Short reports

Primary malignant fibrous histiocytoma of the mediastinum

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Abstract

A malignant fibrous histiocytoma of the mediastinum was successfully treated by surgery and radiotherapy. Magnetic resonance imaging was able to show the site of invasion, the two different components of the tumour, and the blood supply.

Malignant fibrous histiocytoma is one of the most common forms of soft tissue sarcoma in older people, arising most frequently in the extremities and retroperitoneum.¹ We present a case of malignant fibrous histiocytoma of the mediastinum.

Case report

A 47 year old woman presented with mild pain in the right upper chest and back and right shoulder, a dry cough, and slight exertional dyspnoea. A systolic murmur was heard over the right upper thorax posteriorly. Conventional radiography and tomography revealed a large, round soft tissue mass in the right upper chest of possible pulmonary or mediastinal origin; computed tomography showed a tumour 11 × 9 cm adherent to the superior mediastinum, but not invading the chest wall. Bronchoscopy showed the dorsal segmental bronchus to be compressed, but the mucosa was normal. A brush biopsy was unhelpful, and a fine needle aspirate consisted only of blood. Intra-arterial digital subtraction angiography showed a hypervascular tumour supplied by the subclavian artery and its branches. Magnetic resonance imaging with a special surface coil to enhance image quality² suggested that it was vascularised by intercostal vessels and that the tumour originated in the right paravertebral region at the level of the third to fifth vertebral bodies (fig 1). The tumour consisted of a solid cranial component with central necrosis, separated from a cystic caudal part by a septum.

At thoracotomy a vascular tumour supplied by intercostal vessels was found in the upper posterior mediastinum, infiltrating the paravertebral region and adherent to the right

upper lobe. Its cranial part was solid with central necrosis; the caudal part was better defined, and had a lucent appearance. Frozen section studies suggested a malignant mesenchymal neoplasm. The tumour was excised completely after ligation of large capsular vessels. The postoperative course was uneventful.

On subsequent histological examination the tumour proved to be a malignant fibrous histiocytoma with a polymorphic-storiform and myxoid pattern (fig 2), containing fibroblastic cells immunoreactive for vimentin. In view of this diagnosis radiotherapy was instituted six weeks after surgery, with 50 Gy in 25 fractions to the tumour site and 14 Gy to the posterior chest wall and paravertebral region over 12 weeks. When last seen 24 months after surgery the patient was well.

Discussion

Malignant fibrous histiocytoma has a wide morphological and cytological spectrum, and is characterised by a mixture of elements resembling histiocytes and fibroblasts, often arranged in a storiform pattern and accompanied by giant cells and inflammatory cells.³ Because of the diverse appearance of this tumour, a combination of electron microscopy and immunohistochemistry may be necessary to distinguish it from anaplastic carcinoma and other types of sarcoma. Although it frequently metastasises to the lungs, an intrathoracic origin is rare. Thirty nine cases arising in the lung have been reported,⁴ but only eight in the mediastinum.^{3 5-10}

The clinical manifestations, treatment, and

Figure 1 Magnetic resonance image showing tumour invasion in the paravertebral region (white arrows). The tumour consists of two parts: the cranial part has a lower signal intensity at the periphery representing solid tissue, and a high signal intensity at the centre (*) representing necrosis; whereas the caudal part has a homogenous high signal intensity reflecting a more fluid like consistency. There are also vessels visible (arrow head).

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Figure 2 Malignant fibrous histiocytoma, showing a storiform pattern.

subsequent course of these latter patients have all been different.⁵⁻¹⁰ Preoperative computed tomography and open lung biopsy have been recommended in the planning of radical resection.⁹ In our case, however, magnetic resonance imaging had advantages over other diagnostic procedures in that it showed the site of invasion in the paravertebral region, the two different

components of the tumour, and its blood supply.

At present there is no consensus about the treatment of choice for patients with mediastinal malignant fibrous histiocytoma. The eight patients reported underwent radiotherapy, surgery, chemotherapy, or a combination of these. The high incidence of local recurrence and distant metastases associated with this tumour would support the case for initial radical en bloc excision. Further data are needed to establish the optimal management.

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