

A non-chromaffin paraganglioma of the lung

H. MOSTECKÝ, J. LICHTENBERG, AND M. KALUŠ

From the Second Clinic of Tuberculosis, and the First Surgical Clinic, Medical Faculty, Charles' University, Prague, and Hlava's Institute of Pathological Anatomy, Prague

Paraganglioma occurs approximately as frequently in the suprarenal glands as in other sites. In the neck these tumours originate most frequently in the carotid or the jugular body, and in the ear from the tympanic ganglion. In the chest they grow from the aortic or the vagal bodies. They arise in the abdominal cavity in the para-aortal chromaffin tissue and the remnants of the organs of Zuckerkandl. Besides these usual sites they may exceptionally appear elsewhere—namely, in the femoral region, in the orbit, in the tongue muscles, in the diaphragm, and in the jugular vein (Pryse-Davies, Dawson, and Westbury, 1964; Heppleston, 1958; Smithers and Gowing, 1965).

Genetically and functionally paragangliomata may be divided into two groups (Valach, 1956): (1) those originating in the chromaffin paraganglia of the sympathetic chain—these are the true paragangliomata and they usually show hormonal activity; (2) those originating in the achromaffin paraganglia of the parasympathetic nerves, the vagus and glossopharyngeus. From these the achromaffin paragangliomata, or chemodectomata, usually grow. The achromaffin paragangliomata are more frequent. One of their most usual sites is in the anterior mediastinum in relation to the ascending and the horizontal parts of the aortal arch (Chodkowska and Pawlicka, 1961; Pachter, 1963; Zak and Chabes, 1963). They generally occur in women in their thirties and their size varies from 1 cm. to 17 cm. They are usually spherical, rigid tumours of an elastic consistence, sometimes lobular, and of a brownish colour on the cut surface. They are richly vascularized and surrounded by a capsule. Quite frequently they adhere firmly to neighbouring organs (Pachter, 1963). Their symptomatology is minimal. Most of these tumours are found by chance as a result of routine radiography of the chest. Only when the tumour enlarges considerably do symptoms caused by pressure appear; these symptoms are dyspnoea, or pain in the chest. Outside the mediastinum non-chromaffin paragangliomata appear exceptionally.

The object of our paper is to present a non-chromaffin paraganglioma situated in the lung. This is a rarity, and so far only one example has been described by Heppleston (1958).

CASE REPORT

A man of 46 years, working as a locksmith in a grindstone factory, had been a moderate smoker of some 10 to 12 cigarettes daily since the age of 25 years. In July 1958, on the occasion of routine radiography, an oval shadow, situated paracardially on the right side, was discovered. The patient was subjected to regular radiological controls and the shadow showed no changes. In January 1964 he was admitted to hospital for examination. On admission he had no complaints; his weight was 70 kg. and his height 172 cm. All laboratory investigations were negative. The electrocardiogram showed only non-specific changes. On the chest radiograph, at the level of the fifth rib paracardially on the right side, there was an oval shadow, 5 by 4 cm. in size, sharply circumscribed, which partly overlapped the heart shadow (Fig. 1). On the lateral projection the shadow impinged upon the inferior pole of the hilum and extended into the central lobe of the lung (Fig. 2).

Pneumomediastinoscopy, bronchoscopy, and bronchography were not helpful. Cytological examination of the sputum was negative. Thoracotomy for a probable benign tumour situated in the medial or inferior lobe of the right lung was recommended. The operation was performed on 4 February 1964. A right-sided thoracotomy through the fifth intercostal space was done; the pleural cavity was free. At the base of the middle lobe, in the vicinity of the hilum, a spherical solid mass the size of a mandarine was palpated. After dissection of the visceral pleura we were able to separate the tumour from the lung. A number of vessels connecting the tumour with the surrounding tissues were divided and ligated. The tumour had a smooth surface, was of reddish-grey colour, and had a vascular, laminated structure.

HISTOLOGY Examination shows a slightly encapsulated neoplasm which is very vascular and composed of large, often polymorphous cells with light, granulated protoplasm. The general arrangement is solidly

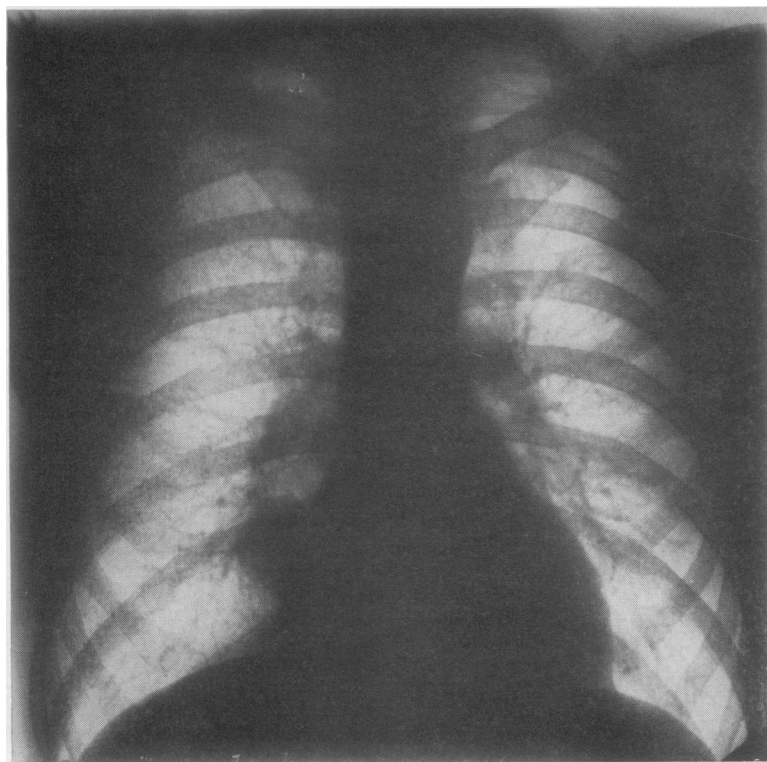


FIG. 1. *Postero-anterior radiograph showing an oval shadow paracardially on the right side.*

FIG. 2. *Lateral tomograph with the shadow in the middle lobe.*

FIG. 1

alveolar. There are no mitoses. In the protoplasm of the tumour cells a small amount of lipids, glycerides, and PAS-positive material is present. Single cells contain argentaffin granules which give a positive Bodian's reaction. The stroma, separating the alveoli from the supporting tissue of the tumour cells, is very cellular. The walls of the ectatic venules show hyaline changes, and in some places calcification is present. There are spaces lined by one layer of cylindrical epithelium that resemble rudimentary bronchioles. These are the characteristic features of an angiomatous form of a non-chromaffin paraganglioma of the lung (Figs 3 and 4).

DISCUSSION

Only one case of a non-chromaffin paraganglioma of the lung has been described in the literature, the case of Heppleston (1958). His patient was a man of 53 years. An oval tumour, 4 by 3 by 3 cm., was removed from almost the same site as in our patient. The tumour was well circumscribed and was easily separated from the lung after incision of the visceral pleura. But it was somewhat different from our case and from the other paragangliomata described in the litera-

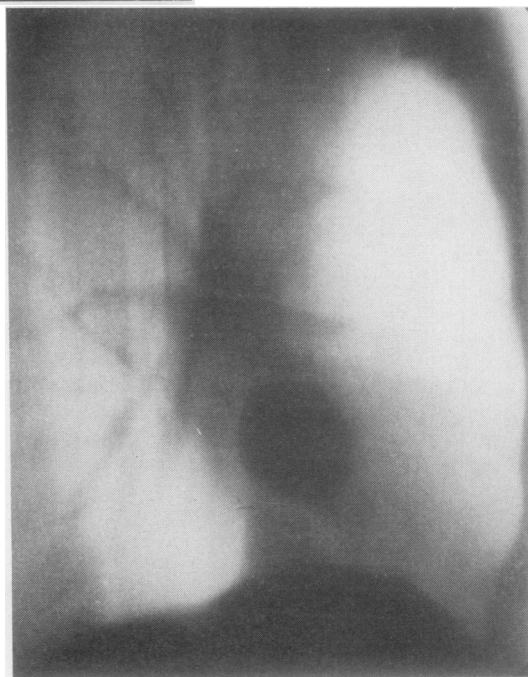


FIG. 2

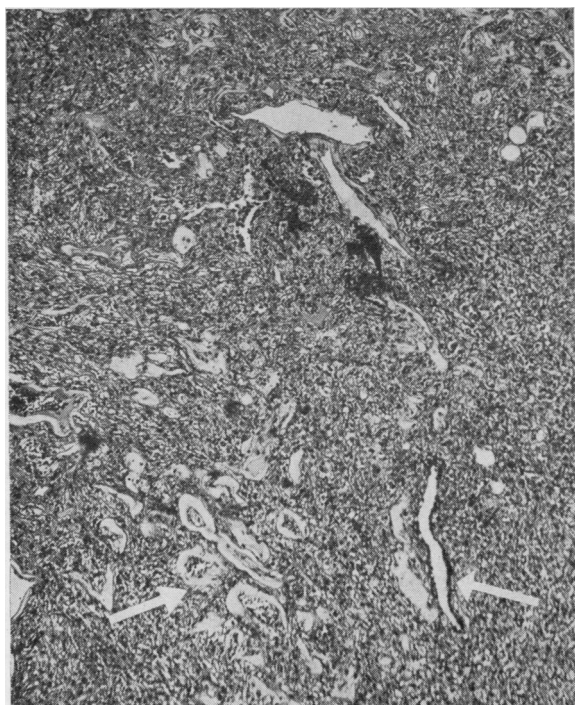


FIG. 3. Non-chromaffin paraganglioma, highly vascularized with rudimentary bronchioles (arrows). H. and E., $\times 52$.

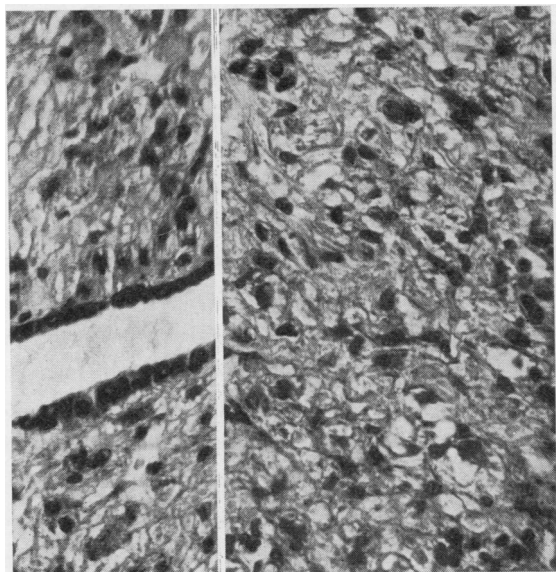


FIG. 4. Solidly alveolar arrangement with cellular fibrous stroma. To the left there is a rudimentary bronchiole. H. and E., $\times 305$.

ture. It was greyish in colour, rather brittle, surrounded by a fibrous capsule and with septa projecting into the lung. This macroscopic difference was probably due to the relation between the tumour tissue and the supporting stroma, which determines the consistence of the tumour.

The biological properties of paraganglioma can be determined only very approximately from the microscopical appearance, because a certain degree of polymorphism of the tumour cells is usual, and even an infiltrative growth of paraganglioma into the neighbourhood does not always signify malignancy. The vascularity of paraganglioma is variable and may reach such a degree that the tumour is considered to be a haemangioma. As far as the differentiation between a chromaffin and non-chromaffin paraganglioma is concerned, Bodian's technique of demonstrating argentaffin granules in the protoplasm of the tumour cells is effective. Chromaffin tumours give a positive reaction, and in the others the reaction is negative. The difference between both groups is consequently only quantitative. An insignificant partial argentaffinity in our observation places the tumour into the group of non-chromaffin paragangliomata.

In our case the discovery of rudimentary bronchioles is interesting; this testifies to its relation to the lung and it resembles a pulmonary hamartoma. The discovery of a non-chromaffin paraganglioma in the lung is surprising, because until recently nothing was known about the presence of chemoreceptive tissue in the lung, though there was physiological proof of chemical reflexes in animals. Recently Zak and Chabes (1963) proved that chemoreceptive tissue exists in the neighbourhood of bronchial arteries and the sensitive nerves in the lung of man. Bednár (1966) emphasized the presence of chemoreceptive tissue at the branching of pulmonary venules. Chemo-dectomata, which usually have a close relation to the pulmonary vessels, also testify to its existence. These are not true tumours but a simple hyperplasia of chemoreceptive tissue, which appears more frequently during congestion in the lesser circulation (Barroso-Moguel and Costero, 1964). Similar small tumours, frequently multiple, were found in approximately 3% of the normal population, mostly women of middle and advanced age (Zak and Chabes, 1963). These findings support the possible appearance of a non-chromaffin paraganglioma in the lung of man.

SUMMARY

A non-chromaffin paraganglioma of the lung is described. It was removed from a man of 46 years.

It was situated in the base of the middle lobe. The origin of paraganglioma of the lung is supposed to be in the chemoreceptive tissue, which is hypertrophied in 3% of the population, especially in the presence of pulmonary hypertension.

REFERENCES

- Barroso-Moguel, R., and Costero, I. (1964). Some histochemical tests in Zak's chemoblastosis. *Amer. J. Path.*, **44**, 17a (Sci. Proc.).
- Bednář, B. (1966). Hypertensive changes of pulmonary venulae. *Acta Univ. Carol. Med. (Praha)*, in the press.
- Chodkowska, S., and Pawlicka, L. (1961). Pulmonary chemodectoma. *Nowotwory*, **11**, 421.
- Heppleston, A. G. (1958). A carotid-body-like tumor in the lung. *J. Path. Bact.*, **75**, 461.
- Pachter, M. R. (1963). Mediastinal nonchromaffin paraganglioma. *J. thorac. Surg.*, **45**, 152.
- Pryse-Davies, J., Dawson, I. M. P., and Westbury, G. (1964). Some morphologic, histochemical, and chemical observations on chemodectomas and the normal carotid body. *Cancer*, **17**, 185.
- Smithers, D. W., and Gowing, N. F. C. (1965). Chemodectoma in the region of the aortic arch. *Thorax*, **20**, 182.
- Valach, V. (1956). Extraadrenal paraganglioma. *Acta Univ. Carol. Med. (Praha)*, **2**, 411.
- Zak, F. G., and Chabes, A. (1963). Pulmonary chemodectomatosis. *J. Amer. med. Ass.*, **183**, 887.