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False aneurysm of the pulmonary artery with peripheral venous thrombosis

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Aneurysms of the pulmonary artery are usually of its extrapulmonary portion (Boyd and McGavack, 1939; Deterling and Clagett, 1947). Intrapulmonary aneurysms are less common, and, when present, are usually mycotic.

A rare association of aneurysms of medium-sized branches of the pulmonary artery, venous thrombosis of the legs, and cerebral venous sinus thrombosis as part of a systemic illness has been described (Pirani, Ewart, and Wilson, 1949; Hughes and Stovin, 1959). Pirani *et al.* (1949) thought the process in the pulmonary artery was mycotic from infected emboli originating in the leg veins. Hughes and Stovin (1959) also associated the pulmonary aneurysm with embolism of the pulmonary vessel in the presence of abnormal bronchial arteries.

This case report is of a similar triad, leg vein thrombosis, sagittal sinus thrombosis, an aneurysm of a branch of the left superior pulmonary artery, and, in addition, an aneurysm of a systemic artery.

We suggest that these lesions are due to a 'collagen disease'.

CASE REPORT

A.J., a man of 29 years, was well until January 1961, when he felt listless and tired easily. During the next few months he had several episodes of 'influenza' consisting of fever, generalized aches and pains, a sore throat, and often a dry cough. By October of the same year he complained of loss of weight, anorexia, and night sweats. On December 19 he was admitted to Bridgend General Hospital with a deep-vein thrombosis of the right leg which was treated with anticoagulants until he was discharged in January 1962. He was readmitted one month later with lower abdominal and right loin pain and severe headache, accompanied by intense photophobia and neck stiffness of two weeks' duration. A few days before this readmission he had coughed up some blood on one occasion and may have passed some blood in his urine. At this time he had an irregular fever up to 101° F. Clubbing of the fingers was noted, but he stated that this had been present all his life, and his only brother was healthy and clubbed. He had considerable neck stiffness, a positive Kernig sign, and bilateral papilloedema with a small haemorrhage close to the right disc. The cerebrospinal fluid pressure was raised (300 mm. of water) but the fluid showed no increase in cells or protein; the Lange test was normal and the W.R. was negative. Blood counts showed a progressive anaemia with a persistent leucocytosis between 10,000 and 15,000/c.mm., predominantly polymorphonuclears; eosinophils were never recorded above 2%. The E.S.R. was 50 mm. in one hour. The serum alkaline phosphatase was 28·4 K-A. units, the serum albumin 2·3 g./100 ml. and the serum globulin 4·2 g./100 ml., but the other liver function tests were normal. The Rose Waaler test was weakly positive. Urine examination, blood urea, tests of bleeding and clotting function, blood and bone marrow cultures, Widal and virus agglutinations, and repeated examinations for L.E. cells were all negative. The electroencephalogram showed phase reversal of slow waves occurring symmetrically about the mid-line and transverse leads, suggesting that the abnormal process lay along the sagittal plane. It was thought that the intracranial findings were due to thrombosis of the superior saggital sinus.

In March 1962, the patient developed some red, tender swellings on the legs and a pulsatile swelling near the angle of the right jaw. The latter was explored surgically in the Cardiff Royal Infirmary and was found to be an aneurysm of the right external carotid artery. The closely related superior thyroid artery and small related lymph nodes were biopsied. The superior thyroid artery was histologically normal, but small arterioles showed fibrinoid change, and small venules were infiltrated by neutrophil polymorphs (Fig. 1). The lymph nodes showed peculiar foci of necrosis, and around this area several haematoxylin bodies were seen (Fig. 2). An attempted biopsy of a nodule on the leg and a pectoral muscle biopsy revealed no arteritis. A pericardial friction rub was heard from time to time, and the chest radiograph showed fluid in one costo-phrenic angle which cleared within two weeks. Treatment with prednisone, 60 mg./day, was begun, and there was considerable improvement. However, when the dosage was reduced four months later, he became breathless, had abdominal pain, the pain and swelling in the legs returned,

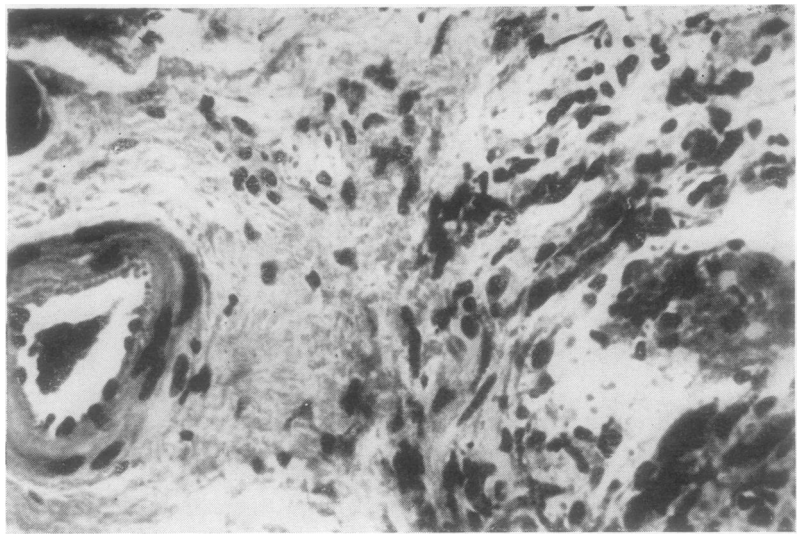

FIG. 1. *Shows fibrinoid change in arteriole (on the left) and a vein with infiltration by inflammatory cells (on the right). H. and E.* × 120.

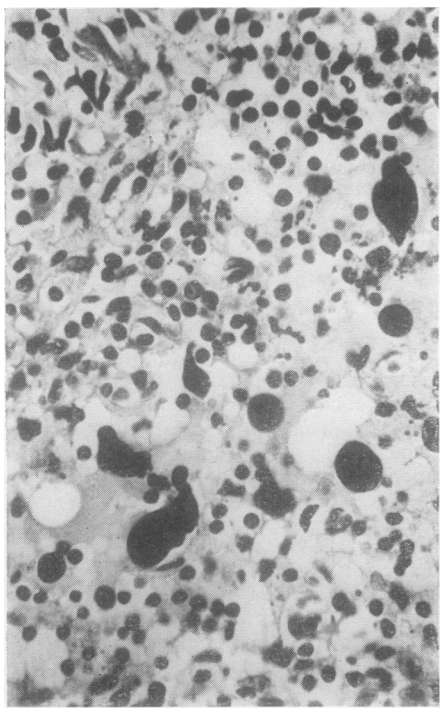

FIG. 2. *Lymph node biopsy revealing several haematoxylin bodies. H. and E.* × 140.

and he had several small haemoptyses. There was again fever to 101° F. and the E.S.R. was 105 mm. in one hour. A barium meal, barium enema, and an intravenous pyelogram were performed at this time and all were normal. As soon as the prednisone dosage was increased, there was considerable subjective and objective improvement. Anticoagulants were also recommenced at this time.

In October 1962, he suddenly coughed up about one pint of bright red blood. His general condition remained good and his blood pressure was 120/80 mm. Hg ; he had always been normotensive. The chest radiograph then showed a round opacity above the left hilum with a less well-defined opacity in the anterior segment of the left upper lobe (Figs. 3 and 4). Subsequent radiographs and tomography, taken at Sully Hospital, showed that the peripheral opacity was clearing and that the rounded opacity was related to a branch of the left superior pulmonary artery. Since October there had been some ill-defined shadows in the right upper lobe and left lower lobe which altered during the succeeding months. It was thought that the opacity above the left hilum was an aneurysm of a branch of the left pulmonary artery and was the cause of the haemoptysis. The segmental opacity was considered to be due to aspiration of blood. As there were no signs clinically of active disease, he was afebrile, and the E.S.R. and serum globulins were normal, it was considered that if this aneurysm were to bleed again it might prove fatal and that his immediate prognosis was dependent upon the fate of the aneurysm rather than the activity of the disease. It seemed unlikely that the lesions in the right upper lobe and left lower lobe were also due

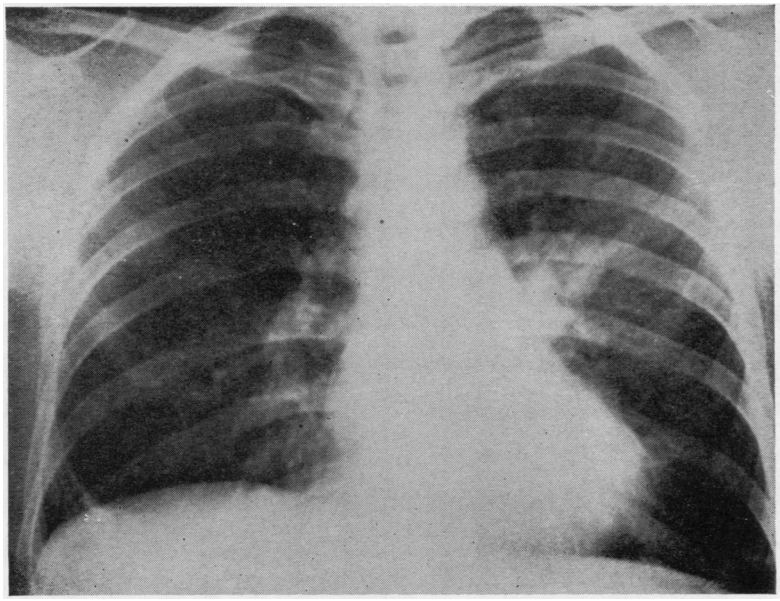

FIG. 3. *Shows a 'round opacity' above the left hilum and an ill-defined peripheral opacity.*

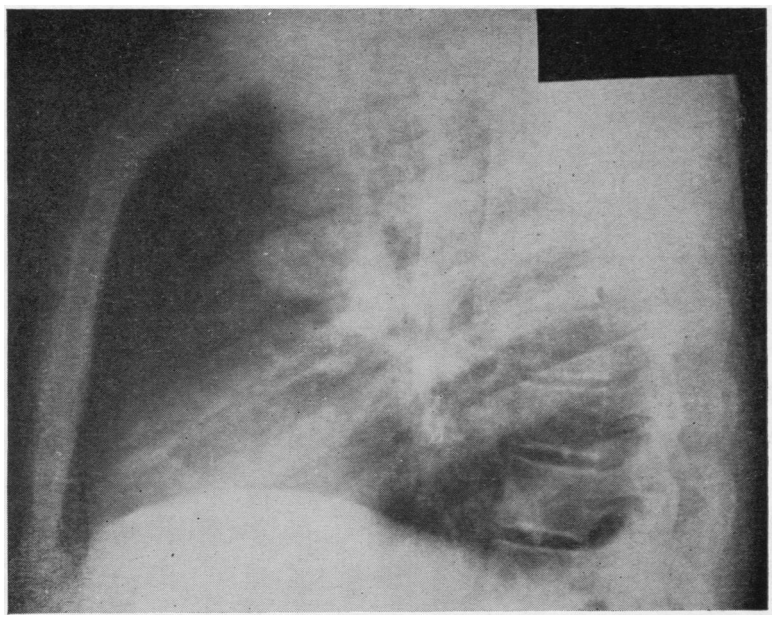

FIG. 4. *Left lateral radiograph showing the round opacity related to a branch of the pulmonary artery, and a related anterior segmental lesion.*

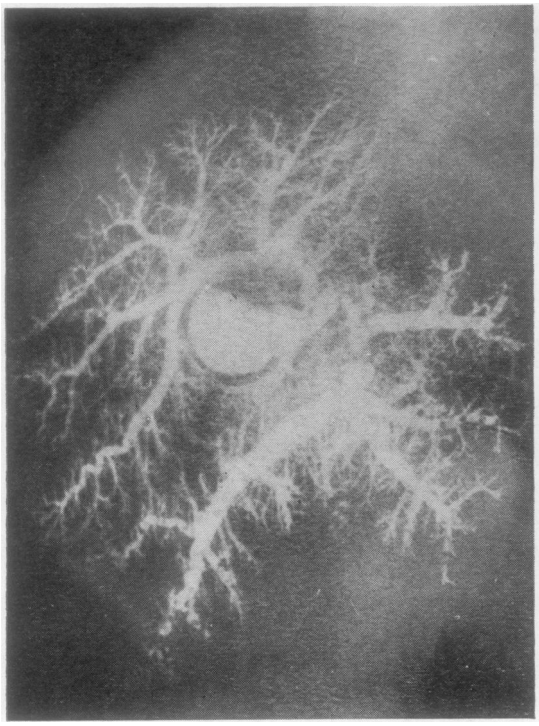

FIG. 5. *Radiograph of a lobectomy specimen after injection of the segmental arteries and veins with radiopaque material showing the filling of a false aneurysm.*

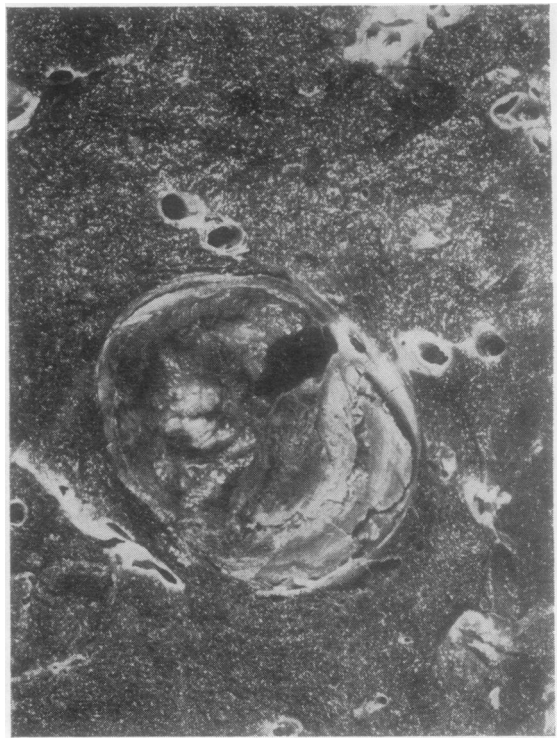

FIG. 6. *Photograph of the false aneurysim with laminated thrombus at one edge.*

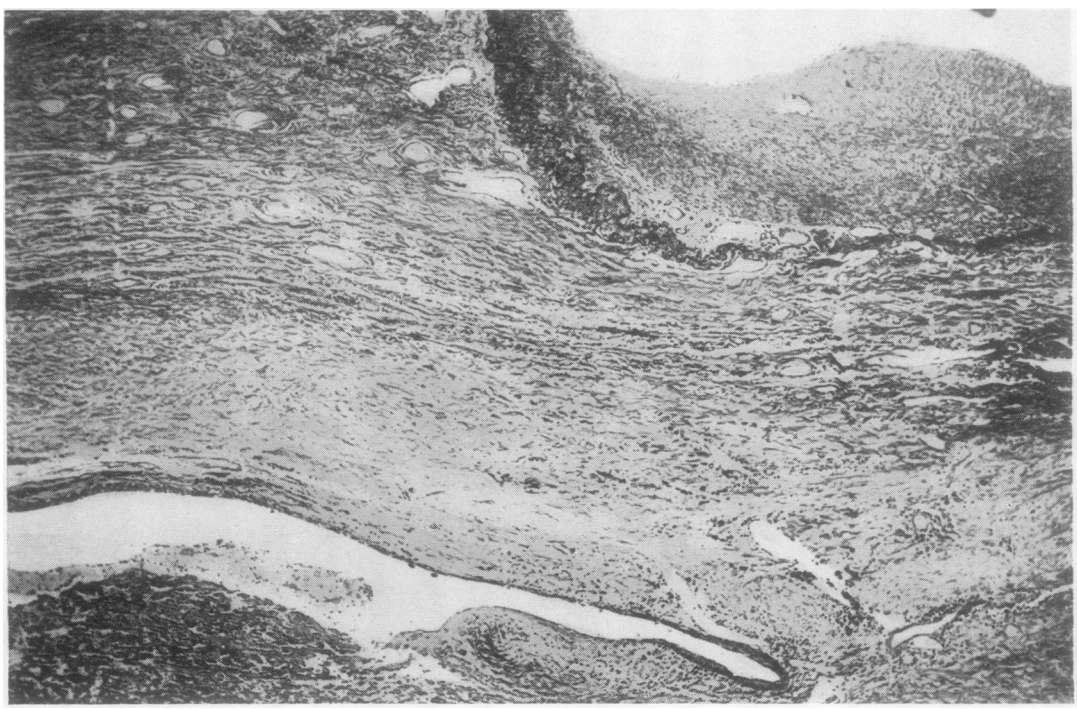

FIG. 7. *Shows the edge of the false aneurysm (below) encroaching on a bronchial lumen and a branch of the pulmonary artery (above) with disruption of the elastica. Elastic and van Giesen* × 40.

to the aneurysm, as these lesions were shown to be ill-defined, lying peripherally in the lung, and were not related to any vascular structures.

A left thoracotomy was performed on December 18, 1962, when a pulsatile mass was felt in the left upper lobe, and the whole lobe was resected. The lesion in the lower lobe felt fibrotic and was not pulsatile. His post-operative course was uneventful.

The veins and arteries of the lobectomy specimen were injected with radiopaque material before fixation (Fig. 5) and showed that the lesion was aneurysmal, arising from a branch of the pulmonary artery, and that there was no connexion with the pulmonary veins. The cut surface of the lobe (Fig. 6) revealed the aneurysm, 2 cm. in diameter, with much laminated thrombus at the edge. The rest of the lobe was normal. Microscopically the aneurysm proved to be a false aneurysm with a segmental disruption of the elastica of the pulmonary artery of origin. The clot itself was largely outside the vessel and much of its wall was an expanded bronchus. The organizing thrombus was separated from the bronchial lumen by a thin layer of respiratory epithelium, and in places squamous metaplasia had occurred. Figure 7 shows the clot protruding into the lumen of the bronchus, and, above, a branch of pulmonary artery showing intimal thickening and segmental disruption of the elastica. There were haemosiderin-laden macrophages in the rest of the lobe, which were otherwise normal. No abnormality of the bronchial arteries was noted, and there were no changes of pulmonary hypertension in the arterioles.

DISCUSSION

Our patient suffered from a generalized disease affecting the systemic and pulmonary arteries and large veins. The involvement of leg veins, venules, and saggital sinus and the association of systemic and pulmonary aneurysms and repeatedly negative blood and bone marrow cultures make a mycotic process seem unlikely. The negative Wassermann reaction and the histology of the lesions deny a syphylitic aetiology, and no congenital lesions were found.

In the absence of other explanations and in the presence of aneurysms of both the systemic and pulmonary circulations, it was natural that polyarteritis nodosa should be considered. This diagnosis is supported by the segmental disruption of the elastica with intimal thickening, in the absence of evidence of infection and pulmonary infarction, the weakly positive Rose Waaler test, and the presence of haematoxylin bodies in the lymph node in a patient with a systemic illness responding to steroids. However, in polyarteritis nodosa with lung involvement there is generally a preceding respiratory illness, usually a blood eosinophilia, necrotizing pulmonary lesions, and granulomatous lesions sometimes with giant cells, and occasionally systemic hypertension (Rose and Spencer, 1957). Our patient had none of these, with the possible exception that the ill-defined lesions in the left lower lobe and right upper lobe might have been granulomatous. The picture presented does not comply with that of polyarteritis nodosa with lung involvement described by Rose and Spencer.

The evidence in favour of a 'collagen disease' appears sufficient for this to be considered in relation to our patient, but what makes the picture so atypical is the presence of venitis microscopically and thrombosis of the veins of the leg and cerebral sinuses, features not associated with a collagen disease. Of the previous similar cases, one of those described by Hughes and Stovin may be explained by a similar mechanism, as there was involvement of both pulmonary and common carotid arteries and segmental disruption of elastica, consistent with healed polyarteritis nodosa, and the systemic illness responded to steroids.

SUMMARY

A case of pulmonary artery aneurysm, with a preceding systemic illness, in which peripheral venous thromboses and signs of raised intracranial pressure predominated, is presented and illustrated.

It is considered that the lesions are part of an unusual collagen disease.

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