

be very useful in case 2), its structural rigidity and, in case of malignant disease, resistance to infiltrative growth by tumour. Disadvantages include a greater tendency to displace, and the risk of obstruction by dried airway secretions. Gianturco Z stents do not move and probably do not induce drying of secretions; furthermore, they allow airflow through the stent itself – for example, when placed over the right upper lobe bronchus. Both types of stents probably could have been used in our patients. We preferred the silicone Dumon stents because of their structural rigidity, the ease by which they can be removed, and our greater experience with them.

We conclude that an endoscopically inserted silicone endoprosthesis may be a valid alternative to surgery for tracheal obstruction

due to intrathoracic goitre in patients in whom surgery represents an unacceptable risk.

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## Massive haemothorax secondary to angiosarcoma

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### Abstract

**A patient who presented with recurrent haemoptyses was found to have an angiosarcoma of the adrenal gland which disseminated throughout the pleural space.**

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### Case report

A 44 year old man presented with daily haemoptysis for one week, but was otherwise well. He had smoked 20 cigarettes per day for 20 years and had not been exposed to asbestos, arsenic, thorotrast, vinyl chloride, or radiotherapy. Examination was unremarkable, with no finger clubbing, a clear chest, and no carotid or vertebral bruits. Chest radiography and fiberoptic bronchoscopy were normal. He continued to have haemoptysis and four weeks later he was admitted as an emergency, acutely short of breath. A chest radiograph now showed large bilateral pleural effusions. Aspiration proved these to be haemothoraces. His haemoglobin concentration was 6.0 g/dl. He was transferred to the regional cardiothoracic centre and four and one litres of blood were drained via left and right chest drains respectively. A computed tomographic (CT) scan of the thorax demonstrated the haemothoraces but was otherwise normal. However, a CT scan of the abdomen revealed a left adrenal mass measuring 5 × 4 × 4 cm (fig 1).

A further 20 litres of blood were drained over the following five days. Clotting screens were normal and the adrenal tumour appeared to be non-functioning (urinary levels of 17-ketosteroids and vanillylmandelic acid were normal). Pulmonary angiography and selective angiography of the intercostal arteries failed to demonstrate a definite bleeding point.

On the sixth day adrenalectomy and exploratory thoracotomy were performed. The adrenal mass was a well circumscribed cystic lesion filled with organising blood clot. Two litres of blood were removed from the left hemithorax to reveal visceral and parietal pleura covered in organising blood clot and studded with multiple haemorrhagic cysts 5–30 mm in diameter. The lung parenchyma appeared to be normal. Frozen sections of the lesions were reported as angiosarcoma. Bleeding was controlled and the chest closed.

Postoperatively the patient had a dense right sided hemiplegia. However, he remained stable

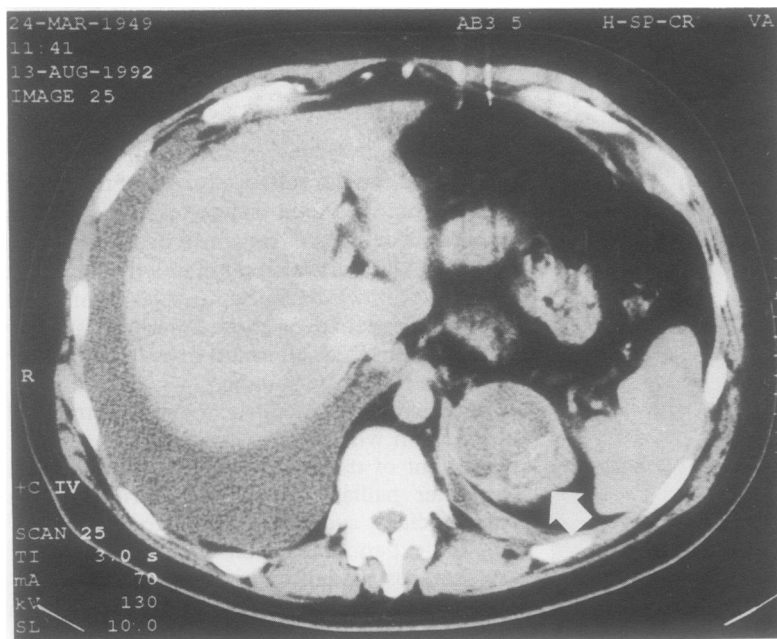


Figure 1 Computed tomographic scan of the abdomen showing a large mass in the left adrenal gland and right sided haemothorax.

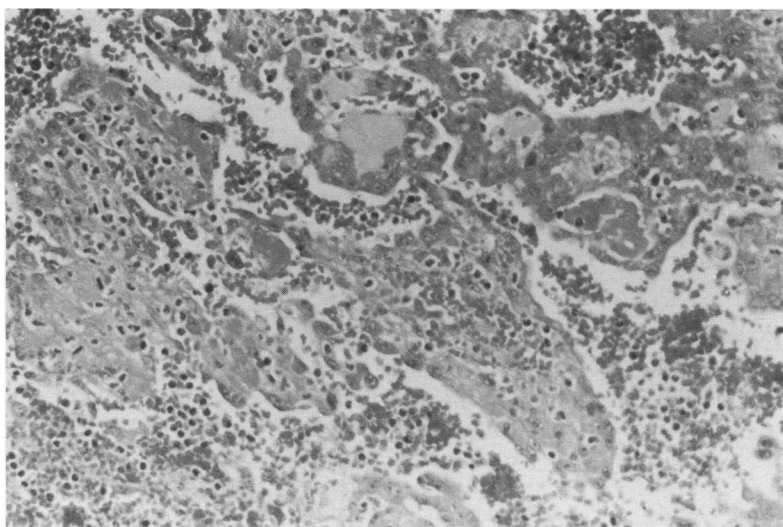


Figure 2 Photomicrograph of section through the adrenal tumour. The typical appearances of an angiosarcoma are demonstrated with vascular channels lined by pleomorphic cells that are either spindle shaped or epithelioid forming papillae and cords. Stain: haematoxylin and eosin. Original magnification  $\times 100$  reduced to 66% on origination.

until the third postoperative day when the bleeding recommenced. Despite administration of tranexamic acid and transfusions of blood, platelets and fresh frozen plasma he continued to bleed. Given the prognosis of the malignancy and the neurological deficit, active treatment was discontinued and the patient died on the tenth postoperative day.

#### **PATHOLOGY**

A full post mortem examination was performed. Multiple cysts found on the pleura of both lungs consisted of loose blood clot surrounded by a rim of grey tissue. The adrenal mass consisted of organising clot and necrotic tissue surrounded by viable adrenal tissue. Histologically, both adrenal and pleural lesions exhibited the typical appearance of angiosarcoma (fig 2). Along with the classic appearance, positive staining for Factor VIII related antigen and *Ulex europaeus* agglutinin I confirmed the diagnosis of angiosarcoma.

The lung parenchyma was normal and no other deposits of tumour were found. The brain was found to have multiple infarcts that were thought to have been secondary to extreme fluctuations in blood pressure perioperatively.

#### **Discussion**

Angiosarcoma is a rare tumour that occurs in skin, bone, breast, and visceral organs, especially the liver. It is associated with exposure to vinyl chloride, thorotrast, arsenic, radiotherapy, and to chronic post mastectomy lymphoedema.

Primary angiosarcomas of the adrenal gland or lung are exceptionally rare with only two convincing reported cases of primary adrenal<sup>1,2</sup> and three of lung origin.<sup>3-5</sup> There are further reports of adrenal involvement in disseminated disease<sup>6</sup> and angiosarcomas frequently metastasise to the lungs.

In the case described here it is unclear where the tumour originated. The single large adrenal tumour with multiple pleural deposits tends to suggest an adrenal primary. Indeed, the pattern of multiple small haemorrhagic nodules studing the pleura is common in metastatic disease. However, it is also described in two of the three cases of primary lung disease.<sup>3,4</sup>

The histological diagnosis of angiosarcoma is often difficult. The characteristic features are well described by Kareti *et al.*<sup>1</sup> Demonstration of the endothelial markers Factor VIII related antigen and *Ulex europaeus* agglutinin I strongly support the diagnosis. Thrombomodulin is said to be a more sensitive and specific marker.<sup>7</sup>

The prognosis of angiosarcoma is poor. Forty cases of cardiac angiosarcoma had a mean survival of nine months.<sup>6</sup> Cutaneous angiosarcoma has a better prognosis with one series reporting a five year survival of 12%.<sup>8</sup> In isolated cases prolonged survival has been reported following radical surgery,<sup>1</sup> radical radiotherapy,<sup>8</sup> and a combination of surgery, radiotherapy and chemotherapy.<sup>6</sup> Interferon alpha-2a induced regression in a child.<sup>9</sup>

Death is often related to pulmonary involvement. In a series of six patients with cardiac angiosarcoma five died as a consequence of lung metastases.<sup>6</sup> Lung deposits may cause respiratory failure by a combination of local infiltration, intrapulmonary, and intrapleural haemorrhage.<sup>3,5</sup> In the case reports where patients have presented with respiratory symptoms, survival has been only a matter of weeks.<sup>3-5</sup>

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