Thorax, 1981, **36**, 66–68

Short reports

Intrathoracic chemodectoma with noradrenaline secretion

BISHAN K NIGAM, STEPHEN L HYER, EVELYN J TAYLOR, AND TUPAS GUHA

From the Department of Cardiothoracic Surgery, Walsgrave Hospital, Coventry, West Midlands

Tumours arising in the chemoreceptor system are termed chemodectomas. Intrathoracic chemodectoma is a rare tumour, only 74 cases having been described by 1977.[1] Systemic catecholamine secretion by an intrathoracic chemodectoma is very uncommon and we have been able to trace only one other published case report of proven catecholamine secretion. We describe a second case and discuss briefly these interesting tumours.

Case report

A 38-year-old housewife was referred to the cardiac unit for investigation of hypertension which had persisted since her first pregnancy three years previously. She was a non-smoker and was not taking the contraceptive pill. At presentation the blood pressure was 160/110 mm Hg. There was no radio-femoral delay or renal bruits and both optic fundi were normal. The chest radiograph and tomograms showed a lobulated mass in the paravertebral region at the level of T6/T7 (fig 1 a, b) which appeared highly vascular at aortography. Computerised axial tomography was performed to define the relationship to the vertebral bodies, and this revealed that the mass entered the body of the vertebra of T6 (fig 2).

Two 24-hour urine collections were analysed for urinary catecholamines and showed grossly raised normetadrenaline (NMA): 3323 μg/24 hours and 2802 μg/24 hours (NR: 204-585). Urine homovanillic acid (HVA) and metadrenaline (MA) excretions were normal.

Operation was performed and the arterial pressure and ECG were monitored continuously. Blood pressure was controlled by an intravenous infusion of phentolamine 1 mg/ml at 3 mg/min, supplemented by intravenous boluses of 2 mg propranolol, especially when the tumour was being manipulated. After removal of the tumour there was a fall of the systolic blood pressure to 80 mm Hg which responded to blood transfusion, and thereafter the blood pressure remained stable.

At thoracotomy a mass 6 × 3·5 cm was lying along the sympathetic chain. The mass was stripped from the body of the sixth dorsal vertebra revealing

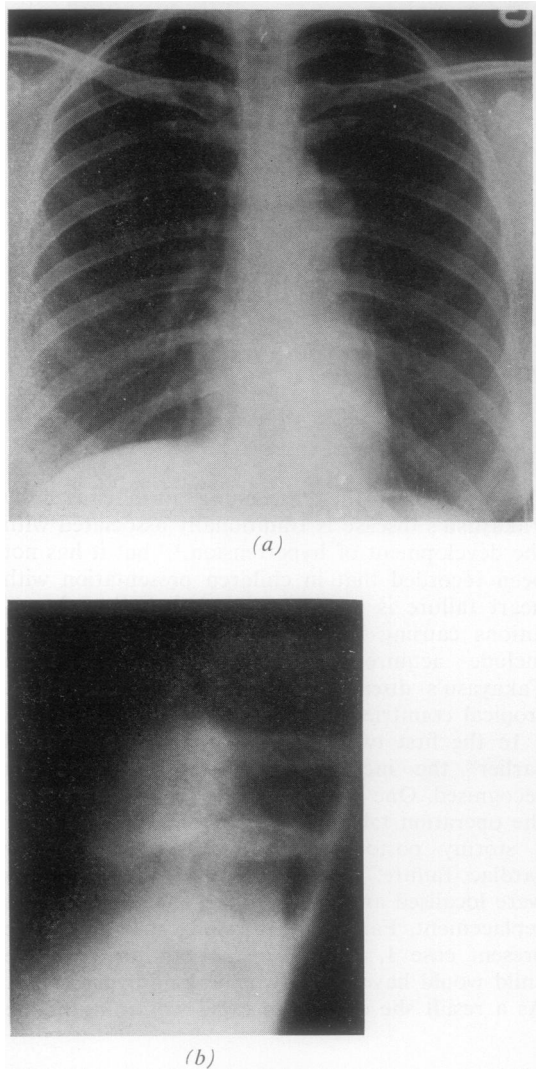


Fig 1 *PA chest radiograph (a) and lateral tomogram (b). Note paravertebral mass.*

Address for reprint requests: Dr BK Nigam, Cardiothoracic Unit, Walsgrave Hospital, Coventry, West Midlands CV2 2DX.

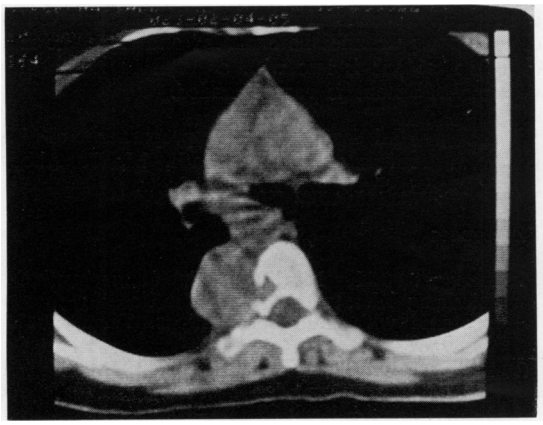


Fig 2 *Computerised axial tomogram, level T6. Paravertebral mass erodes the body of the vertebra, closely applied to the descending aorta.*

a sclerotic cavity, as had been shown by the CT scan. The wall of the cavity was curetted. After operation the patient made an uneventful recovery and the blood pressure and urinary catecholamines returned to normal (urine NA: 120 μg/24 hours).

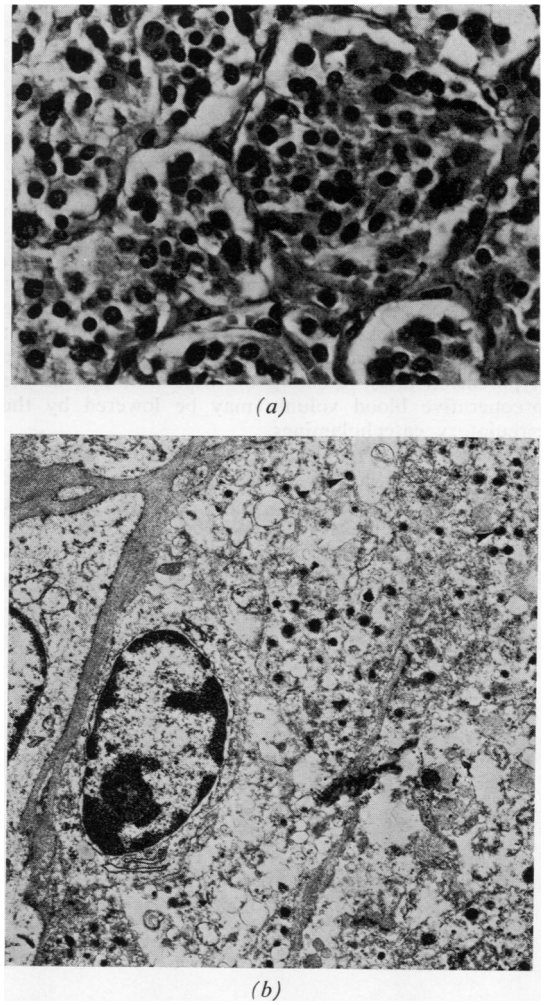
Pathology

The macroscopic appearance was of a lobulated, encapsulate mass $5 \times 3 \times 2{\cdot}7$ cm. The cut surface was grey and haemorrhagic and after fixation in chromate the tissue showed a dark brown colour. Sections (fig 3a) showed this tumour to be composed predominantly of large polygonal and ovoid cells with plentiful eosinophilic granular cytoplasm and vesicular nuclei arranged in alveoli with a highly vascularised intervening stroma. In some areas there was an admixture of large polygonal cells with small cells containing hyperchromatic round nuclei. The cells showed mild nuclear pleomorphism and occasional mitoses. Diffuse yellowish brown chromaffin positive granules were seen in the cytoplasm of the tumour cells fixed in dichromate solution, and the granules showed characteristic tinctorial properties when Serki's, Schmorl's, and Masson Fontana staining techniques were used.

The electron microscopic study of the formalin-fixed tumour tissue (fig 3b) showed poor preservation of the tumour cells, but membranebound electron-dense core granules of the neuro-secreting "arrow" type seen in APUD cell tumours were present in the cytoplasm. Biochemical analysis of the tumour showed noradrenaline 1200 μg/gm wet tissue and dopamine 37 μg/gm wet tissue, confirming noradrenaline secretion.

Discussion

Duke *et al*[2] described a case of retroperitoneal and mediastinal chemodectomas in a 42-year-old negress



(a)

(b)

Fig 3 *(a) Tumour cells showing nuclear polymorphism and fine granular cytoplasm. Cells arranged in an alveolar pattern (Haematoxylin and Eosin, original magnification $\times 720$). (b) Low power electron micrograph showing dense membrane-bound granules in the cytoplasm (original magnification $\times 18\,270$).*

with a five-year history of episodic hypertension, palpitation, and nervousness. Edwards and Heath[3] reported a case in which a nodule of chemoreceptor tissue was found deep in the lung of a lady who died from a pontine haemorrhage complicating systemic hypertension. This may have been a further secreting chemodectoma. Fawcett and Husband[4] described a case of chemodectoma of the lung with hypertension. Histologically the chromaffin reaction was negative although this may have been caused by formalin fixation.

The most useful diagnostic tool before operation

is arteriography as these tumours are highly vascular. In our case computerised tomography was able to define accurately the relation of the tumour to aorta and vertebral body as confirmed at operation. Additional studies to exclude the possibility of endocrinopathy should include urinary catecholamine determination. When catecholamine secretion is demonstrated all further operative procedures, including the aortogram, should be performed with α and β adrenergic blockade, either as phentolamine infusion supplemented by propranolol or with labetolol. These measures will reduce the risk of hypertension, tachycardia, and arrhythmia during tumour removal. Replacement of blood loss at operation is of major importance, to prevent severe hypotension after resection of these tumours, as the preoperative blood volume may be lowered by the circulatory catecholamines.

Operation is often hazardous because of the large vascular supply and a tendency of the tumour to encircle vital structures, in addition to the physiological effects of catecholamine secretion. Although these tumours may remain asymptomatic for years, there is usually continued growth resulting in morbidity from local pressure.[5] Since the possibility of malignancy cannot be excluded and since the risk of complications is less with a small tumour, early surgical treatment is recommended.

We are grateful for help in the preparation of this paper to Professor Donald Heath, Mr R Abbey Smith, Dr Ian Gray, and Dr Julie Crow.

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