

Intrathoracic desmoids: report of two cases

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Extra-abdominal desmoids form part of a group of conditions known as the fibromatoses.¹ Other members of the group include plantar and palmar fibromatosis, Peyronie's disease, and abdominal and intra-abdominal desmoids. All are characterised by fibroblastic proliferation, which is locally invasive and may recur after treatment but never metastasises. Symptoms arise as a result of contraction, constriction, and local invasion.

The most common sites for extra-abdominal desmoids, which are sometimes known as aggressive fibromatoses,² are the shoulder, chest wall, and thigh. Very rarely intrathoracic lesions have been reported.³ We describe two cases of desmoids arising in the chest after thoracic surgery.

Case reports

CASE 1

A 19 year old caucasian man underwent Heller's cardiomyotomy for achalasia via a left 8th interspace thoracotomy in December 1982. He made an uncomplicated recovery.

He returned 32 months later with increasing dyspnoea, productive cough, and pleuritic chest pain. Four months previously he had noticed enlargement of a keloid scar on the left shoulder from a childhood vaccination, and his thoracotomy scar was now undergoing the same change. On examination there were nodular keloids of both vaccination and thoracotomy scars. The trachea had deviated to the right and there was stoney dullness to percussion over the left hemithorax with absence of breath sounds. Abdominal palpation revealed a left subcostal mass. A ranging radiograph obtained as a preliminary to computed tomography (figure) showed that the left chest was filled with a homogenous mass everting the left hemidiaphragm and pushing spleen and stomach downwards and medially.

The thoracotomy scar was excised, exposing a plaque of abnormal fibrotic tissue that enveloped the ribs above and below. Sections of three ribs were removed in continuity, multiple vascular adhesions to the chest wall were divided, and a segment of adherent diaphragm at the hiatus was excised en bloc.

Pathological examination revealed a lobulated fibrous mass weighing 5.2 kg, comprised of uniform spindle cells, with elongated nuclei, arranged in poorly defined bundles separated by an abundant mucoid stroma. Review of the sections by Dr F M Enzinger of the US Armed Forces Institute of Pathology confirmed a diagnosis of extra-abdominal desmoid.

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Accepted 20 May 1986

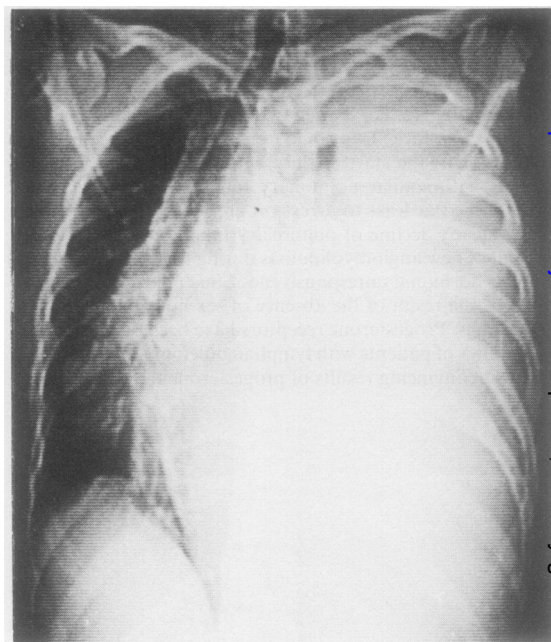
The patient remains well six months after operation, with no evidence of local recurrence.

CASE 2

An 18 year old man had been operated on in Turkey in August 1982 for dysphagia and regurgitation, via a left thoracotomy. In June 1984 he was seen at a London hospital with recurrence of symptoms. A radiograph showed a mediastinal mass.

At operation the patient was found to have a mass involving the mediastinum and right lung, with oesophageal constriction and spread into the diaphragm. Right pneumonectomy and clearance of fibrotic tissue from the mediastinum were carried out via bilateral thoracotomies. Histological examination confirmed a diagnosis of extra-abdominal desmoid.

A chest radiograph four weeks after operation was unremarkable but by nine weeks a mass 1.5 cm in diameter was visible at the left hilum. By 14 weeks it had grown to 5 cm



Desmoid tumour filling the left chest and arising from the site of previous left thoracotomy; the left lung is compressed up into the apex.

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in diameter. Six weeks later the patient returned to Britain with severe dysphagia and increasing respiratory distress. Chest radiography and computed tomography showed a 9 × 9 cm mass affecting the left hilum.

Surgical clearance was not feasible. Palliative radiotherapy (20 Gy (2000 rads) in eight fractions) and a trial of non-steroidal anti-inflammatory drugs (indomethacin 25 mg thrice daily) failed to reduce the size of the mass or relieve symptoms. He returned to Turkey with a feeding gastrostomy in situ.

Discussion

Trauma has been suggested as a factor in the development of desmoids of the abdominal wall, perhaps related to muscle stretching in pregnancy, and extrathoracic desmoids have occurred after mastectomy and after resection of a neurofibroma.³ A single case has been reported of an intrathoracic desmoid arising at the site of a previous thoracotomy⁴ and another was related to rib fractures.⁵ In these, as in our two cases, the lesion presented within three years of the initial trauma.

Recurrence rates of 60% are reported for extra-abdominal desmoids.² The rarity of intrathoracic lesions makes assessment of the natural history difficult, but both of the patients mentioned above developed a recurrence, as did our second patient. Local recurrence is best avoided by primary excision well beyond the visible extent of tumour.³ In our cases radical excision was limited because the masses affected thoracic wall or mediastinal structures.

Desmoids do not metastasise¹ and control by five or more resections has been reported, with very successful prolongation of life.² Radiotherapy has been claimed to control recurrences in up to 80% of patients,⁶ but there is little experience with lesions in the thorax, where the lung's tolerance to irradiation may be a limiting factor. In a recent

study⁷ non-steroidal anti-inflammatory drugs, with ascorbic acid or tamoxifen in some instances, inhibited growth of desmoids in six of seven patients, including one with an intrathoracic lesion.

At present surgery remains the first line of treatment. Radical resection and reconstructive procedures, as suggested for desmoids of the thoracic wall,⁸ may not be technically feasible for intrathoracic lesions and repeated local excision may be required for recurrences. The rapidity of growth seen in our patients dictates that frequent follow up visits should be maintained to permit early, effective treatment.

We are most grateful to Professor J H Wyllie, Academic Unit of Surgery, Whittington Hospital, and to Mr S C Lennox, Cardiothoracic Surgery Unit, Brompton Hospital, for permission to report on their patients.

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