Intrathoracic chemodectoma with multiple localisations

L. K. LACQUET, A. C. MOULIJN, C. M. JONGERIUS, M. LIMBURG, AND J. B. RENSING

From the Department of Thoracic, Cardiac, and Vascular Surgery, St. Radboud Hospital, University of Nijmegen; Lung Clinic, Medical Centre Dekkerswald, Groesbeek; and Laboratory of Pathology, St. Canisius Hospital, Nijmegen, The Netherlands

Lacquet, L. K., Moulijn, A. C., Jongerius, C. M., Limburg, M., and Rensing, J. B. (1977). *Thorax*, 32, 203–209. **Intrathoracic chemodectoma with multiple localisations.** In 1958 a mediastinal tumour was discovered in an asymptomatic woman with a history of vomiting and an oesophageal anomaly which had not been treated. A tumour of the anterosuperior mediastinum in relation to the aortic arch was extirpated and proved to be a chemodectoma or non-chromaffin paraganglioma.

At subsequent follow-ups the mediastinum was never normal and the heart size progressively increased, the oesophageal anomaly remaining unchanged. A small opacity appeared in the left lung in 1974. An operation performed in 1975 revealed an osteochondroma in the lung, pericarditis, and an intrapericardial chemodectoma. The oesophagus was not explored.

An intrathoracic chemodectoma is rare. The importance of angiography in its diagnosis is emphasised. Malignant degeneration is seldom observed. Therapy is surgical, the tumour being radioresistant. The possibility to be considered in our patient was either relapse of the tumour with degeneration or multiple localisations of the tumour.

An intrathoracic chemodectoma or paraganglioma is a rare tumour. Lattes (1950) described the first two mediastinal localisations. Shaw and Kennedy (1956) described a chemodectoma in relation to the ninth intercostal space. Barrie (1961) reviewed 12 cases of intrathoracic chemodectoma from the literature and added two personal observations. Elders and de Boer (1961) reported three personal cases, including one with an intrapulmonary localisation, and, in a thesis on paragangliomas, Elders (1962) described six intrathoracic cases in The Netherlands (including our patient after her first operation); in addition he collected the data on 19 patients from the literature. Smithers and Gowing (1965) collected 28 cases including one personal observation. Reboud *et al.* (1967) reported the first case in France, and Wilkinson and Forgan-Smith (1969) the first in Australia. In a review of 111 intrathoracic neurogenic tumours, Oosterwijk and Swierenga (1968) identified one benign and one malignant paraganglioma.

According to Lattes (1950), paraganglionic tumours can, on the one hand, arise from the ortho-sympathetic nervous system, being localised in the

adrenal cortex, in a few mediastinal cell groups or along the abdominal aorta, and on the other hand can be related to the parasympathetic nervous system (eg, carotid glomus and aorticglomera) or be localised in cell groups in the vicinity of the vagus nerve and glossopharyngeal nerve. When stained with chromium salts, the tumours of the former group prove to contain chromaffin cells and to be functionally active—phaeochromocytomas or chromaffin paragangliomas. Tumours of the latter group are functionally inactive—non-chromaffin paragangliomas, which Mulligan (1950) described as chemodectomas because they arise from the chemoreceptor system.

According to Totten (1973) and Glenner and Grimley (1974) paragangliomas are neuro-endocrine neoplasms arising from the paraganglion system containing the adrenal medulla and the extra-adrenal paraganglia, ie, carotid, aortic or aorticopulmonary paraganglia. Biochemical, histochemical, and ultrastructural studies have shown that all the paraganglia store catecholamine granules in the chief cells, and that the tumour cells are neoplastic chief cells containing catecho-

lamine granules (Alpert and Bochetto, 1974). The previous distinction between chromaffin and non-chromaffin has therefore become obsolete.

In the human organism, the aortic glomera or intrathoracic chemoreceptors are usually found at four sites (Boyd, 1937; LeCompte, 1948): (1) where the ductus arteriosus or ligamentum arteriosum arises from the pulmonary artery; (2) at the origin of the left coronary artery, between aorta and pulmonary artery; (3) at the bifurcation of the brachiocephalic trunk or lateral to the origin of the right subclavian artery; (4) along the left anterolateral aspect of the aortic arch, at the origin of the left subclavian artery.

Case report

A woman aged 31 was admitted to hospital in 1958 because routine screening had revealed a mediastinal opacity. Her only complaints were fatigue and vague backaches. The history showed an episode of vomiting at age 16 (in 1942); the patient reported that an oesophageal anomaly had been diagnosed but, owing to wartime conditions, the investigation had not been completed and no treatment had been instituted.

Clinical and laboratory findings were normal. The chest radiograph disclosed an opacity in the left anterosuperior mediastinum (Fig. 1). The bronchogram was normal. At contrast radiography of the oesophagus the contrast medium passed

unobstructed but the proximal two-thirds showed dilatation, and there were multiple lacunae (Fig. 2). Oesophagoscopy revealed a proximal

Fig. 2 Contrast radiography of the oesophagus: dilatation of the proximal two-thirds and multiple filling defects.

Fig. 1 Chest radiograph, anteroposterior projection (4 July 1958): opacity in the left upper mediastinum.

megaesophagus with pronounced mucosal folds which readily yielded to pressure. A biopsy specimen showed normal oesophageal mucosa. Since the patient was not inconvenienced by oesophageal symptoms she refused further examination of the oesophagus but consented to an operation for the mediastinal tumour.

After left-sided thoracotomy an egg-sized tumour was found in the anterosuperior mediastinum; the tumour was amply vascularised and partly attached to the aortic arch. During extirpation of this tumour the phrenic nerve was damaged.

Pathological examination of the resected specimen revealed a well-defined tumour measuring 6×5×4 cm and weighing 75 g. Microscopic examination identified it as a 'parasympathetic paraganglioma or chemodectoma' (Fig. 3).

The patient continued to report regularly for review and was doing well apart from transient complaints of regurgitation when supine. The consecutive chest radiographs showed, however, that a mediastinal opacity persisted; from 1969 on the heart shadow increased in size. In 1974, moreover, a small tumour formed in the central area of the left lung. In 1975 both the heart shadow and the tumour had increased in size (Fig. 4), and the patient was therefore readmitted for further investigation.

The tomogram showed that the shadow in the central area of the left lung was sharply defined, and the bronchogram showed occlusion of a segmental bronchus of the lingula. The oesophageal anomaly was unchanged. A cine-angiogram of the right atrium revealed an opacity between the atrial cavity and the edge of the heart shadow (Fig. 5), and an angiogram of the ascending aorta disclosed an amply vascularised tumour between the ascending aorta and aortic arch on the one hand and the trunk of the pulmonary artery on the other, immediately above the left coronary artery (Fig. 6).

The condition was tentatively diagnosed as relapse of the chemodectoma with possibly malignant degeneration and pericarditis as well as a lung tumour which might be an intrapulmonary chemodectoma.

The patient was still asymptomatic and there were no indications of tamponade. A reoperation was proposed and accepted by the patient. It was performed on 21 April 1975 via a left anterolateral thoracotomy with subsequent transverse splitting of the sternum. The tumour in the lingula was enucleated and examination of frozen sections identified it as a benign osteochondroma (Fig. 7). The oesophagus was not explored.

When the pericardium was opened, 600 ml of clear fluid drained. There was an amply vascular-

Fig. 3 Tumour in superior mediastinum 1958. Typical alveolar pattern with vascularised stroma. Parasympathetic paraganglioma or chemodectoma. Haematoxylin and eosin ×400.

Fig. 4 *Chest radiograph, anteroposterior projection (3 February 1975): upper mediastinum broadened; heart shadow increased in size; small tumour in the central area of the left lung.*

Fig. 5 *Cine-angiogram of the right atrium: opacity between right atrial cavity and edge of heart shadow.*

Fig. 6 *Cine-angiogram of the ascending aorta: vascularised tumour along ascending aorta immediately above the left coronary artery.*

Fig. 7 Tumour of lingula 1975. Tumour composed of mature bone and rather cellular cartilage. Benign osteochondroma. *H and E* $\times 160$.

ised intrapericardial tumour the size of a plum between the aortic arch and the trunk of the pulmonary artery. The tumour extended as far as the origin of the left coronary artery; it was resected, together with the adventitia of the aorta and the pulmonary artery. After resection a rupture in the trunk of the pulmonary artery caused massive blood loss and cardiac arrest. The tear was oversewn, and after blood transfusion and cardiac massage the heart resumed its contractions. The patient was ventilated artificially until the next day, the further postoperative course being uneventful.

Pathological examination of the tumour, which measured $8 \times 4 \times 4$ cm and weighed 55 g, confirmed the diagnosis 'benign aortopulmonary chemodectoma'. The histological features corresponded with those of the tumour resected in 1958 (Fig. 8).

Discussion

The collective surveys of patients with an intrathoracic chemodectoma show an equal age and sex distribution (Barrie, 1961; Elders, 1962; Smithers and Gowing, 1965). The patients are often asymptomatic, the tumour being discovered in 50% of cases at routine screening or at post-mortem examination (Gremmel *et al.*, 1966). If

present, symptoms are usually vague and not demonstrably related to the tumour. Complaints of pain depend on the site of the tumour and prevail in particular in association with tumours of the costovertebral sulcus. Respiratory symptoms result from displacement of bronchi and lungs. Dysphagia can result from external compression of the oesophagus (Nickels and Friman, 1972). Severe general symptoms are suggestive of a malignant change.

Chemodectomas can be localised at the site of the aortic glomera in the anterior mediastinum, or in the posterior mediastinum at the level of the costovertebral sulcus if the tumour arises from paraganglionic tissue in the adventitia of the aorta (Duncan and McDonald, 1951; Taylor and Evans, 1958); intrapulmonary localisations have also been reported (Heppleston, 1958; Elders, 1962). Aberrant paraganglionic cells, and therefore paragangliomas, can also be present along the course of intrathoracic rami of the vagus nerve, eg, in the wall of the oesophagus (Taylor, 1951).

In our patient, a benign chemodectoma along the anterolateral aspect of the aortic arch was resected in 1958, and a benign chemodectoma between the ascending aorta and the pulmonary artery was removed in 1975. Since the mediastinal shadow never completely disappeared after the first operation, both tumours may already have

Fig. 8 Tumour relapse in mediastinum 1975. The same histological picture as in 1958. Benign chemodectoma. H and E $\times 400$.

been present in 1958. It is also possible (but unproven) that the oesophageal tumour is a third chemodectoma.

Arteriography is of great importance as a diagnostic aid in view of the vascular nature of the chemodectoma; angiography can determine the vascularisation as well as resectability (d'Altorio *et al.*, 1974).

Madden (1958) studied the clinical course and prognosis of these tumours in 11 published cases. Malignant degeneration in this series was exceptional and was similar to that in the carotid glomus tumour. Depending on the malignancy criteria applied, the incidence of malignancy is sometimes reported to be as high as 50%. With the occurrence of distant haematogenous metastases as the sole criterion, however, the malignancy incidence is only 3%. Benign aortic glomus tumours can, as a rule, be totally extirpated. As in the case of carotid glomus tumours, resectability is largely determined by the possibility of damaging the large vessels during resection. As in the case of carotid glomus tumours, operative morbidity and mortality certainly offset the advantages of resection of the tumour. Total resection of malignant aortic glomus tumours is impossible and radiotherapy is ineffective (Monro, 1950; Gillis *et al.*, 1956). As a chemodectoma is potentially malignant the treatment should be surgical.

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- Request for reprints to: Professor L. K. Lacquet, Department of Thoracic, Cardiac and Vascular Surgery, St. Radboud Hospital, University of Nijmegen, The Netherlands.