

Malignant mesenchymoma of the chest wall in an adult

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Abstract

Mesenchymoma (hamartoma) of the chest wall is an extremely rare tumour presenting in early infancy or fetal life. Pleural, pulmonary, and lymph node metastases developed in a young man with malignant mesenchymoma of the chest wall. The tumour had several characteristics that differ from the mesenchymoma reported from the other parts of the body.

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Mesenchymoma (hamartoma) of the thoracic wall is a rare entity presenting in infancy or fetal life.¹⁻⁴ Its malignant counterpart is exceedingly rare.⁵ Mesenchymoma of chest wall, benign or malignant, has not been described in an adult. We report a case of malignant mesenchymoma of the chest wall in a young adult with pulmonary, pleural, and axillary lymph node metastases.

Case report

A 20 year old man presented in January 1990 with a history of progressive, slow enlargement of a painless lump in the right pectoral region over two years. There was no swelling elsewhere or any constitutional symptoms. A firm, non-tender, freely mobile, 1.5 cm lymph node was palpated in the right axilla. The presenting mass was hard, immobile, non-tender, measuring 8 × 12 × 15 cm, and the overlying skin and nipple were freely mobile. Breath sounds were diminished in the right pectoral and infra-axillary regions. The rest of the systemic examination showed nothing abnormal. There was no evidence of von Recklinghausen's disease.

The results of routine blood biochemistry investigations were within normal limits. Urine gave a negative result for vanillyl-mandelic acid. A chest radiograph showed a huge opacity on right side with well circumscribed margins and a shifted mediastinum. Computed tomography of the chest showed a solid, non-enhancing tumour crossing the midline, containing areas of degeneration and focal, scattered calcification (fig 1). The third and fourth ribs were destroyed anteriorly and adjacent lung tissue was atelectatic. There was a moderate right pleural effusion. An ultrasound scan of the abdomen was normal. A total body scan with technetium-99m

showed increased radioactivity in the region of right third and fourth ribs. The cytological appearance of a fine needle aspirate was inconclusive but a Tru-Cut needle biopsy specimen was reported as showing a low grade fibrosarcoma.

Operative findings

The tumour was invading the pectoral muscles. From the pectoralis major a 10 cm long, shiny, pearl white structure was extracted. The tumour was widely excised, with the lower halves of pectoral muscles, the anterior halves of the second to fifth ribs, and the right half of the body of the sternum taken en bloc. About 700 ml of serous, exudative, non-malignant effusion was evacuated. The lower reaches of the parietal and diaphragmatic pleura contained many nodules of 1-5 cm. The lateral basal segment of the right lower lobe contained two nodules 1.5 cm each that were wedge resected. No hilar or mediastinal lymphadenopathy was noted and the rest of the lung was normal on palpation. The chest wall was reconstructed with Marlex mesh and free split rib grafts. The patient was ventilated for 48 hours and made an uneventful recovery. The pleural effusion had not recurred at follow up six months later.

Pathological findings

The tumour measured 12 × 15 × 18 cm and weighed 2.5 kg. The cut surface was greyish white in the centre and surrounded by a jelly like periphery. Microscopically it showed collagenous stroma focally hyalinised with spindle shaped malignant cells and myxoid tissue. There was abundant osteoid and chondroid formation and some areas contained adipose and rhabdoid tissue (fig 2). The axillary lymph node showed similar tumour cells but with a predominantly neural element, mimicking a schwannoma. Nodules from the lung and the pleura showed cells similar to primary tumour. Immunohistochemically the primary tumour was negative for desmin, myoglobin, S-100, and cytokeratin but was strongly positive for



Figure 1 Computed tomogram of the chest showing extent of the tumour, focal degeneration, and scattered calcification. Densities measured at 1, 2, and 3 are 37.1, 20.1, and 24.1 respectively.

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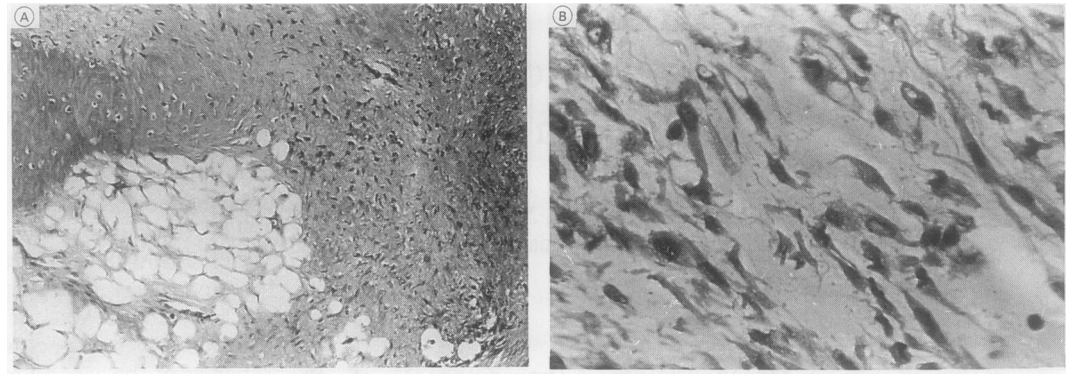
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Figure 2
Photomicrograph showing different elements:
(A) chondromatous and lipomatous tissue in malignant fibrous stroma;
(B) differentiation into early rhabdoid tissue.
(Haematoxylin and eosin).



vimentin, which confirmed its mesenchymal nature.

Discussion

Mesenchymoma was defined by Stout⁶ as a tumour composed of two or more unrelated mesenchymal derivatives apart from fibrous tissue. These tumours are thought to arise from embryonic mesenchyme that have the potential to differentiate into any type of connective tissue.

Blumenthal *et al*⁵ reported malignant mesenchymoma of the chest wall in infants but the diagnosis was contested by Brand *et al*.¹ Even benign mesenchymoma of the chest wall is extremely rare. In a recent review only 21 cases were found to have been reported worldwide, all in fetuses or infants.⁷ Our case is unique in many respects, such as late presentation in young adult life, malignant nature, metastases in different sites and a composite histological picture. The tumour had spread by local implantation of pleural seedlings by lymphatic permeation to an ipsilateral axillary lymph node, and by blood-borne metastases to the right lower lobe. The pleural metastases were less cellular with more myxoid tissue and less collagenous stroma. The pulmonary metastases were more cellular and less myxoid and looked more like the primary tumour. In the axillary lymph node the major element was neural, which substantiated the totipotent nature of the cell of origin. In a similar case a pulmonary metastasis from a malignant mesenchymoma of the right thigh was only chondromatous.⁸ Being totipotent every constituent element of a malignant mesenchymoma can grow as a

metastasis. By comparison with mesenchymomas of other parts of the body^{2,8} this tumour was poorly differentiated into rhabdomyoblastic tissue—hence its desmin negativity. Desmin positivity of rhabdomyosarcoma depends on the degree of differentiation and can be as low as 32%.⁹ Adipose tissue was more scanty than in a mesenchymoma of the head and neck region, where adipose tissue and smooth muscle are often present.¹⁰ Furthermore, elements of aneurysmal bone cyst like formation were not seen in our tumour, in contrast to benign mesenchymomas of the chest wall.¹⁻³

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