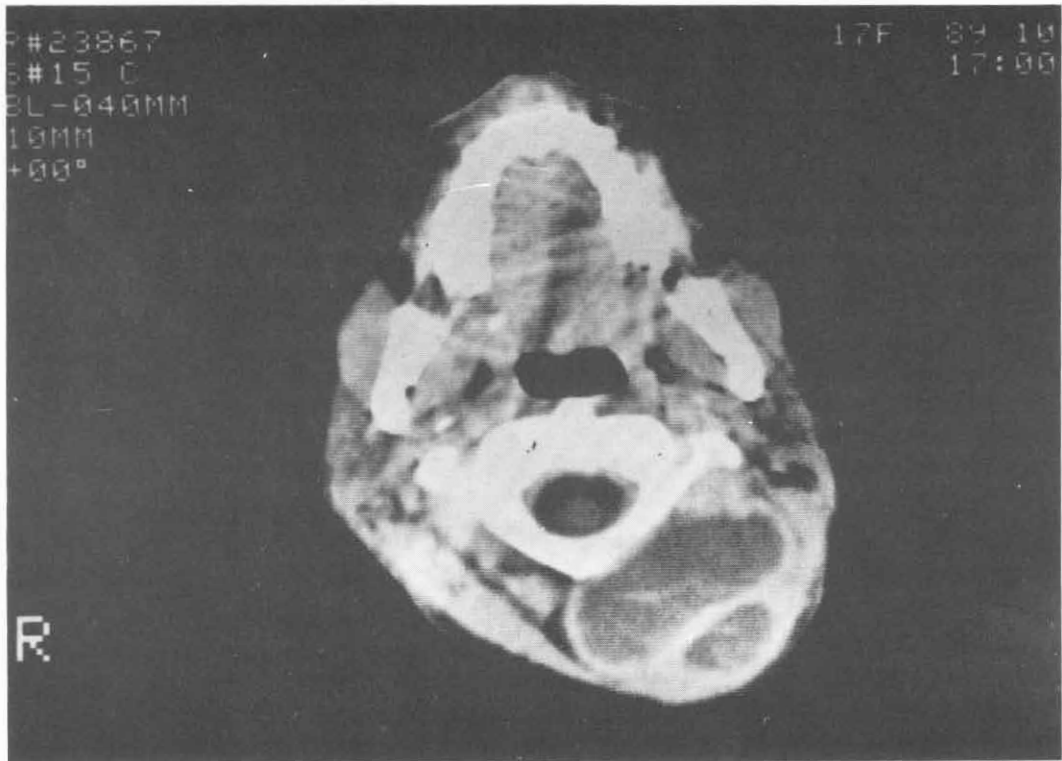


Fig. 2. A CT scan showing a large, well circumscribed, cystic multilobulated mass in the left occipital region with no evidence of bony erosion.

like anaemia, weight loss and fever vary significantly between the two major series.<sup>1,2</sup>

## DISCUSSION

Angiomatoid malignant fibrous histiocytoma is a rare soft tissue tumour, commonly affecting the young. It is usually misdiagnosed. With early diagnosis and wide local excision most of the patients now have a chance to survive. In 1979 Enzinger reported on 41 cases studied retrospectively and the tumour was classed as low grade malignant fibrous histiocytoma with incidence of recurrence in 63%, metastases in 20% and mortality in 12%. In the series of 20 cases reported by Pettinato et al<sup>3</sup>, the local recurrence rate was reported as 25% and a mortality of 5%.

In another recent series of 108 patients with a 10 years follow-up reported by Costa and Weiss, local recurrence developed in only 12% of patients, all of whom were cured by reexcision. Five patients developed metastasis, four of which were local (a second nodule developing in the region or extremity but not in the operative field). All the local metastasises underwent wide local excision with no further problems. The one with distant metastasis presented late with a large tumour and was completely excised but developed lung metastasis 15-months and cerebral metastasis 19-months after excision. She did not respond to chemotherapy or radiotherapy and died 26-months after surgery. Costa and Weiss<sup>1</sup> reaffirm the entity of angiomatoid malignant fibrous histiocytoma but indicate that the lesion appears

such a low grade neoplasm that assessment of various histologic parameters or grading does not provide any information. They also feel that angiomatoid malignant fibrous histiocytoma should be reclassified with fibrohistiocytic tumour of intermediate malignancy rather than with the conventional malignant fibrous histiocytoma, most of which are high grade sarcomas.

## REFERENCES

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