Intrathoracic Phaeochromocytoma

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SUMMARY

Paroxysmal or sustained hypertension in a patient with a tumour in the costovertebral region of the thorax should suggest the presence of a phaeochromocytoma. Adequate roentgenographic examination of the chest and determination of urinary catechol excretion are the principal diagnostic aids.

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Intrathoracic tumours of the nervous system are usually found in the posterior mediastinum. If such a tumour is accompanied by paroxysmal or even sustained hypertension, the possibility of an epinephrine- or norepinephrine-secreting phaeochromocytoma should be considered in the differential diagnosis.

The purpose of this article is to report the rare occurrence of a functioning intrathoracic phaeochromocytoma in a young female patient who was referred for exclusion of an intracranial tumour because of headaches and poor vision.

CASE REPORT

A girl aged 15 years was admitted to hospital with the complaint of persistent headache and failing vision. On examination, her blood pressure was raised (300/180 mmHg) and she had gross papilloedema. No localising signs were present. The full blood count was normal but the erythrocyte sedimentation rate was raised. Chemical analysis showed normal blood urea and electrolyte levels. Examination of her urine revealed the presence of albumin. Creatinine clearance was normal. The 24-hour urine analysis for normetadrenaline and metadrenaline was 8,2 mg/24 hours (normal $<$ 1 mg/24 hours).

A postero-anterior chest roentgenogram suggested a paravertebral mass to the left of vertebrae D8 - 10 (Fig. 1), which could not be adequately demonstrated on the lateral projection. This was confirmed on tomography, which demonstrated a soft-tissue mass producing pressure erosion on the lateral aspect of the adjacent vertebral body and pedicle of D4 (Fig. 2). Excretory urography with tomography demonstrated the normal situation of both kidneys. No abnormal masses were demonstrated. At thoracotomy, a large paravertebral tumour was removed (Fig. 3), and histology showed that it was a phaeochromocytoma (Fig. 4). Postoperatively, the patient's

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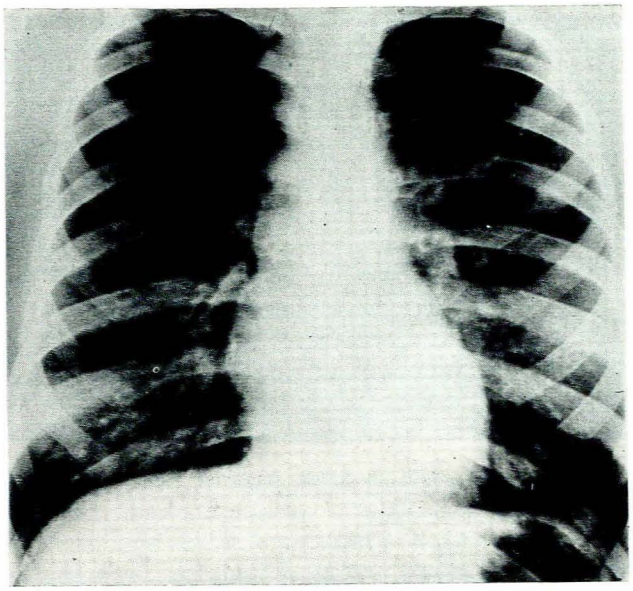

Fig. 1. Postero-anterior roentgenogram of chest. A mass is present behind the heart.

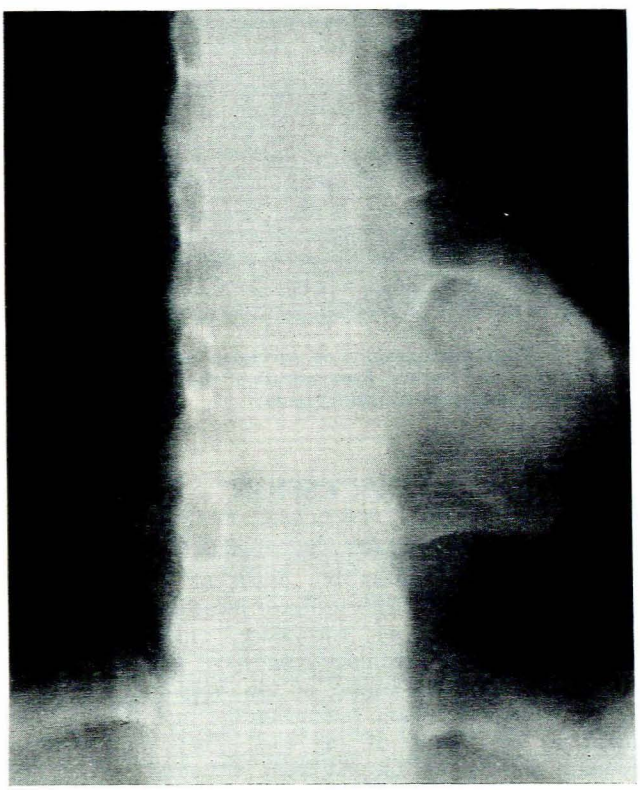

Fig. 2. Anteroposterior tomography of the paravertebral region, demonstrating a soft-tissue mass on the left, eroding the adjacent pedicle and vertebral body of D9.

blood pressure remained elevated for 4 days, with an accompanying tachycardia (probably due to circulating catecholamines still present in the blood), and then gradually returned to normal levels. She is at present still asymptomatic, with a normal blood pressure.

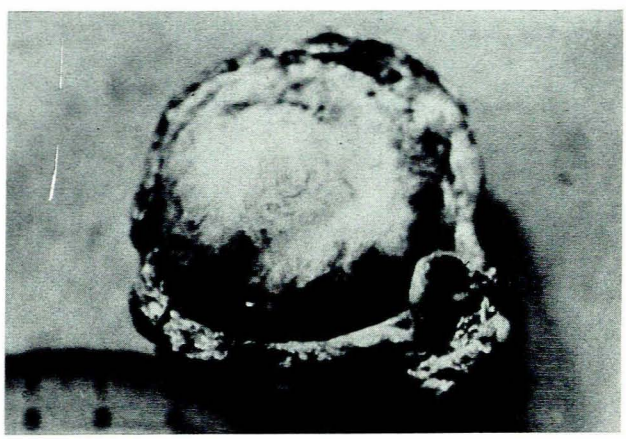

Fig. 3. Macroscopic specimen removed from left hemithorax.

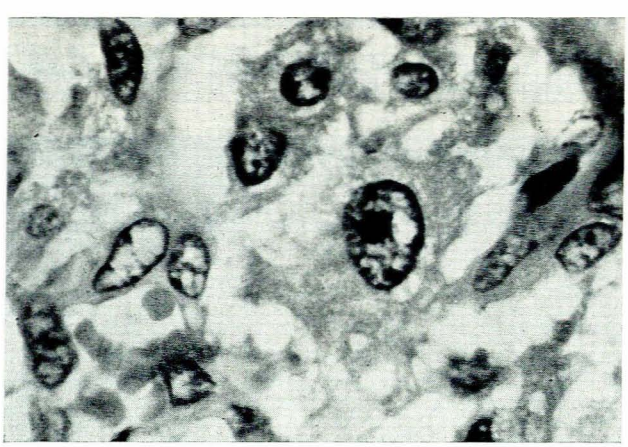

Fig. 4. Photomicrography of specimen stained with haematoxylin and eosin, demonstrating the architecture of a functioning phaeochromocytoma.

DISCUSSION

Ten per cent of all phaeochromocytomas are extra-adrenal in location, arising from the organ of Zuckerkandl or sympathetic ganglia.[1] The thoracic location is the rarest.[2]

To the best of our knowledge, only 24 cases of intrathoracic phaeochromocytoma have been reported.[3-7] They arise along the course of the sympathetic chain in the paravertebral region retropleurally, and they lie just lateral to the heads of the ribs. They may produce pressure erosion of the adjacent vertebral bodies and pedicles.

Roentgenographic demonstration of an intrathoracic phaeochromocytoma may not be easy, unless the tumour is large. On a postero-anterior film, the growth may be obscured by those mediastinal structures which project laterally beyond the lateral margins of the vertebral column (Fig. 1). Furthermore, the lateral film may not give the help which it often does in costovertebral tumours, since they may not cause a recognisable density in the costovertebral area on a postero-anterior radiograph of the chest. It is therefore important to include a penetrated postero-anterior chest view—as well as screening of the patient, oblique views and tomography—to exclude such a tumour with certainty (Fig. 2).

Biochemical tests have made the diagnosis of a phaeochromocytoma easier, but localisation of the tumour can still be extremely difficult. The majority of tumours are functioning,[2] but in those that do not function, the diagnosis may only be made at surgical laparotomy. Therefore, even in the absence of hypertension, the possibility of a clinically non-functioning phaeochromocytoma should always be considered in the differential diagnosis of posterior mediastinal tumours, since excessive manipulation of such a tumour at operation may result in a sudden rapid rise in blood pressure—with a mortality rate of 50%.[8] Von Euler[9] observed that postganglionic nerve fibres secrete norepinephrine instead of epinephrine. It is therefore possible to suggest that a tumour secreting norepinephrine is more likely to be located in an extra-adrenal than in the adrenal situation. This is important, since there is a general tendency to concentrate only on the abdomen when using radiography with its specialised techniques.

In the radiological differential diagnosis of this posterior mediastinal mass, a neurofibroma was also considered, but was thought to be less likely because of the tumour's broad base adjacent to the dorsal spine and the erosion of the adjacent pedicle from without.

About 10% of phaeochromocytomas are malignant,[10] and of these, one-third are extra-adrenal in situation. Pathological evaluation of this specimen demonstrated the absence of malignancy, but it is reported that histological evaluation of malignancy is unreliable in this condition and that malignancy must be watched for by continued follow-up of every individual patient.

This case emphasises the importance of the extra-adrenal situation of phaeochromocytomata and is a further indication for a penetrated routine chest X-ray film in the evaluation of patients with hypertension.

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REFERENCES

1. Palmieri, G., Ikkos, D. and Luft, R. (1961): Acta endocr., **36**, 549.
2. Green, W. O. jun. and Bassett, F. R. (1961): Amer. J. Clin. Path., **35**, 142.
3. Maier, H. C. and Humphreys, G. H. (1958): J. Thorac. Surg., **36**, 625.
4. Pampari, D. and Lacerenza, C. (1958): *Ibid.*, **36**, 174.
5. Cone, T. E. jun. and Pearson, H. A. (1963): Pediatrics, **32**, 531.
6. Cueto, J. C., McFee, A. S. and Bernstein, E. F. (1965): Dis. Chest, **48**, 539.
7. Fries, J. G. and Chamberlin, J. A. (1968): Surgery, **63**, 268.
8. Phillips, B. (1940): Arch. Path., **30**, 916.
9. Von Euler, V. S. (1951): Ann. Surg., **134**, 929.
10. Graham, J. B. (1951): Int. Abstr. Surg., **92**, 105.