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FIBROMA OF THE VISCERAL PLEURA*

BY

SIR CLEMENT PRICE THOMAS AND C. E. DREW

From the Brompton and Westminster Hospitals, London

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Intrathoracic fibromas are rare tumours, but as might be expected they have been described in many different situations within the chest in relation to areas of connective tissue. In a personal series of 198 primary intrathoracic neoplasms, excluding carcinoma of the lung and oesophagus, Harrington (1950) encountered 13 fibromas or lipofibromas, all of which arose in the posterior mediastinum, but most of those which have been fully described appear in single case reports. Study of the available literature suggests that these tumours may be classified in four groups depending upon their position. Some have been found in the larger bronchi and occasionally diagnosed after biopsy through a bronchoscope. Others have been identified in relation to the parietal pleura, although some of these are undoubtedly fibrous tumours of neural origin. The third group occurs in the mediastinum and the fourth comprises those which appear to arise beneath the visceral pleura. The growth may assume giant size before the onset of symptoms referable to the chest, and may obtain additional blood supply through adhesion to neighbouring structures. It is then difficult—and sometimes impossible—to define its origin.

Clagett and Hausmann (1944) claim the biggest fibroma yet removed from the chest, a mass weighing 5 kg., filling the lower part of the right pleural cavity and extending behind the heart to the left side. This was so densely adherent that it was not possible to decide whether it arose beneath the visceral or parietal pleura.

This paper is intended to describe our findings in some examples which fall in the fourth group, i.e., those which we believe arise in close relation to the visceral pleura.

CASE REPORTS

CASE 1.—The patient was a man of 62 years of age when surgical operation was advised in 1947. At the age of 13 he developed right-sided pleurisy. When he was 21 years of age he spent two months in a sanatorium after complaining of cough and pain in the chest, but no diagnosis was made. He had suffered similar pleuritic pain intermittently ever since. In 1939 he was noticed to have clubbed fingers and he stated that his hands and feet were getting bigger. In 1943 he was admitted to hospital for six months where he was investigated and treated for severe joint pains in the hands and feet. At this time he stated that an attempted aspiration was made of the right side of his chest, but no diagnosis was made.

In early 1945, following a severe exacerbation of similar pain, he was once more admitted to hospital, when radiographs showed what was thought to be a raised right hemidiaphragm and pleural changes compatible with the history of recurrent pleurisy. Bronchography demonstrated bronchiectasis of the basal bronchi.

The arthritis was treated symptomatically without complete relief and after 18 months he was forced to give up his work. He was completely bed-ridden with joint pains for three months before his last admission in March, 1947.

At this time he complained of pain in the lower part of the right chest with cough productive of purulent sputum. The joints of the hands and feet were swollen, tender, and painful, with gross drumstick clubbing of the fingers. Radiography now showed a rounded opacity above the right cupola of the diaphragm which in retrospect was present in the films taken two years previously, but had since increased in size. There appeared also to be collapse of the middle lobe and part of the lower lobe. Bronchoscopy showed rigidity and narrowing of the lower lobe bronchus, but no evidence of intrabronchial tumour.

At thoracotomy (C.P.T.) a large mass was found which appeared to be arising from the middle lobe with the lower lobe attached to it. It did not appear to be a carcinoma because the bulk of the tumour was outside the pulmonary substance. A lower and middle lobectomy was performed and the patient made a good recovery. His joint pains disappeared at once and a month after operation he was walking for the first time in nine months. He is now fit and well and the finger clubbing is now no longer present.

Examination of the specimen showed it to be a well-encapsulated fibroma which, in part had undergone necrosis, presenting in the fissure between the lobes which had been removed and probably arising in relation to the visceral pleura of the middle lobe.

*Based on a paper read to the Thoracic Society at the Royal College of Surgeons in London on February 22, 1952.

Although the result obtained from this operation was gratifying, it was felt that with careful dissection it might have been possible to remove the tumour without sacrifice of lung tissue.

CASE 2.—In the same year a woman of 59 years of age was admitted to hospital, and her history briefly is as follows.

She had been well until three years previously when she developed intermittent pain and swelling in the feet and hands. Occasionally the attacks were accompanied by a fever up to 101° F. Each episode lasted between two and three weeks. For many years she had suffered from asthma and hay fever. She had been admitted to hospital in October, 1945, when investigations showed no cause for her symptoms and radiographs of the chest were said to show no abnormality. At that time there was marked clubbing of the fingers. In June, 1946, her condition was worse. There was hard, pitting oedema of the lower limbs extending half-way up the leg and similar oedema of the wrists and forearms, with pain, tenderness, and swelling of the small joints of the extremities and well-marked clubbing of the fingers. The clinical picture was that of severe osteo-arthropathy and radiographs of the bones showed obvious periosteal reaction.

Radiographs of the chest were reported on as normal, but the left cupola of the diaphragm was higher than that on the right side. However, six months later, in December, 1946, further radiography demonstrated a rounded opacity at the left base which screening showed to be above the diaphragm. Bronchoscopy revealed narrowing of the left lower lobe bronchus.

At thoracotomy (Mr. N. R. Barrett) there were no pleural adhesions. The tumour was situated in the base of the pleural cavity, and was about the size of a grapefruit and quite smooth, except for one area which was nodular. It was entirely free except for a slender vascular pedicle by which it was attached to the pleura at the diaphragmatic surface of the lower lobe. The lower segments contained large pulsating vessels which supplied the tumour. The vascular pedicle was ligated and divided and the tumour removed. The patient made a good recovery and the pain and swelling in the limbs disappeared immediately following operation.

The specimen was a well-encapsulated tumour which was microscopically a cellular fibroma.

CASE 3.—A married woman, who was 45 years of age in 1939, had suffered from "rheumatism," chiefly in the hands, for about 12 years. She was very obese, and as she wished to reduce her weight radiographic examination of her chest was carried out before treatment. A small opacity, stated to be a cyst, was discovered in the right lung, but she was reassured about this and told to forget it. Five years later she felt vaguely unwell and the examination was repeated. There was said to be no change in the size of the opacity. Early in 1947 she was again radiographed and there was a marked increase in the size of the opacity. She was admitted to hospital, and bronchoscopy showed that the right lateral tracheal wall was displaced medially and the posterior wall about an inch above the carina was displaced forwards. The right main bronchus was reduced to a transverse slit. No tumour was seen. These bronchoscopic appearances suggested that the patient was suffering from an inoperable malignant tumour.

One year later, i.e., in March, 1948, she was still feeling reasonably well and maintaining her excess weight, but was suffering from increasing dyspnoea and the polyarthritis was more severe. The radiographic appearances were unchanged. By this time two cases of intrathoracic fibroma had undergone successful surgical operation and a review of this patient, known to have had an intrathoracic opacity for nine years which had become very much larger and was associated with polyarthritis, suggested that this, too, was an example of fibroma of the visceral pleura.

Thoracotomy was performed (C.P.T.) and the tumour was found in the main fissure. There were many large subpleural vessels. A line of cleavage between the lung and the neoplasm was found with some difficulty because of dense adhesions, and the latter removed. The patient made an excellent response to the operation and a few days later was able to take a ring off her finger which she had not been able to do for about ten years. Two months later she was feeling well, but once more she was unable to remove the ring. At this time there was a small opacity in the right hilar region which was thought to be due to a haematoma. Seven months after operation this shadow was still present. Four months after this she complained of recurrence of the arthritis, and the hilar opacity was definitely increasing in size. In April, 1949, the chest was reopened (C.P.T.). A large mass was found between the upper, lower, and middle lobes, no longer encapsulated, but invading the lung. There were several enlarged hilar lymph glands. Pneumonectomy was carried out and she was discharged one month later, again free from arthritis. The first tumour removed seemed to be encapsulated and was reported on as a fibroma, which was moderately cellular in places with some pleomorphism of the tumour cells. The report on the lung showed that there were multiple deposits of a tumour which was invading the lung substance. The histological appearances were similar to that of the first specimen, but there was increased cellularity and pleomorphism of the cells. There was clearly invasion of the lung by what appeared to be a spindle cell fibrosarcoma. The glands removed showed no evidence of metastases. Since the second operation, the patient has remained well.

In view of the recurrence after the first operation, it was felt that the disappointment in removing two lobes in Case 1 for a tumour which might have been excised alone was unjustified, and it was resolved that in future such lesions when adherent should be removed together with adjacent pulmonary tissue.

CASE 4.—A woman of 58 years of age attended the out-patient department in November, 1949, complaining of swelling and pain in the ankles, wrists, and fingers.

She volunteered the information that her thimble no longer fitted the tip of her middle finger. She had been treated for rheumatism for many years. Recently she had noticed increasing dyspnoea and non-productive cough with occasional pain in the upper part of the right side of the chest.

Radiographs showed a dense, clear-cut, homogeneous opacity in the upper part of the right hemithorax, which from its shape and position could well be caused by a tumour in the greater fissure.

The patent was bronchoscoped and this disclosed a bulge in the right lateral wall of the lower part of the trachea and that the right upper lobe orifice was almost occluded. A biopsy was taken from the orifice and was described as bronchial mucosa infiltrated by squamous cell carcinoma.

The bronchoscopic findings suggested that such a tumour would be ineradicable and accordingly the patient was submitted to radiotherapy. This made no difference to the patient or to the tumour.

The clinical and radiological findings were so characteristic of the condition with which by now we were familiar that a careful examination was made of the circumstances surrounding the original report of malignancy in the bronchoscopic biopsy. By mischance the specimen had been confused with that of a patient with undoubted carcinoma of the lung, but in whom bronchial biopsy gave negative findings.

Thoracotomy was carried out (C.P.T.), and on opening the chest there appeared to be a firm mass in the lung which dissection showed to be in the main fissure. The upper lobe was completely atelectatic and lying posteriorly against the mediastinum. The tumour was firm, white, and slightly lobulated and was easily dissected free. In two areas on the upper and lower lobes a sliver of lung was purposely removed in order to get well beyond the territory of the growth. This patient showed no change in joint symptoms after operation, because during prolonged bed rest before it, her pains had become less apparent. However, at no time had the joint symptoms been as severe as those in the three examples previously described.

Examination of the specimen showed the tumour to be a fibroma, which was moderately cellular but showed no invasive characteristics.

CASE 5.—This patient was an Egyptian of 56 years of age who was first examined in June, 1950. Four years before he underwent routine chest radiography and an opacity was noted in the right lower lobe. No treatment was undertaken, but when he came to England radiography was repeated. On this occasion there was a large oval shadow in the lower part of the right hemithorax. He had never complained of symptoms referable to the chest and he had no joint pains. There was slight clubbing of the fingers. Tests for hydatid infection were negative. Bronchoscopy showed no abnormality.

Thoracotomy was advised and at operation (C.P.T.) a large mass was found in relation to the pleural surface of the lower lobe projecting from the lung, but to a certain extent within its substance. Right lower lobectomy was performed and the patient made a good recovery.

The specimen showed a pale, nodular mass which appeared to be pushing the lung aside rather than invading it. Histological examination showed that, while the bulk of the tumour was encapsulated, at some points tumour cells had penetrated the capsule and were in contact with the lung. In parts the cells were very pleomorphic and mitoses were abundant with extensive necrosis. It was considered to be a fibrosarcoma.

CASE 6.—The last case in this series was a woman of 59 who may be claimed as a result of mass miniature radiography, but in fact she sought radiological examination because of a harsh, dry cough. The chest films showed a small opacity in the right hilar region but in the lateral view, where the fissures are particularly well marked, it is obvious that it could well be caused by a lesion in the fissure rather than in the lung. She gave no history of joint pains, there was no clubbing of the fingers, and no other abnormality on clinical examination or bronchoscopy.

At thoracotomy (C.E.D.) a lobulated tumour, roughly ovoid, was found in the fissure between the upper and lower lobes. It was about 3 cm. long and attached by a narrow friable pedicle to the visceral pleura of the under surface of the upper lobe, about 2½ cm. from the right upper lobe bronchus, although part of the tumour was resting against that structure. An attempt to dissect the tumour together with a small slice of lung tissue resulted in the mass tearing from the lung surface, so fragile was its attachment. This small tumour appeared to be a well-encapsulated fibroma.

CLINICAL FINDINGS AND DIAGNOSIS

Of the six patients in this series, four were women and all were over 55 years of age when submitted to operation.

Symptoms referable to the chest include pain, cough, and dyspnoea, but one patient was symptomless. Four complained of "rheumatism" and three were severely incapacitated by pain, swelling, and tenderness in the joints of the extremities, although all enjoyed occasional remissions. They showed in varying degree what is called hypertrophic pulmonary osteo-arthropathy and the joint symptoms in all but the first case described antedated those referable to the chest by many years. They were all dramatically relieved by operation. Joint pains returned in one patient in whom the tumour recurred and she was again relieved by pneumonectomy.

Osteo-arthropathy is a most dramatic and striking feature of the clinical picture. Other published case reports show the same association between it and fibrous tumours of the visceral pleura or lung (Clagett and Hausmann, 1944; Massachusetts General Hospital, Case 31271, 1945), but

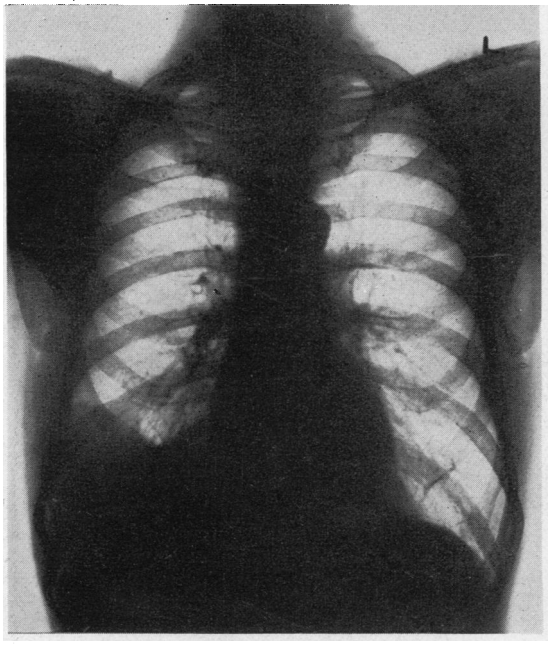

FIG. 1.—Case 1: radiograph showing opacity at right base. March, 1945.

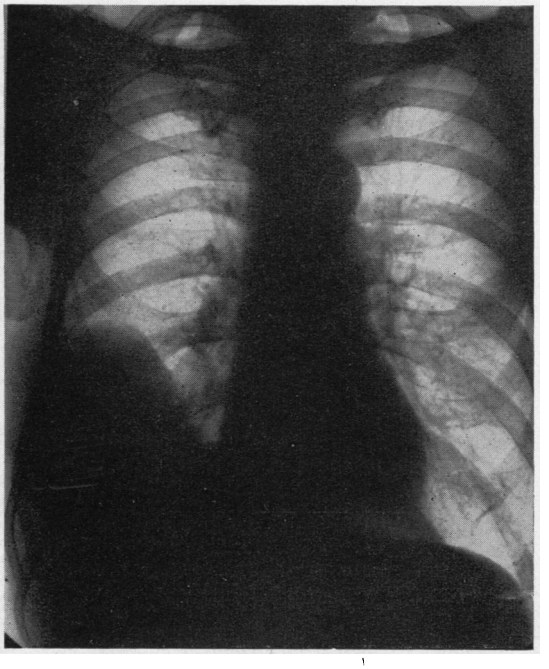

FIG. 2.—Case 1: radiographic appearance January, 1947.

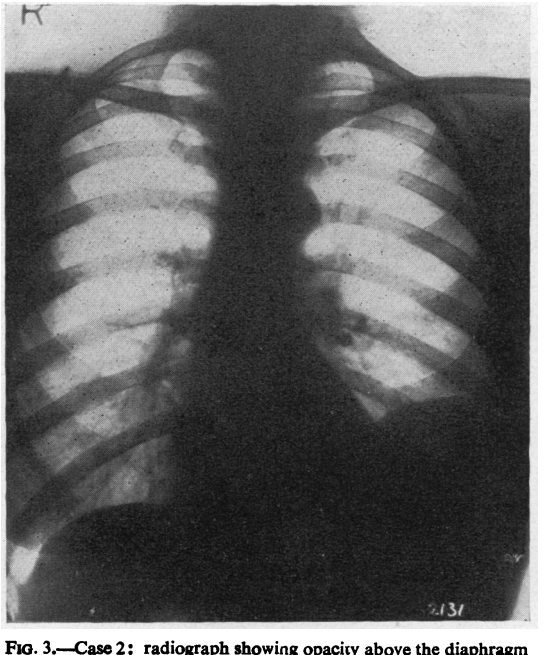

FIG. 3.—Case 2: radiograph showing opacity above the diaphragm on the left side.

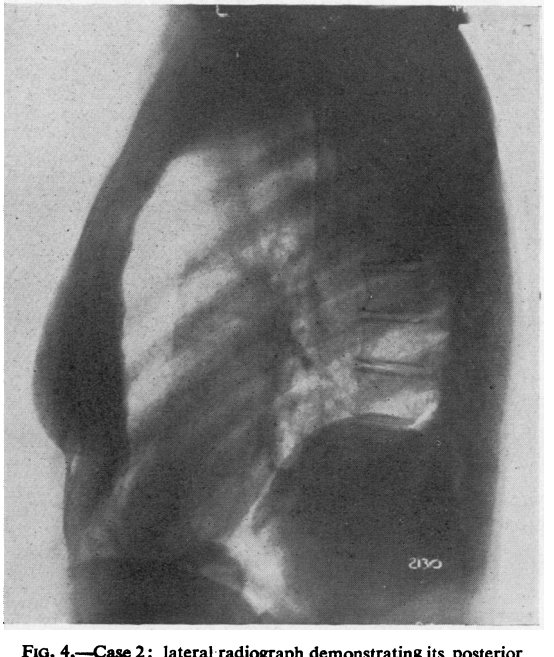

FIG. 4.—Case 2: lateral radiograph demonstrating its posterior position.

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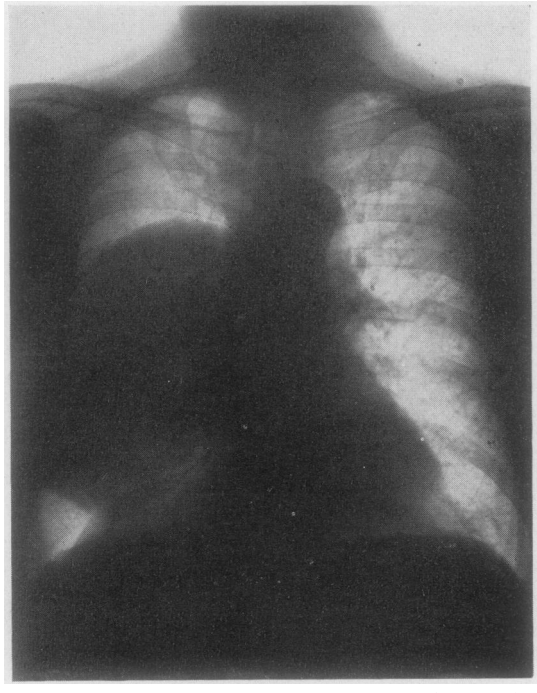

FIG. 5.—Case 3: radiograph showing oval circumscribed opacity, May, 1948. Diagnosed one year previously as a carcinoma of the lung which was bronchoscopically inoperable.

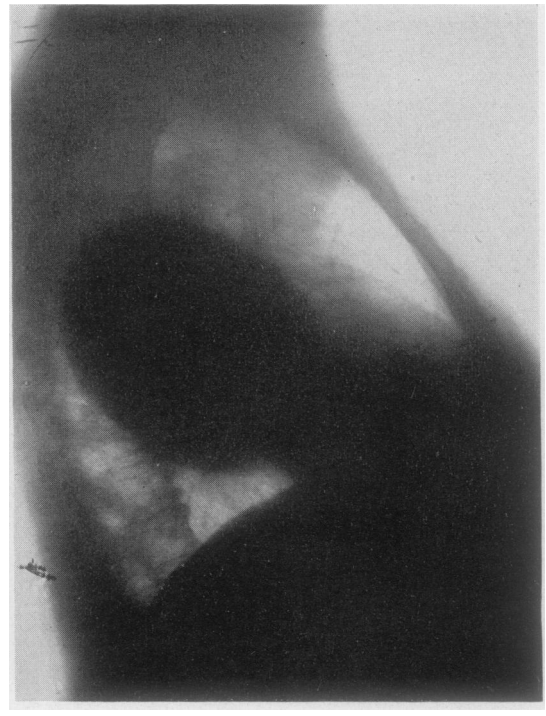

FIG. 6.—Case 3: lateral radiograph showing the tumour which was subsequently located in the fissure between upper and lower lobes.

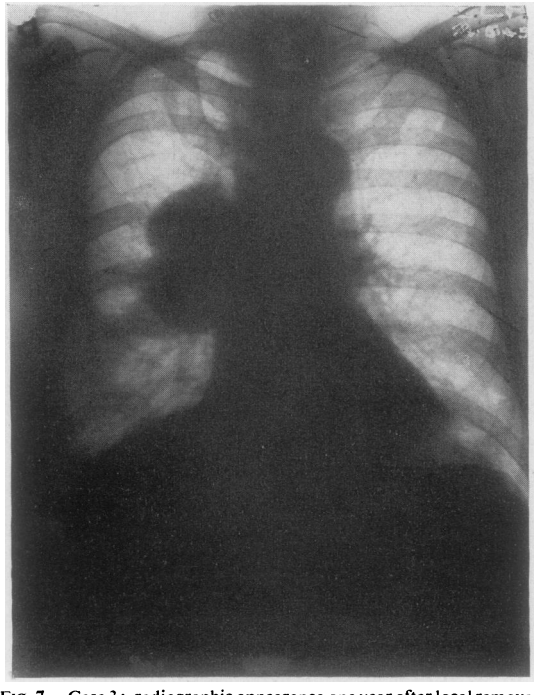

FIG. 7.—Case 3: radiographic appearance one year after local removal of the tumour.

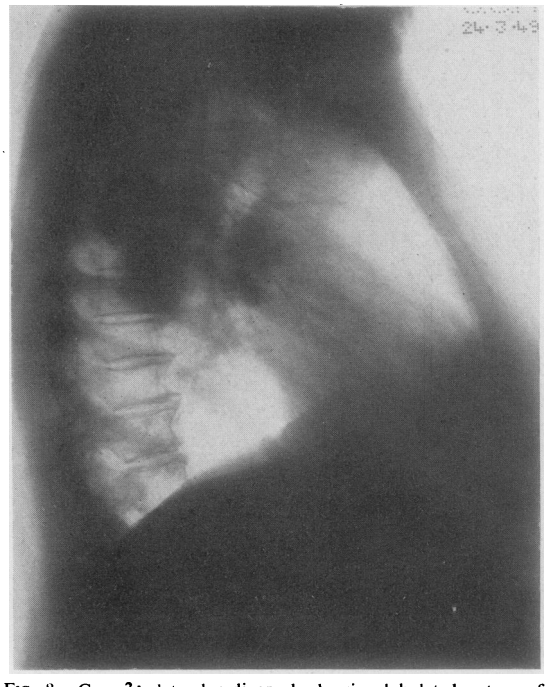

FIG. 8.—Case 3: lateral radiograph showing lobulated nature of the recurrence.

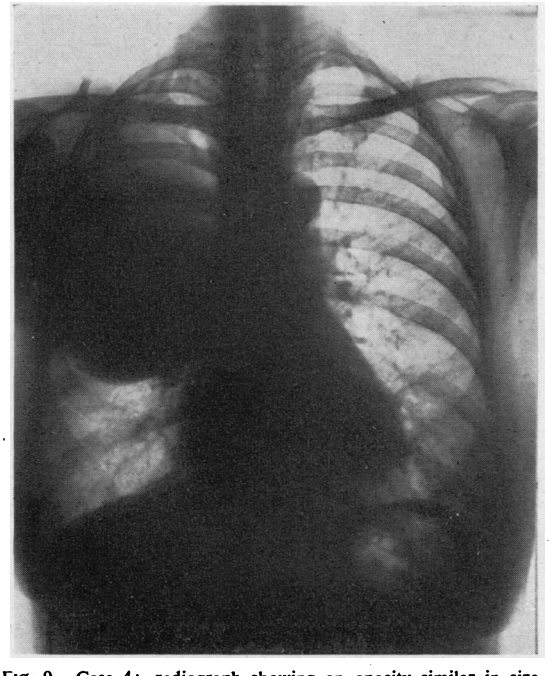

FIG. 9.—Case 4: radiograph showing an opacity similar in size, appearance, and position to that in Case 3.

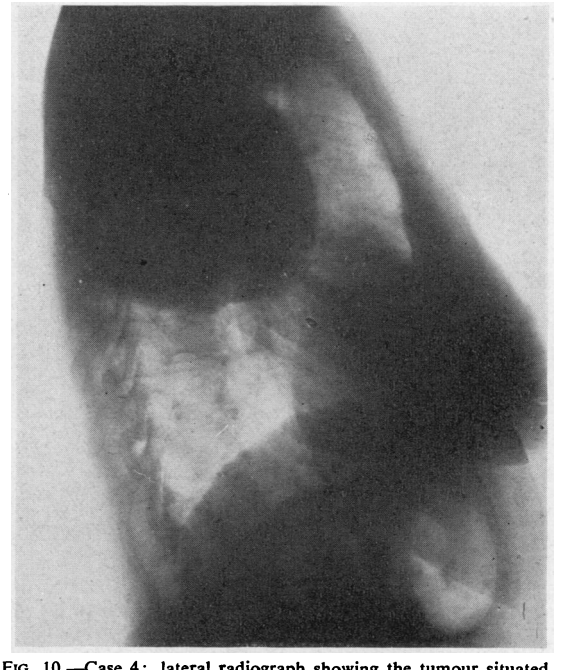

FIG. 10.—Case 4: lateral radiograph showing the tumour situated in the interlobar fissure.

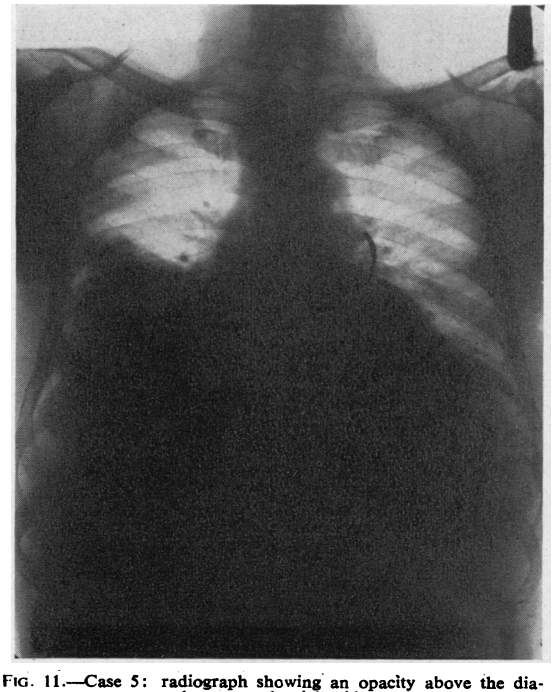

FIG. 11.—Case 5: radiograph showing an opacity above the diaphragm on the right side.

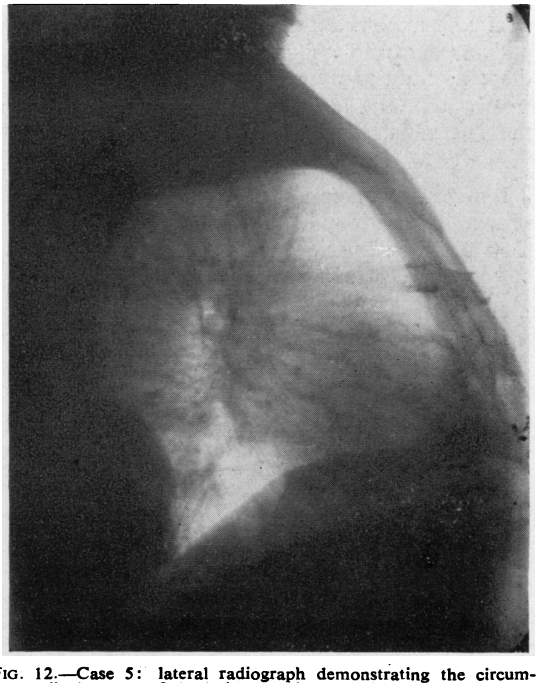

FIG. 12.—Case 5: lateral radiograph demonstrating the circumscribed nature of the lesion and its posterior situation, similar to that in Case 2.

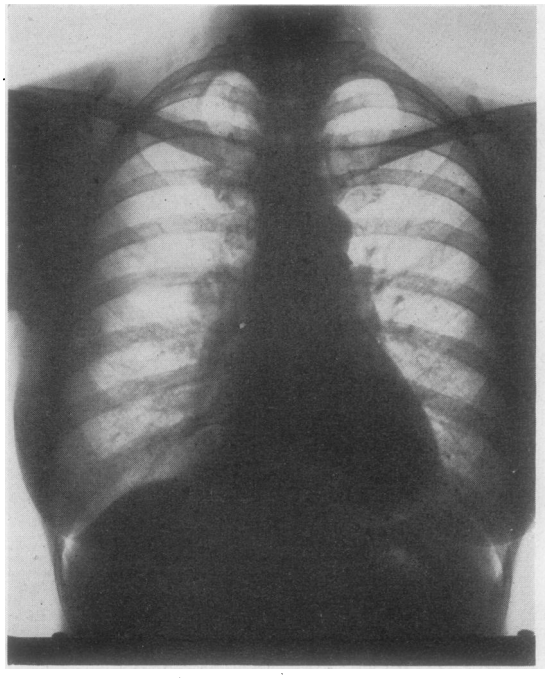

FIG. 13.—Case 6: radiograph showing a small opacity near the hilum of the right lung.

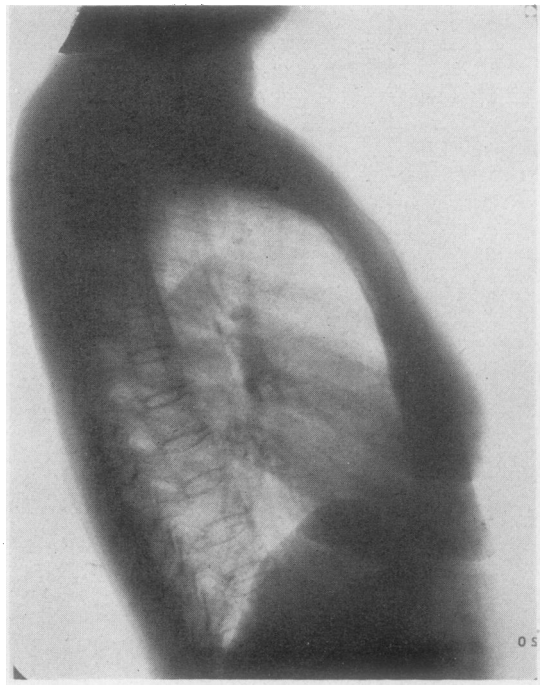

FIG. 14.—Case 6: lateral radiograph showing the lesion immediately behind and above the hilum. It was possible to identify the greater fissure in the original film, which suggested the location of the tumour subsequently found attached by a narrow pedicle to the under surface of the upper lobe.

it is not a constant finding. In our series, two patients were free from arthritis and in one of these the fingers were not clubbed, but it is noteworthy that the tumour which was removed from the latter patient was very small in comparison with the usual finding of a large mass.

Fawcett (1945) reported the removal of a neoplasm similar in position, histology, and size to that in Case 2, but his patient showed no clubbing of the fingers or arthritis.

Hypertrophic osteo-arthropathy has been described associated with various lesions within and without the thoracic cavity. It sometimes accompanies chronic intrathoracic infections and is commonly found in patients with carcinoma of the lung.

No lesion is as constantly associated with this phenomenon as that which is the subject of this paper. We cannot find the same link with fibrous tumours elsewhere in the chest, although this observation is based only on the available literature which suggests that they are even more rare than those arising beneath the visceral pleura. In our experience fibrous neurogenic tumours do not give rise to osteo-arthropathy.

The radiological appearance, when the tumour is large, is that of a dense, sharply circumscribed opacity, and on this evidence alone the correct pre-operative diagnosis cannot usually be made, except that when the mass arises in a fissure its position, particularly from the lateral aspect and its ovoid shape with the long axis in the direction of the fissure, might suggest a pleural tumour. In two cases the radiological abnormality was at first missed because it presented in the floor of the hemithorax and its outline merged with that of the diaphragm. The radiographs do not suggest the slight lobulation which is almost always present. Bronchoscopy was helpful in excluding intrabronchial lesions, but in two cases in which the tumour arose in the upper part of the thoracic cavity, distortion of the trachea and right main bronchus suggested inoperability had this been associated with carcinoma of the lung.

The diagnosis can usually be established when the chest is opened. Two specimens, covered with extensions of the visceral pleura and attached by narrow pedicles to the lung, were easily removed, but a tumour presenting in a fissure, particularly when adherent to lung folded over it, may be mistaken for an intrapulmonary lesion.

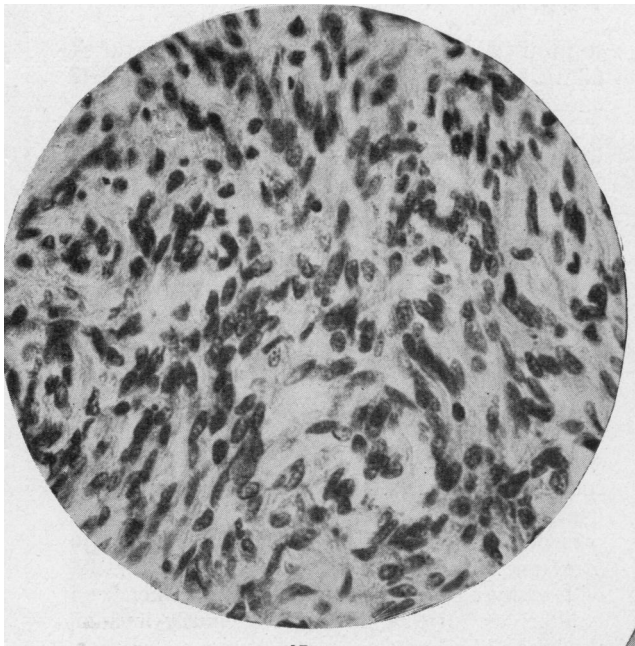

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FIG. 15.—Case 1: photomicrograph showing cellular area without collagen fibres.

FIG. 16.—Case 2: photomicrograph showing few cells with masses of collagen.

FIG. 17.—Case 2: photomicrograph showing sharp transition from a fibrous to a cellular area.

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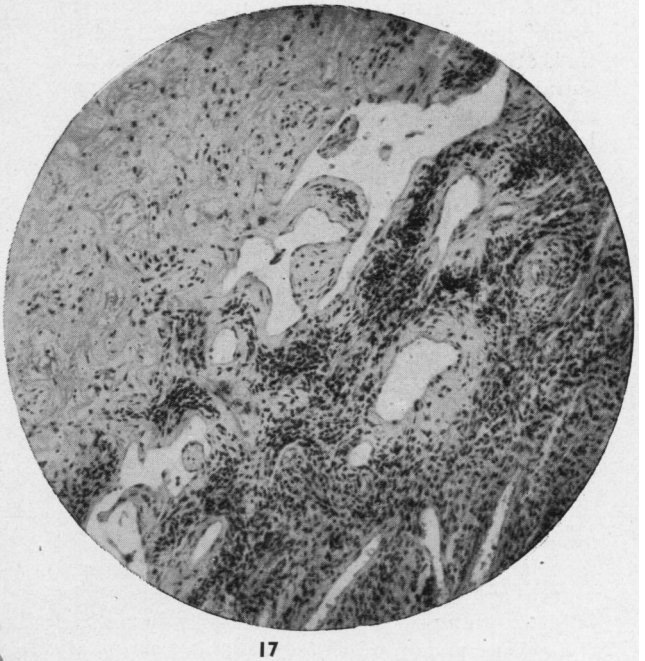
PATHOLOGY

Klemperer and Rabin (1931) divided pleural neoplasms into localized and diffuse types and suggest that the former arise from the tissues beneath the superficial flattened cells of the pleura. These authors collected from different sources 16 examples of so-called giant sarcoma of the visceral pleura, some of which caused death by vascular and cardiac compression. In each case the tumour was enclosed in a capsule covered by a smooth membrane continuous with the visceral pleura. Some were adherent to surrounding structures, making it difficult to establish the exact place of origin. They were variously described as



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fibroma, fibrosarcoma, myxofibrosarcoma, spindle cell sarcoma, but in no case was there evidence of metastasis. Four examples from the authors' experience are described in detail, two of which are classified as fibrosarcomata.

Lilienthal completely removed one tumour which microscopically was a cellular fibroma, but because the capsule was infiltrated in a few places by tumour cells it was considered that it should be more correctly termed a fibrosarcoma. The same surgeon was unable to remove the tumour in the second case. It was large, hard, and fixed, but a small piece about the size of a tennis ball was excised and shown to be a fibrosarcoma similar in structure

to that in the first patient. Four years later further fragments of tumour were removed at thoracotomy, and showed a startling change in histology to that of a highly malignant neoplasm. At this time there was radiological evidence of secondary deposits in the other lung.

The tumours which we have removed are similar in most respects to the localized fibrous neoplasms described by Klemperer and Rabin. On macroscopical examination five out of six appeared to be benign, easily removed from the chest, and apparently well encapsulated. Their origin beneath the visceral pleura was well demonstrated. One tumour recurred and was then obviously infiltrating the lung necessitating pneumonectomy. At the first operation the mass was adherent to the upper lobe and it is assumed that the tumour was incompletely removed. The sixth specimen showed the bulk of the tumour as an excrescence on the lung surface, but part was intrapulmonary and invasive, so that it was thought advisable to perform lobectomy.

The histological appearances in all the examples quoted displayed similar characteristics. In any one tumour some parts appear benign, acellular with masses of collagen, yet other areas may show necrosis with marked cellularity and pleomorphism suspicious of malignancy. Unless there is obvious permeation of the capsule, it is impossible to say whether the lesion is benign or malignant. It seems likely that they behave as simple tumours for a very long time, but that they may become locally invasive, particularly after surgical intervention, as suggested by Klemperer and Rabin.

We have described these neoplasms as fibroma or fibrosarcoma, although it is realized that only the passage of time will decide how many of those thought to be benign will recur in a malignant form.

Stout and Himadi (1951) collected eight examples of fibrous tumours arising in relation to the parietal and visceral pleura, which they described as solitary (localized) mesothelioma. Of these, two closely resembled the tumours which we have described in that they were connected to the lung by a pedicle and were microscopically identical. We prefer not to use the suggested terminology which implies the existence of a multiple or diffuse type of tumour, which, as far as we know, has not been described. The term mesothelioma (endothelioma) has been so misused, particularly in the nomenclature of the diffuse type of pleural tumour, that until greater numbers of pleural neoplasms have been studied it would be preferable to describe them in terms suggestive of their predominant constituent tissue. Klemperer and Rabin used the term mesothelioma only for a diffuse type of tumour, and Willis (1948) doubts whether such an entity exists.

Treatment

In a short experience of six cases seen during the past five years it seems obvious that treatment should be directed at removal of the tumour, even in the absence of symptoms. It is potentially malignant by local invasion and may reach a size which could cause death by compression of surrounding structures. The disability caused by hypertrophic pulmonary osteo-arthropathy when present is completely relieved by its extirpation. The tumour should be removed completely together with a sliver of lung or parietal pleura wherever it is adherent. If the tumour is obviously invading the lung, lobectomy or even pneumonectomy may be required. One patient who was inadvertently subjected to radiotherapy did not react to the treatment.

Summary

Six patients with fibrous tumours of the visceral pleura were treated surgically, the tumour alone being removed in four while lobectomy was required in the remaining two cases. One tumour which was incompletely removed subsequently recurred and pneumonectomy was carried out. No other recurrence has been observed, but the longest period of observation is five years. Four patients who showed hypertrophic pulmonary osteo-arthropathy were dramatically relieved by operation.

Our thanks are due to Mr. N. R. Barrett for permission to publish Case 2 and to Dr. A. Morgan and Dr. J. Clegg for pathological reports.

References

Clagett, O. T., and Hausmann, P. F. (1944). *J. thorac. Surg.*, **13**, 6.
Fawcett, A. W. (1945). *Brit. med. J.*, **2**, 425.
Harrington, S. W. (1950). *Penn. med. J.*, **53**, 1164.
Klemperer, P., and Rabin, C. B. (1931). *Arch. Path., Chicago*, **11**, 385.
Massachusetts General Hospital (1945). Case 31271. *New Engl. J. Med.*, **233**, 18.
Stout, A. P., and Himadi, G. M. (1951). *Ann. Surg.*, **133**, 50.
Willis, R. A. (1948). *Pathology of Tumours*, 1st edit., p. 646. Butterworth, London.