

sitive analysis of intrathoracic major airways obstruction and should be mandatory investigations in patients with large mediastinal cysts. Abnormalities in these values may indicate appreciable obstruction before the onset of symptoms and thus lead to early intervention.

Cytotoxic chemotherapy is not standard treatment for bronchogenic cysts, though this patient had previously had dramatic relief of airways obstruction when given chemotherapy for a misdiagnosed lymphoma. Cell turnover and secretory activity may have been inhibited to such an extent that the volume of the cyst was reduced below a critical level. Instillation of sclerosant agents has been used for hepatic cysts<sup>7</sup> and recurrent swelling of thyroid cysts,<sup>8</sup> but we believe that this is the first time a sclerosant has been instilled percutaneously into a bronchogenic cyst. Although follow up here has been only for six months, this case suggests that instillation of bleomycin and alcohol may have a role in controlling recurrent bronchogenic cysts that cannot be surgically removed.

### Addendum

The latest follow up, 16 months after instillation of bleomycin and alcohol, showed no change in the size of the residual cyst on the computed tomogram and lung function remained normal.

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## Treatment of desmoid tumours in Gardner's syndrome

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### Abstract

**A 24 year old woman with Gardner's syndrome developed a massive chest wall desmoid tumour, which required radical excision and prosthetic reconstruction. In view of the local aggressiveness of this tumour and the fact that it does not metastasise a policy of radical surgery when possible is recommended.**

Desmoid tumours are rare, accounting for 0.1% of all tumours, with an 8:1 female predominance. The overall annual incidence is about 2.4/1 m population, but in Gardner's syndrome the incidence ranges from 3.5% to 17%. Gardner's syndrome, first described in 1953,<sup>1</sup> consists of adenomatous polyps of the gastrointestinal tract, osteomas, epidermoid cysts, lipomas, dental abnormalities, and periampullary carcinomas as well as desmoid tumours. The incidence of the syndrome is 1:14 025 with an equal sex distribution. It appears to be determined by the same gene as familial polyposis coli, Gardner's syndrome representing a more aggressive phenotypic

expression. The gene is inherited as an autosomal dominant. We report a case of a desmoid tumour in a young woman with a family history of Gardner's syndrome and discuss treatment.

### Case report

A 24 year old woman presented with a six month history of a rapidly enlarging mass related to a surgical scar beneath her left breast. A large left atrial appendage had been removed through a left posterolateral thoracotomy five years earlier. She had never been pregnant. Her mother had had two desmoid tumours excised from the anterior abdominal wall and had undergone colectomy for carcinoma of the colon complicating polyposis coli. Her maternal grandmother had died of carcinoma of the colon and her brother had died several years earlier after the removal of a 3 kg tumour from his arm.

Physical examination showed a fit young woman with a 20 cm diameter firm, fixed mass related to the anterior aspect of a thoracotomy wound and inferior to her breast, displacing it upwards and medially. She also had a 3 × 4 cm firm, modle mass behind her right ear which had the clinical characteristics of an epidermoid cyst. Physical examination, including sigmoidoscopy and ophthalmoscopy, otherwise showed no abnormalities.

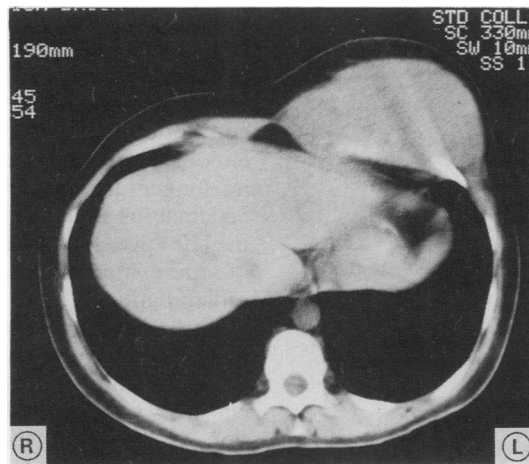
Full blood count, electrolyte concentrations, and results of liver function tests were normal. A barium enema performed earlier that year also showed nothing abnormal. Computed tomography showed a very large soft tissue mass on the left lower anterior chest wall, destroying ribs and invading the left rectus muscle (figure). There was no

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Computed tomogram showing the desmoid tumour on the left lower anterior chest wall.

evidence of metastases in the mediastinum, liver, or lungs.

At operation the tumour extended from the left rectus muscle into the previous thoracotomy scar; the tumour was radically excised with part of the chest wall. The resected specimen measured 30 × 20 × 9 cm and weighed 2.3 kg. The defect in the chest wall was filled by a prosthesis composed of methylmethacrylate cement sandwiched between two layers of Marlex mesh. The remaining abdominal defect was closed by using the contralateral rectus muscle as a pedicled flap to allow primary closure.

Her postoperative course was complicated by an area of flap necrosis, infection, and wound dehiscence beneath the left breast, exposing the chest wall prosthesis. Cover of the prosthesis was achieved with an omental graft, which in turn was grafted with split skin. Her recovery thereafter was uneventful and she was discharged home two weeks later.

Histological examination showed the tumour to consist of spindle cells with small, elongated nuclei and small nucleoli infiltrating between fat and the fibres of the surrounding skeletal muscle. The appearances were consistent with a desmoid tumour.

When the patient was last seen, 12 months after the operation, there was no evidence of tumour recurrence.

### Discussion

The term desmoid, derived from the Greek *desmos* (band or bond), was first used by Johannes Müller in 1838<sup>2</sup> because of the tendon or bandlike appearance of the tumours, which arise from fascial or musculoaponeurotic structures. In a recent series of 89 patients<sup>3</sup> the most common sites for a desmoid tumour were in the abdomen (49%), with extra-abdominal sites accounting for 43%,

and the mesentery for 8%. Seven patients had multiple tumours. They do not metastasise but are locally invasive; the incidence of recurrence after surgery is up to 60%.<sup>4</sup>

Trauma, particularly surgical trauma, has been implicated as an important aetiological factor<sup>3,5,6</sup> in genetically susceptible individuals, but the growth of the tumour seems to be modulated by other factors, especially the female sex hormones and oestrogen in particular. This explains why desmoid tumours are most common in women of childbearing age and grow most rapidly in this group.

Although there is widespread agreement in published reports that surgery should be the principle therapeutic manoeuvre, there is disagreement over the extent of the resection margin. Most authors, however, believe that excision should be as radical as possible but that vital structures should be preserved. Local recurrence should be treated by simple excision.

The role of radiotherapy is not clear. Most reports suggest that desmoid tumours are relatively radioinsensitive and, although radiotherapy may provide local control of non-resectable or recurrent tumours,<sup>7-9</sup> the success in such cases might have been due to ovarian ablation and the consequent reduction in female sex hormone production.

The use of oestrogen antagonists, testosterone, progesterone, and prednisolone has been disappointing. Non-steroidal anti-inflammatory drugs and ascorbic acid (which lower cyclic adenosine monophosphate in tumour cells and inhibit ornithine decarboxylase, an enzyme associated with tumour cell proliferation<sup>10</sup>) have also failed to control the size of tumours consistently or, when used as adjuvants, to prevent recurrence.

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