

Cartilage-containing tumours of the lung

Relationship between the purely cartilaginous type (chondroma) and the mixed type (so-called hamartoma): an unusual case of multiple tumours¹

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An unusual case is reported of a woman aged 27 years who presented with four intrapulmonary cartilage-containing tumours which were resected from the left lung. The appearance of two new shadows in the chest several years later suggested that two of the resected tumours had recurred. Three of the four resected tumours consisted entirely of cartilage and bone and other connective tissues. The fourth tumour, although consisting almost entirely of cartilage and connective tissue, also contained epithelial tissue in the form of two small clefts, one in the periphery and the other in a connective tissue septum between the lobules of cartilage of the tumour. These tumours are regarded as a variation of the more typical cartilage-containing tumour of the lung which contains many spaces lined by respiratory epithelium and is regarded as a neoplasm arising in the connective tissue beneath the mucosa of a small bronchus with subsequent expansion into its lumen and enclosing spaces lined by the mucosal epithelium during its eccentric growth. The tumours consisting almost entirely of cartilage without spaces lined by epithelial cells are thought to expand into the adjacent lung tissue and not into the bronchial lumen. Therefore there is no inclusion of respiratory epithelium from the mucosa of the bronchus of origin.

It is not uncommon to find benign tumours containing cartilage within the thoracic cage. These tumours may occur within the major bronchi (endobronchial) or within the lung parenchyma (intrapulmonary). There has been much controversy in the literature concerning the relationship between endobronchial and intrapulmonary cartilage-containing tumours (Bateson, 1965), but there is good evidence that both these forms are the same tumour, composed of the same tissues, and differing only in their site of origin (Sutherland, Aylwin, and Brewin, 1953; Hasche and Haenselt, 1960; Bateson, 1965).

The intrapulmonary cartilage-containing tumours are composed predominantly of cartilage, and because of this were originally thought to be chondromata of the lung (Hickey and Simpson, 1926; Klages, 1931). The cartilage is usually arranged in lobules separated by septa of connective tissue in which are found groups of fat

cells, bundles of smooth muscle fibres, collections of round cells, blood vessels, and branching and elongated or cystic spaces lined by various types of respiratory epithelium. Within the lobules of cartilage, bony trabeculae enclosing marrow spaces may be present. Since these tumours are composed of connective and epithelial tissues normally present in the bronchi and lung, they were regarded as hamartomata (Bayer, 1929; Jaeger, 1935) or adenochochondromata (McGlumphy, 1924). Möller (1933) referred to her case as a mixed tumour, a description recently adopted by Willis (1958).

Although the majority of intrapulmonary cartilage-containing tumours show a lobular structure consisting of a mixture of epithelial and connective tissues, there are a few exceptions which consist almost entirely of cartilage (sometimes ossified with marrow spaces and areas of degeneration) and minimal amounts of other connective tissues including fat but without the presence of branching and elongated or cystic spaces lined by respiratory epithelium within the

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mass of the tumour. Examples of tumours of this type have been reported by Bikfalvi, Molnár, and Horányi (case 8, 1954), Edling (1938), Franco (1958), Hodges (1958), Kirschner and Kny (case 6, 1957), and probably case 3 in the series reported by Weisel, Glicklich, and Landis (1955).

The presence of two types of cartilage-containing tumours, one consisting entirely of cartilage and connective tissue and the other consisting predominantly of cartilage and connective tissue but with epithelial tissue also, has given rise to controversy. Franco (1958), Hall (1948), and Hochberg and Pernikoff (1950) make a distinction between these two types and regard those consisting almost entirely of cartilage as chondromata and those with epithelial tissue as hamartomata (adenochondromata, hamarto-chondromata). It is with this controversy in mind that the following unusual case is reported and discussed.

CASE REPORT

A white woman aged 27 years was found on routine radiological examination of the chest in 1957 to have four shadows in the left lung (Fig. 1). These consisted of a small, round shadow behind the left clavicle, a large, well-defined shadow in the left lower lobe, and two medium-sized shadows which were heavily calcified, one in the anterior segment of the left upper lobe and the other at the left base.

A chest radiograph taken one year later showed an increase in the diameter of the largest shadow. The patient had no symptoms referable to the respiratory system, and a Casoni test was negative. In August 1958, thoracotomy was performed, all four tumours were easily located, and all shelled out of the lung without difficulty. Her post-operative course was uneventful and she was discharged from hospital in September 1958.

Macroscopically, the four resected tumours measured 7.5, 2.5, 2.5, and 0.75 cm. in diameter, and they weighed 118, 33, 13, and 3 g. respectively. The largest specimen was rather ragged towards one end but was covered elsewhere by a thin capsule. The cut surface showed pale, fairly firm tissue, which was slightly translucent, and many areas of cystic change. The cysts were filled with a yellowish sticky fluid. The other three specimens had a lobulated external surface and were of bony hard consistency.

Microscopically, the large tumour revealed a composition of cartilage and fibrous tissue. The latter was seen centrally, where there was hyaline change, oedema, and regions of cystic degeneration. Some of the cartilage was calcified. In one connective tissue septum between the lobules of cartilage a double epithelial layer was identified (Fig. 2). There was a similar structure at one point on the periphery of the tumour. Elsewhere, a layer of fibrous tissue surrounded by compressed lung was present around

the periphery of the tumour. The smaller specimens were a mixture of cartilage, bone, and adipose tissue, the proportion of cartilage being progressively less as the tumours decreased in size.

At follow-up in 1959, a radiograph of the patient's chest revealed clear lung fields. A radiograph in 1963 showed the presence of two new shadows, one below and one lateral to the left hilum, and a further radiograph in 1965 showed that these shadows had increased slightly in size. A comparison of these with the pre-operative radiographs showed that the two new shadows corresponded in position with the shadows of two of the original tumours, and the new shadows were presumed to be due to a recurrence of two of the tumours. A complete radiographic survey of the skeleton at this time was negative, excluding the possibility of metastases from a chondrosarcoma.

DISCUSSION

This case presents several unusual features of cartilage-containing tumours of the lung. These are the age and sex of the patient, the multiplicity of lesions, the possibility of recurrence, and the histological structure.

The age and sex of the patient are unusual, as the majority of these tumours occur in men over the age of 40 years (Bateson, 1965).

The presence of several intrapulmonary cartilage-containing tumours of the lung in one patient appears to be extremely rare. Cases with multiple pulmonary hamartomata have been reported by Keers and Smith (1960), and Logan, Rohde, Abbott, and Meltzer (1965), but in each of these cases the tumours were of fibro- leiomyomatous type, did not contain cartilage, and were different lesions from those under discussion. Wilkins (1955) reported a series of asymptomatic isolated pulmonary nodules; one of the series had two hamartomata (presumably cartilaginous) lying adjacent to each other in the basal segment of the left lower lobe. Muendel and Yelin (1955) quote Virchow, who described a case with three chondromata in one lung.

The appearance of the two new shadows several years after the removal of and corresponding in position with two of the original tumours suggested the possibility of recurrence. In a previous survey of the literature (Bateson, 1963) no recorded case of recurrence of an intrapulmonary cartilage-containing tumour could be found with the exception of the case reported by Schiødt and Jensen (1960). This case was different in that the recurrent tumour had the histological structure of a sarcoma. It is also possible that the two new shadows in the present case may be new tumours

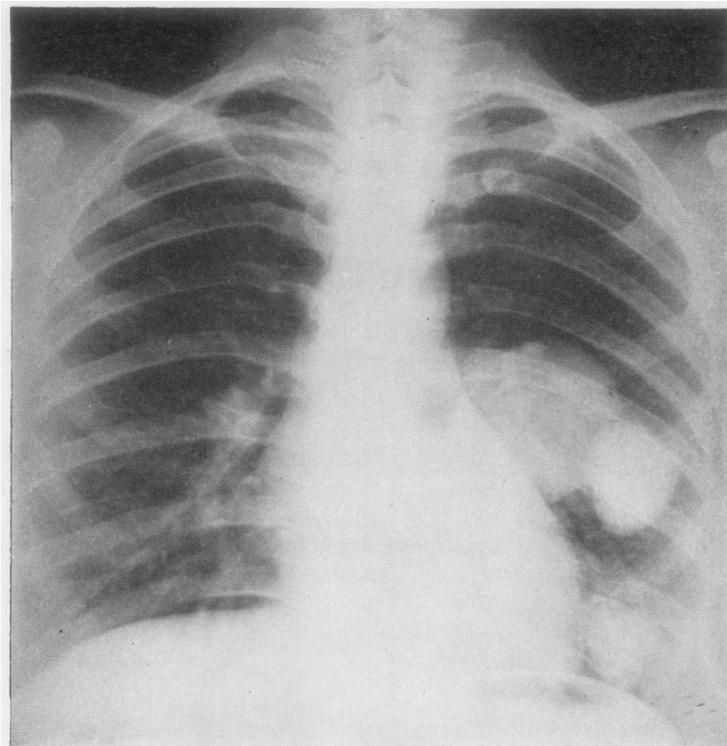


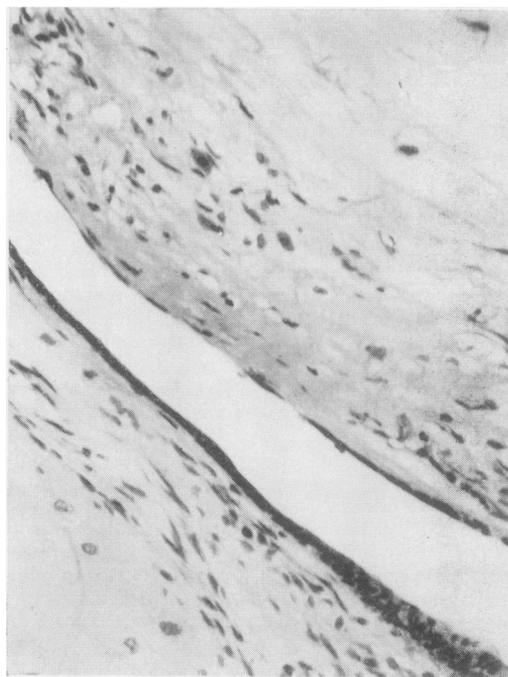
FIG. 1. (Left) Postero-anterior radiograph of the chest showing the four shadows in the left lung. The smallest shadow lies behind the inner end of the left clavicle. The calcification in the two medium-sized shadows is well shown.

FIG. 2. (Below) Photomicrograph of the largest tumour showing a cleft lined by respiratory epithelium in connective tissue septum between lobules of cartilage. H. and E. $\times 335$.

and not due to a recurrence of two of the original tumours.

Finally, the most interesting feature was that the structure of these four tumours was different from that of the majority of cartilage-containing tumours. Three of the tumours consisted of cartilage, bone, and adipose tissue without the presence of epithelial-cell-lined clefts and spaces which are characteristic of these neoplasms. The cartilage gradually merged into the surrounding connective tissue, which was in turn surrounded by compressed lung parenchyma. The largest tumour showed a similar structure except for the absence of bone and the presence of two elongated spaces lined by respiratory epithelium, one lying in a connective tissue septum between areas of cartilage and the other at the periphery of the tumour.

There are two possible interpretations of the nature of these four tumours. First, that they are pure chondromata and are therefore distinct and different from the usual type of intrapulmonary cartilage-containing tumour of the lung (which contains epithelial tissues); and, secondly, that they are of the same nature as the more usual type. The absence of any connexion with the



cartilage of a bronchial ring contraindicates the first possibility, and the second possibility is supported by the fact that one of these four tumours contained epithelial tissues, and similar cartilage-containing tumours with only a minimal amount of epithelial tissue have been reported (Bateson, 1965; McDonald, Harrington, and Clagett, 1945, case 7).

The occurrence of tumours consisting entirely, or almost entirely, of cartilage and connective tissue within the lung parenchyma supports the theory that these lesions arise in the connective tissue of the wall of small bronchi (Bateson, 1965) with metaplastic change in the connective tissue giving rise to cartilage, fat, smooth muscle, and lymphoid tissue. The presence of epithelial-cell-lined spaces within the majority of these tumours results from the inclusion of the epithelium lining the bronchus of origin by the lobular and eccentric growth of the cartilage and connective tissue into the lumen of the bronchus (Bateson, 1965). The absence of epithelial-cell-lined spaces in three of the four tumours in the present case and in those reported by Bikfalvi *et al.* (1954), Edling (1938), Franco (1958), Hodges (1958), Kirschner and Kny (1957), and Weisel *et al.* (1955) can be explained by the proliferation of the connective tissue and the formation of cartilage with expansion of the tumour outside the bronchus into the adjacent lung parenchyma.

It must be mentioned that Möller (1933) regarded these cartilage-containing tumours as neoplasms (mixed tumours), and she believed that it was the proliferation of the bronchial epithelium which induced the underlying connective tissue to form cartilage, fat, and other tissues, implying that they are primarily epithelial tumours. This theory was supported by Willis (1958). Spencer (1962) did not agree with this, as he found it hard to see how respiratory epithelium could have this function of inducing the formation of cartilage and other tissues. However, the existence of intrapulmonary tumours consisting entirely of cartilage and connective tissue, or with only minimal amounts of epithelial tissue, makes it impossible to accept the theory of Möller and supports the origin of these neoplasms from the connective tissue of the wall of small bronchi with the secondary inclusion of epithelium from the bronchus of origin.

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