

“PULMONARY HAMARTOMA” (THE CARTILAGINOUS TYPE)

BY

M. J. T. ADAMS

From the Bland-Sutton Institute of Pathology, Middlesex Hospital, London

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“When I use a word,” said Humpty Dumpty, “it means just what I choose it to mean—neither more nor less” (Carroll, 1908).

The word “hamartoma” (*ἁμαρτία*=error, *ωμα*=tumour) was first suggested by Albrecht in 1904 in his paper “Ueber Hamartome,” as the name of a group of tumour-like malformations in which the normal tissue elements of an organ are abnormally mixed. The abnormality could be in the quantity of the tissue, its arrangement, or in its degree of maturity, or in a combination of these faults. Albrecht, in this paper, was suggesting that tumours fell into two main categories, choristomas, which were tumours consisting of tissues not normally present in an organ, and hamartomas. Hence the term hamartoma was originated to describe a group of tumours which includes the majority of benign growths. When applied to the lungs, however, the term hamartoma has come to be used by many to refer to a specific, well-defined benign tumour composed mainly of cartilage. If the use of the word in this restricted sense was universal there would be no confusion; but in practice it is found that there are still people who use the word in its true, more general sense, embracing pulmonary adenomas, haemangiomas, and other more unusual lesions.

The purpose of this paper is to discuss the specific type of tumour referred to by many as a hamartoma; to show that it has a behaviour peculiarly its own; to discuss its diagnosis and treatment; and to suggest a more restricted nomenclature for it. Four original examples of these tumours are described and compared with a case of another type of benign pulmonary tumour.

The gross and microscopical structure of the tumours to be discussed is illustrated by the following case.

CASE 1.—Mr. J. H. died aged 76.

This man was first seen at the Middlesex Hospital in November, 1949, complaining of epigastric pain which was found to be due to a benign ulcer for which a partial gastrectomy was performed by Mr. R. S. Handley.

At this time he had noticed swellings of his fingers and wrists for a month before admission but had no symp-

toms or signs of respiratory disease. A radiograph of the chest showed a spherical mass lying in the left lower zone posteriorly, with calcification in the lower part. This mass was thought to be benign, possibly a dermoid or hydatid cyst, but in view of his age and general condition it was not investigated further except for two Cassoni tests which were both negative.

He was readmitted in November, 1954, with uraemia and congestive heart failure, and died within a few days. A further chest radiograph taken at this time showed a slight increase in size of the pulmonary mass.

A necropsy, performed by Dr. W. W. Richardson, showed that death was due to uraemia, secondary to bilateral pyelonephritis, and quite unrelated to the lung lesion.

The lower lobe of the left lung contained a rounded, firm, irregular white tumour, 3½ in. in diameter, in the posterior basal segment (Fig. 1). The tumour cut like cartilage and contained areas of mucoid material and cartilage with apparent patchy calcification.

Histological examination showed a tumour composed predominantly of cartilage, which was arranged in a lobular pattern. The lobules were separated by clefts lined by cuboidal epithelium. Between this epithelium and the cartilage were varying thicknesses of connective tissue, containing scattered smooth muscle fibres, and an inflammatory cell infiltration with many blood vessels (Fig. 2).

The sections of this specimen showed all the essential features of these tumours, which are the islands of cartilage separated by epithelial-lined clefts, together with varying amounts of connective tissue (Hickey and Simpson, 1926).

The first of these tumours confirmed by microscopy was reported by Lebert in 1845. Further isolated cases were reported, until in 1926 Hickey and Simpson reviewed 38 cases from the literature and reported two of their own, under the title “Primary Chondroma of Lung.” Further reports followed, and among recent reviews those of McDonald, Harrington, and Clagett (1945) and Stein, Jacobson, Poppel, and Lawrence (1953) give good accounts.

The tumours are rare. McDonald *et al.* give a figure of 0.25% among the adult population based on a series of 23 cases in nearly 8,000 necropsies. The majority of the tumours were smaller than that

FIG. 1

described here in Case 2, and would have been difficult, if not impossible, to detect during life. The sex incidence given in their series was three males to one female.

There are conflicting reports about the rate of growth of these tumours. Hall (1948) considered that growth is unusual, but Brewer, Brookes, and Valteris (1953) cited the case of a tumour which appeared over a period of four years in a 30-year-old man. Reports of cases before adult life are rare; the youngest in the series of McDonald *et al.* (1945) was a 21-year-old woman. Jones (1949) reported a related type of tumour in a newborn infant, but this, although containing some cartilage, consisted largely of fibrous tissue and was not strictly comparable. The tumours arise generally during adult life and therefore must grow, although this growth may be slow or erratic. The following case illustrates this slow growth.

CASE 2.—Mrs. B. M. died aged 39.

This patient had been under observation for a long time on account of severe mitral stenosis with attacks of cardiac failure, and her chest had been radiographed many times in the years since 1950. In 1951 a small opacity was noticed in the right mid-zone behind the anterior aspect of the fourth rib (Fig. 3). Subsequent films taken in 1953 and 1955 showed a slight but steady increase in size (Figs. 4 and 5).

It was decided in September, 1955, that, although she was by that time extremely ill, mitral valvotomy offered the only chance of relief, and that operation was performed by Mr. J. R. Belcher; she died shortly afterwards.

A necropsy performed by Dr. P. S. Andrews showed that, apart from the valvular disease, there was a round nodule 1 in. in diameter at the lateral edge of the upper lobe of the right lung. This was found to be a spherical cyst filled with mucoid material. The cyst wall was $3/16$ in.

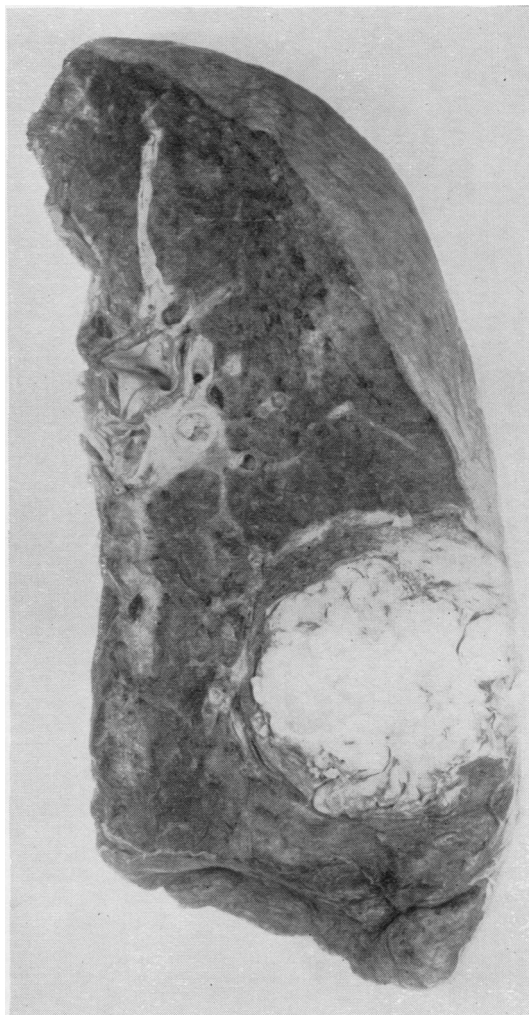


FIG. 1.—Case 1: Vertical section through the left lung to show position and relative size of tumour (by kind permission of Dr. W. W. Richardson).

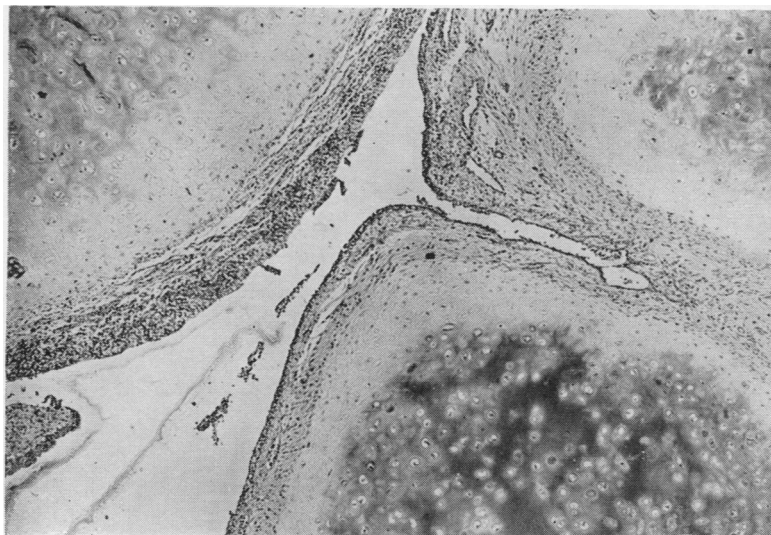


FIG. 2.—Case 1: Photomicrograph showing lobules of cartilage separated by clefts lined with cuboidal epithelium. Haematoxylin and eosin $\times 45$.

FIG. 2

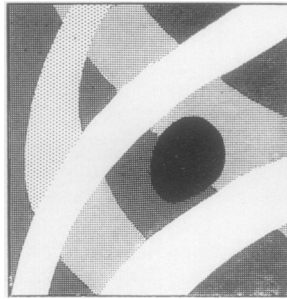
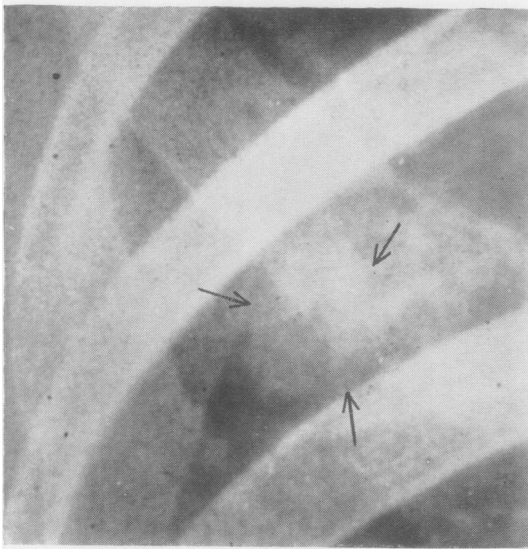
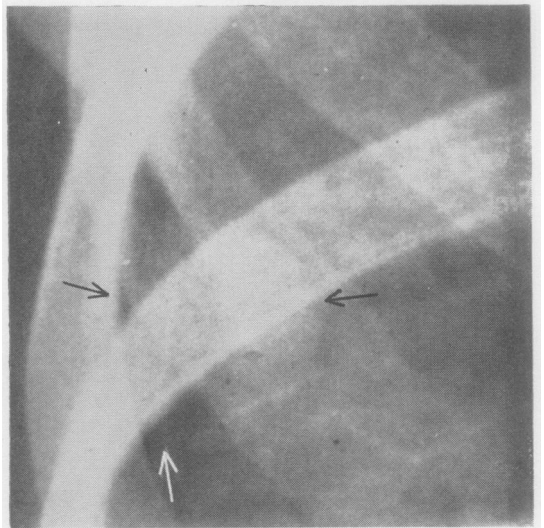
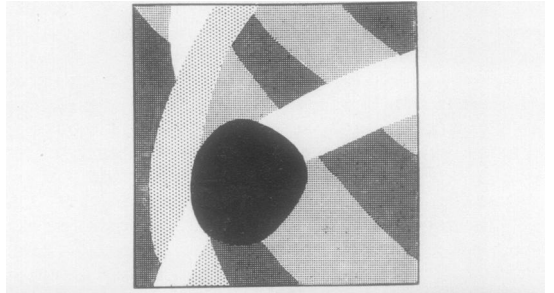
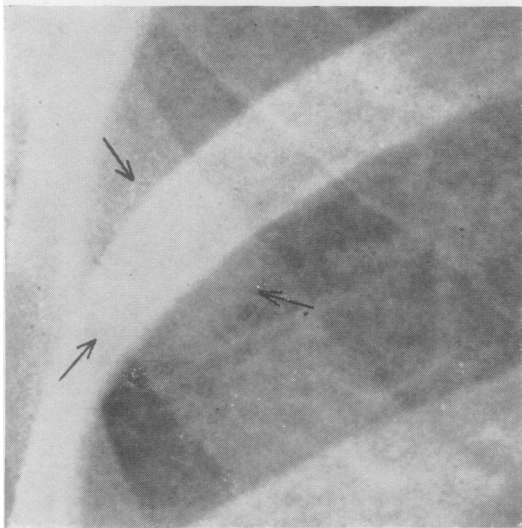
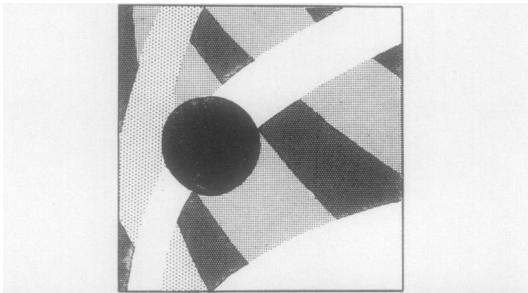


FIG. 3.—Case 2: Enlargement of right mid-zone of postero-anterior chest radiograph in January, 1951, showing tumour (position indicated by arrows and in line drawing). $\times 2.5$.



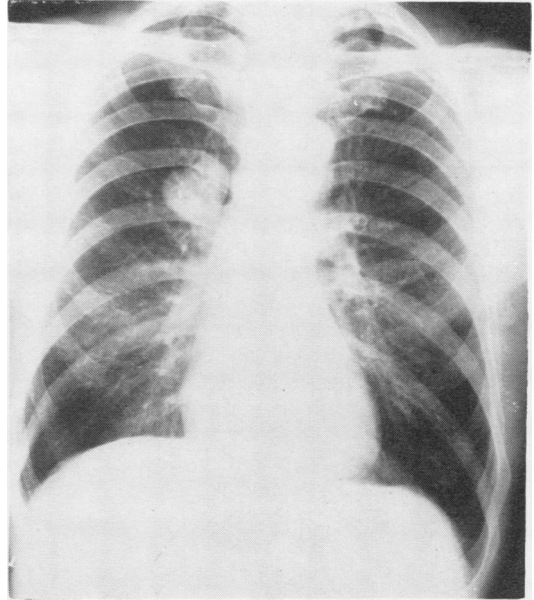
FIGS. 4 and 5.—Case 2: Same region of postero-anterior chest radiograph as in Fig. 3 in February, 1953, and September, 1955, showing progressive increase in size of the tumour, $\times 2.5$.

thick and consisted of cartilage enclosed in a capsule of ciliated columnar epithelium and fibrous tissue, together with a small amount of smooth muscle fibres.

DIAGNOSIS

The larger tumours are easily detectable, and show up radiologically as rounded shadows, sharply demarcated from the surrounding lung fields, and not associated with mediastinal glandular involvement. The tumour, being in lung tissue, moves with respiration and “ falls away ” with the lung if an artificial pneumothorax is induced. This latter test may be of value in distinguishing the tumour from an intrapleural mass, such as a fibroma or a loculated pleural effusion.

Calcification is present in a large proportion of the tumours and was demonstrated radiologically in two of the four cases. It may not be obvious in a straight film, but can often be seen if tomograms are taken. Fig. 6 is a film of the tumour described in Case 3 taken after its removal. This shows the characteristic punctate calcification, and can be compared with the straight radiograph and tomogram of the same patient (Figs. 7 and 8).



Figs. 7 and 8.—Case 3: Postero-anterior radiograph and right lateral tomogram at 9 cm. to show calcification.

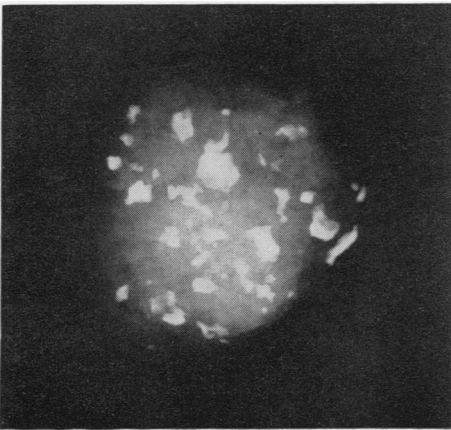
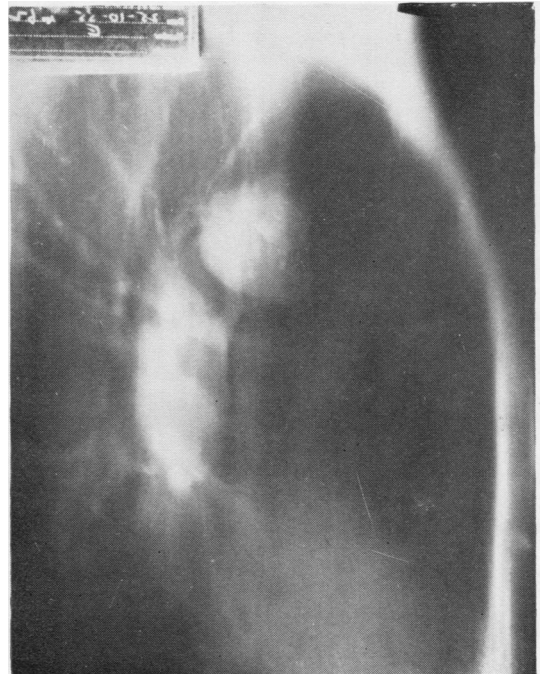


Fig. 6.—Case 3: Radiograph of tumour after its surgical removal showing the characteristic punctate calcification.



In life, the tumours are usually discovered accidentally in the course of radiological examination. There are no pathognomonic symptoms or signs, and no laboratory investigations which can be of assistance. The tumour is almost always out of reach of the bronchoscope, and there is rarely any indication for bronchoscopy. As can be seen from the illustrations to the above case it may be difficult to detect the tumour, although it may subsequently be discovered at necropsy. In this instance the low density of the shadow produced

by the mass, together with its position behind a rib shadow, meant that it was undetectable on several films.

The differential diagnosis in practice is between primary carcinoma of the lung, secondary neoplasms of any type, bronchial adenomas, which are usually situated near the hilum but can occur peripherally (Mayo, 1942), dermoid or hydatid cysts, tuberculomas, fibromas, and encapsulated fluid. If calcification is seen the possibility of malignancy is unlikely, and hence the field of choice will be narrowed considerably. However, small amounts of calcification may very occasionally be seen in large carcinomas of the lung (Davis, Peabody, and Katz, 1956).

The following two cases show that the lesion can be diagnosed radiologically with a fair degree of certainty before operation and an appropriate operative removal planned.

CASE 3.—Mr. G. S., aged 61, attended the Out-patient Department for advice regarding an inguinal hernia. He complained of a chronic cough with the production of an egg-cupful of grey sputum per day. A chest radiograph showed a rounded, solitary tumour in the right upper lobe, which, on tomography, was seen to contain areas of punctate calcification (Fig. 8). On physical examination there was some early clubbing, but no other abnormal signs in the respiratory system.

A diagnosis of pulmonary hamartoma [*sic*] was made and a thoracotomy was performed by Mr. J. R. Belcher on September 26, 1955. A firm tumour was palpated in the medial part of the anterior segment of the right upper lobe; it lay about 1 cm. below the pleural surface. The lung was incised over the tumour which was extruded intact. Two small blood vessels required ligation, and the incision was closed by direct suture. The patient

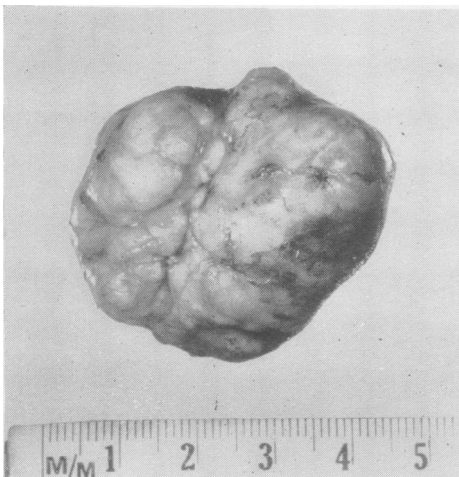


FIG. 9.—Case 3: Tumour after operative removal, showing lobulated surface.

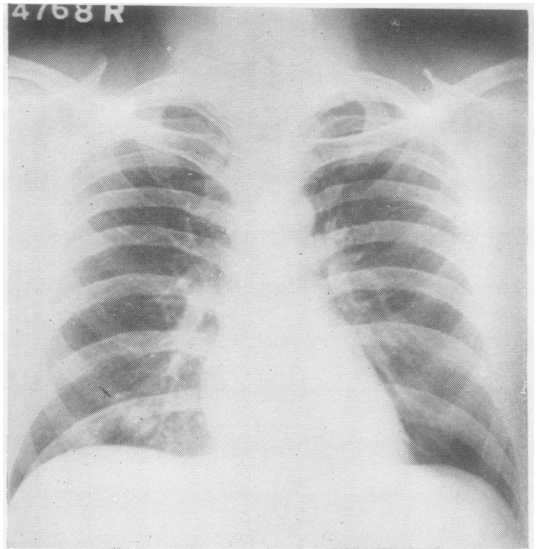


FIG. 10.—Case 4: Postero-anterior radiograph showing lesion in the right lower lobe.

made an uneventful recovery and was discharged from hospital 18 days after the operation. He has remained well since leaving hospital and states that his cough is noticeably better.

The tumour was $1\frac{1}{2}$ in. in its largest diameter, white with a lobulated surface, and rubbery in consistency (Fig. 9).

Sections showed a tumour consisting of lobules of cartilage, around which were areas of connective tissue containing muscle fibres, myxomatous and fibrous tissue, and nerve elements. Between the lobules were deep clefts lined by epithelium, which was mainly squamous but in places was cuboidal in type.

CASE 4.—Mr. W. A., aged 43, worked abroad for a public corporation and had had a chest radiograph in the course of a routine medical examination for re-appointment. A rounded shadow was seen in the right lower lobe (Fig. 10), and his reappointment was deferred until its nature could be determined. He was referred by the examining medical officer to Mr. J. R. Belcher and admitted to hospital for investigation. He had no respiratory symptoms beyond a slight "smoker's cough," and no abnormal physical signs were found in the chest.

At operation on September 21, 1955, a hard tumour was felt in the lateral basal segment of the right lower lobe. The same operative technique as for Case 3 was employed. This patient also made a completely uneventful recovery and was discharged from hospital on the 14th day after operation. When seen six weeks later he was symptom free and able to follow a completely normal life. He had been passed as medically fit by his employers and was returning to the Persian Gulf.

The tumour was a firm, white, lobulated mass $\frac{3}{4}$ in. in diameter. On section it was seen to consist of islands of cartilage separated by clefts lined with squamous epithelium.

TREATMENT

The pre-operative diagnosis of these tumours is uncertain, and, if there is any suspicion that a malignant growth is present, thoracotomy should be undertaken unless there is any contraindication. Apart from this, there is a case for surgical removal of these cartilaginous tumours, and this view is supported by McDonald and his colleagues (1945).

The tumours grow, albeit slowly (see Case 2), and may reach a considerable size (Case 1, and Jackson, McDonald, and Clagett, 1956). If and when this occurs there is some loss of pulmonary function, and the tumour may produce the same disability as a chronic pleural effusion. If with increasing size it should encroach on the hilar region it may cause bronchial obstruction with distal lung damage. Hence, if the tumour can be removed by the enucleation method described above while it is still small and the patient is fit, the possibility of subsequent lung damage is removed.

If the tumour is left alone, neither patient nor practitioner will be content without long-term radiological supervision. This focuses continual attention on the patient's state of health, with consequent anxiety, apart from any possible risks of repeated radiological examinations. If the tumour is removed, after one post-operative check the patient can be assured that there is no further danger.

Lastly, there is the occasion, as in Case 4 above, when a lesion is discovered accidentally, and an employer or insurance company is not satisfied as to its nature. If a man's livelihood is at stake there are good grounds for advising operative removal, even if it were possible to make a confident diagnosis pre-operatively and the patient is symptom free.

The operation of choice is an enucleation as described above. The majority of tumours can be removed in this way, being immediately below the pleural surface of the lung (Hickey and Simpson, 1926; Stein *et al.*, 1953). In these cases the operation is simple. The tumours can usually be located accurately by tomograms, and the incision planned accordingly. The only likely complication is air leakage from the incised lung, and this can be speedily detected and treated if drainage is employed routinely.

NOMENCLATURE

A group of tumours exists characterized by slow growth, a subpleural site, and lack of specific symptoms. The tumours in this group are composed predominantly of cartilage which frequently

becomes calcified, together with epithelial and connective tissue elements. These tumours come within the wide general classification of hamartomas; but this is too general a title for such a well-defined tumour. Other authors agree on this point and many suggestions have been made regarding nomenclature. Womack, first with Graham (1938), and later with Ehrenhaft (Ehrenhaft and Womack, 1952), has included them with adenomas and some other less well-defined tumours as “mixed tumours of lung,” but this does not define them as an entity. Furthermore including them with adenomas has the disadvantage of confusing them with a type of tumour which is potentially malignant. There is evidence in both of Womack's papers, and from other sources, that bronchial adenomas may show malignant properties, and their tendency to local invasion is shown in a recent paper by Ranger, Thackray, and Lucas (1956). I have been unable to discover any well-authenticated report that the tumours under discussion may do so. Greenspan's case (1933) of a primary osteoid chondrosarcoma of lung is often quoted, but there is no evidence that there was any pre-existing benign tumour, and Greenspan himself thought that it arose from the bronchial cartilage. Simon and Ballon's case (1947) of an unusual hamartoma appeared, from its description, to resemble an ossifying adenoma and was comparable to Case 5 below.

Several other names are used or have been suggested for these tumours. “Chondroma of lung” is often used, and this was the name used by Hickey and Simpson in their original review in 1926, but the purists say that a true chondroma cannot contain the other tissue elements which these tumours possess. Lipochoadenoma and adenochondroma have been suggested (Brewer *et al.*, 1953). The latter term, whilst being strictly more accurate than chondroma, describes another type of tumour behaving in a different manner as exemplified by the following case.

CASE 5.—Mrs. E. F., aged 61, was admitted to hospital for investigation of her complaints of general malaise for one year, with poor appetite and recent loss of 11 lb. in weight. The precipitating cause for her seeking advice had been three small haemoptyses in the two weeks before admission.

No abnormality was found on physical examination of the respiratory system, but a radiograph showed a rounded mass at the right base posteriorly, with local pleural adhesions. Examination of the sputum showed “many atypical cells some of which suggest malignancy,” and at bronchoscopy the right middle lobe bronchus was noticed to be displaced downwards, but no growth was seen. A diagnosis of carcinoma was made, and a thoracotomy performed by Mr. J. R. Belcher on June 22, 1955. A mass was palpated in the right lower lobe, proximal

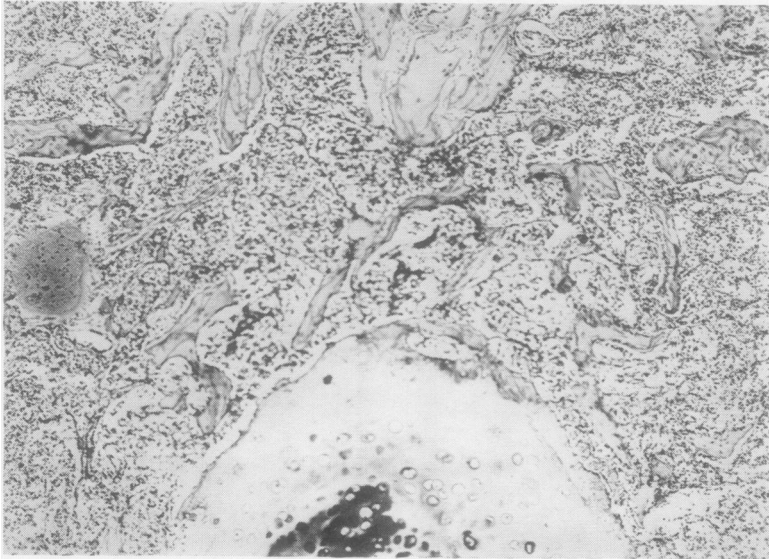


FIG. 11.—Case 5: Photomicrograph showing bone and an island of cartilage lying in an adenomatous matrix. Haematoxylin and eosin, $\times 38$.

to an area of atelectasis, and a dissection lobectomy was performed. She made an uneventful recovery from the operation, and when last seen on March 6, 1956, was symptom free.

Examination of the resected specimen showed a mass $1\frac{1}{2}$ in. in diameter close to the the right lower lobe bronchus, with a peripheral abscess in the lung tissue. On histological examination the tumour was seen to be composed of areas of glandular epithelium with acinar formation in parts, which had undergone extensive ossification, together with an island of cartilage (Fig. 11).

This tumour, which should be described as an ossifying adenochondroma, behaved in a different way from the tumours described earlier. Clinically the patient suffered from haemoptysis, malaise, and loss of weight; radiologically the tumour was central in position with associated lung damage; and histologically the structure was predominantly of adenomatous tissue. The whole clinical picture was one of such severity that the diagnosis of carcinoma of the bronchus was made. A lobectomy at least was essential and a pneumonectomy might justifiably have been performed.

I have quoted this last case, which exemplifies the type of tumour which could rightly be called an adenochondroma of lung, in order to stress the difference between it and the other four tumours described.

All the names so far suggested for the tumours previously discussed appear to be unsatisfactory for one reason or another. I would like to suggest that this specific lesion be known as a "chondro-adenoma of lung." This term, which so far as I know has not been suggested before, is histologically

accurate, and has the merit of stressing the predominance of cartilage, which is structurally the distinguishing feature of the tumours. It is also sufficiently different from the names of allied tumours to avoid confusion with other lung tumours which have a different structure and natural history.

Returning to Humpty Dumpty, one reads a few lines further on that, "when he makes a word do a lot of work he always pays it extra." Albrecht's term "hamartoma" was meant to describe a large group of tumours; let us stop overworking it by applying it to any one specific tumour, and use instead a term which gives a more precise definition.

SUMMARY AND CONCLUSIONS

A type of tumour, often known as hamartoma of lung, is discussed. The tumour represents a distinct entity characterized by subpleural site, slow growth, frequent calcification, and absence of invasive or other malignant properties. It has a histological structure predominantly of cartilage, with associated epithelial and supporting tissue elements. Four cases are described and compared with a fifth illustrating a different type of benign lung tumour. Diagnosis can never be proved pre-operatively, but calcification demonstrated radiologically in a solitary, rounded, peripheral lung shadow, in a man with few or no symptoms, is suggestive of this lesion. Treatment should be by surgical enucleation. The present nomenclature of these tumours is discussed and is considered unsatisfactory. The name "chondroadenoma of lung" is suggested.

I wish to thank Mr. J. R. Belcher for his encouragement and for permission to publish Cases 2 to 5 and Professor A. Kekwick for permission to publish Case 1; Dr. W. W. Richardson for Fig. 1, Mr. M. Turney for Figs. 3 to 10, and Mrs. Julie Honey for the line drawings illustrating Figs. 3, 4, and 5. I am grateful to Dr. A. D. Thomson and my colleagues of the Bland-Sutton Institute of Pathology for their advice.

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