HAMARTOMA

OSCAR CRAIG, F.R.C.S.I., D.M.R.D., F.F.R.,

Department of Radiodiagnosis, St. Mary's Hospital, London, W.2, and Harefield Hospital, Middlesex.

ACCORDING to Gudbjerg (1961) 253 cases of hamartoma were recorded in the literature up to 1960. Since then however many more cases have been added. Hood, Good, Clagett and McDonald (1953) reported that in the Mayo Clinic 16% of solitary tumours resected from the lung were hamartomata.

Bateson (1965) in a review of 155 patients with solitary lung lesions, found 23 to be hamartomata. He states however that the incidence in his series was higher than in most of the earlier reported series.

It is possible in some cases to make a pre-operative diagnosis and so save the patient a thoracotomy. The relevant features of hamartomata will be discussed with the illustration of the following case:

Case Report

The patient, a male aged 51 years, presented at the hospital complaining of waterbrash and heartburn. A barium swallow and meal was requested.

No lesion was found on clinical examination. On barium examination no lesion was seen in the oesophagus, stomach or duodenum. However, it was seen that there was a calcified opacity lying peripherally at the right lung base. (Figs. 1 and 2). This opacity required further investigation and tomography was undertaken. (Fig. 3).

The lesion was found to be well defined and to have a clear cut edge. There was slight lobulation. One of the features was calcification in its centre, and this calcification was "pop-corn" in type. There were no other nodules in its vicinity and the surrounding lung appeared normal. The differential diagnosis included mainly, hamartoma, carcinoma or tuberculoma. The appearances however were considered to be typical of a hamartoma.

Discussion

Albrecht in 1904 was the first to use the term hamartoma. He described these tumours as developmental tumour-like malformations, in which the normal elements of the organ are abnormally represented, in their quality, arrangement, degree of differentiation or all three. The histological picture is one of a stroma of cartilage surrounded by fibrous and myxomatous connective tissue. Lobules covered by respiratory epithelium, and cleft-like spaces lined by respiratory epithelium may be found. There may be areas of calcification, or ossification, glandular elements or smooth muscle. Any variation of these components may be found.

Willis describes two types of hamartoma (a) Infantile and (b) Adult. He states that the infantile type is a true hamartoma, but that the

FIG. 1.—Calcified opacity in chest X-ray—lying just behind diaphragm.

adult type should be referred to as a mixed tumour of the bronchial wall.

Le Roux (1964) discussing this problem agrees with the views of Willis, and includes pulmonary arterio-venous malformations among the hamartomata. He thinks that chondromatous pulmonary hamartomata should be called mixed tumours of the bronchial wall. However, many authors still believe in their origin from embryonic cell rests. If this is so, it seems difficult to explain that these tumours are found most often in the sixth decade, and that only 4% occur before the age of 40 years. (Bleyer and Marks, 1957).

There are two macroscopic types of tumour (a) intra-pulmonary, (b) endobronchial.

The intra-pulmonary type is by far the most common. Bateson and Abbott (1960) reviewed 253 cases and found that only 27 of these were endobronchial. This is an incidence of just over 10%.

FIG. 2.—Calcified opacity, better seen on barium examination film.

The tumour shows a preponderance for the male sex and most authors give this preponderance as approximately 4 to 1. Gudbjerg (1961) described 10 cases, in which there was not so marked a male predilection, and in which 4 of the 10 tumours were endobronchial in origin.

The tumour is found most often on the right side, peripherally and at the base. It is seen less often in the upper lobe. The mode of presentation is vastly different in the two types. The intrapulmonary type, as in the case reported here, is usually found incidentally at routine chest examination. It is exceedingly rare for it to produce any symptoms. However, two cases reported by Bateson and Abbott (1960) did present with hæmoptysis, and it is said that large tumours may press on the hilum and give rise to cough and dyspnoea. This is indeed rare. Bateson subsequently points out that most of these tumours are under 4 cm. in diameter. In fact the majority of these tumours present as coin lesions, which according to Thornton, Adams and Bloch (1944) are 1 to 5 cm. in diameter.

The endobronchial tumours may cause symptoms and signs associated with bronchial blockage. They may therefore cause cough, hæmoptysis and dyspnoea. Areas of atelectasis may occur or even a lung abscess due to distal sepsis. Obstructive emphysema has been described as a result of a ball-valve action of the tumour and because of a similar mechanism, asthmatic attacks may occur.

In the vast majority of cases, the problem presented by a hamartoma is one of differentiation from other causes of coin lesions. Although many lesions can produce coin opacities e.g., abscess, infarct, fungus infections, the most important lesions to be considered are, (1) primary carcinoma (2) secondary deposit (3) tuberculoma (4) arterio-venous malformation (5) adenoma.

In this differentiation, tomography is the most useful additional investigation. On tomography, a hamartoma presents a well-defined and smooth

FIG. 3.—Tomogram showing "pop-corn" calcification and well defined edge of tumour.

outline. There may sometimes be lobulation. No satellite lesions will be seen, and the surrounding lung is normal, except sometimes in the case of endobronchial tumours. Calcification has some importance. Bleyer and Marks state that calcification is not of much diagnostic aid, since it is only demonstrated in 15% of hamartomata. However, when present, the calcification is often characteristic, especially when it assumes a "pop-corn" appearance. Simon (1956) says a blob of central calcification is characteristic of hamartoma. Bateson gives a long list of authors who state that the presence of calcification excludes a primary carcinoma. Tuttle, Barrett and Hetzler (1955) and Taylor, Riokin and Salver (1958) say that calcification can be seen in primary malignant lesions. However, this is exceedingly rare, and a good working rule is that calcification virtually excludes a primary malignancy.

Kerley (1954) and Simon (1956) both state that on tomography, carcinoma gives a lobulated appearance even when it has a well defined edge. Bateson says that a primary carcinoma is always lobulated. Rigler in 1955 described a "notch sign" in carcinoma, seen on tomography. The diagnosis of a primary carcinoma is more easy when an irregular edge is seen, with malignant "strands" extending into the surrounding lung. This is never seen in hamartomata.

Secondary deposits, although seldom solitary, can however present considerable difficulty in differentiation. They tend to increase in size rapidly, whereas hamartomata grow very slowly. Calcification in secondary deposits is very rare indeed. Calcified deposits from ovarian and testicular tumours have been reported by Semple and West (1955). Speed (1943) reported calcified secondaries from osteogenic sarcoma. Secondaries are usually accompanied by a relevant history, whereas intra-pulmonary hamartomata are clinically silent.

Tuberculomata may show irregular punctate or diffuse calcification. They may be lobulated. Satellite nodules are often present in the surrounding lung. These lesions are more common in the upper lobes.

Arterio-venous malformations often show the vascular composition of the tumour itself, and more important, the feeding vessel from the hilum is often demonstrated.

An adenoma is usually found in the larger bronchi, and it rarely presents as a coin lesion. It may not be possible to differentiate it from an endobronchial hamartoma without histological examination. It is however more common in the female and under 40 years of age.

Even with tomography it may not be possible in many cases to diagnose dogmatically hamartoma to the exclusion of carcinoma. It is then necessary to remove the tumour surgically.

Rarely hamartomata may show malignant change in the smooth muscle element, and Cavin (1958) described two tumours showing this change.

Bateson in his review of solitary lung lesions concludes that "if a patient presents with a solitary lung lesion in the chest, which on tomography presents an homogenous shadow, with or without calcification, and with a well-defined smooth outline, and provided there is no history of, or the presence of a malignant tumour elsewhere, the diagnosis of a mixed tumour is most likely."

It would therefore appear possible in many cases to diagnose a hamartoma by plain X-rays and tomography, if the findings are correlated with the history of the patient. In these cases, thoracotomy can be avoided.

There will always remain some cases where no true differentiation from a primary carcinoma can be made, and these must then be subjected to thoracotomy.

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