metabolism, and the syndrome of idiopathic pulmonary haemosiderosis.

It has been postulated that the intrinsic structure of the pulmonary vessels and the alveolar framework is defective, but such a view could not be adequately confirmed in this particular case. Histological sections from grossly affected parts of the lungs stained with special elastic stains show elastic tissue in apparently normal amounts. It must, however, be admitted that for several reasons it is extremely difficult to assess any apparent changes in the elastic structure of the lungs and lung vessels. The elastic elements are in themselves very small, and from their intrinsic property of elasticity the range of normal variation is wide and dependent upon a variety of environmental circumstances. When the alveoli are distended the elastic fibres seem thin, fibrillary, and scanty, whereas in the collapsed alveolus they are thick and rod-like. Stellate aggregations may appear when the elastic fibres are ruptured and contract irregularly to a maximal extent.

It is not suggested that there is any parallel between the modes of production of pulmonary infarction and idiopathic haemosiderosis, but it is interesting to note that, in suitably stained sections of infarcted lung, changes in the elastic tissue such as those described above may be found. No doubt the changes are partly the result of simple mechanical damage produced by alveolar overdistension in the infarcted area.

In view of these observations it is difficult to accept without considerable misgivings the theory that idiopathic haemosiderosis is due to a defect in the lung structure. Possibly the so-called "fragmentation" of elastic fibres and the various fibrous thickenings of the alveolar walls which have been described elsewhere represent the result rather than the cause of the disease, especially since they are noted in the post-mortem reports relating to the more chronic cases of the disease, where reactive and degenerative changes might be expected. Furthermore, it may perhaps reasonably be speculated whether in some reports of this disease there has been insufficient consideration of the range of normality in the elastic content of lung tissue.

Gumpert (1947) has described non-occupational pulmonary haemosiderosis in association with the long-standing pulmonary congestion found in a case of mitral stenosis. Some unrecognized additional factor must obviously operate to account for the rarity of gross haemosiderosis associated with mitral stenosis as compared with the great incidence of mitral stenosis, but by analogy might one speculate whether periodical increases in pulmonary arterial pressure of unknown cause might operate during the development of the idiopathic disease? The manner in which there are radiological indications of greater iron deposition near the hilar regions is reminiscent of the distribution of the hilar shadowing due to vascular congestion and pulmonary hypertension in early left ventricular failure. No myocardial or vascular cause for pulmonary hypertension was, however, recognized in the present case.

Wider recognition of the syndrome will no doubt permit its more extensive study and enable a more satisfactory explanation to be developed.

I am grateful to Dr. T. E. Gumpert for permission to report this case, which was admitted under his care, and to Dr. J. L. Edwards, who carried out the post-mortem examination. I should also like to thank Drs. J. L. Grout and T. Lodge, who helped me with the radiological interpretation, and Dr. A. Jordan for his assistance with the biochemical investigations. The radiographs are reproduced by courtesy of Dr. Grout.

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DISSEMINATED OSSIFICATION OF THE LUNGS IN ASSOCIATION WITH MITRAL STENOSIS

BY

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[WITH PHOTOGRAVURE PLATE]

Heterotopic ossification is a well-known phenomenon occurring in foci of pathological calcification in many parts of the body. In the lungs ossification of calcified tuberculous lesions in the apices has been frequently observed. Much less common are cases of disseminated pulmonary ossification, which occurs in two forms: (*a*) trabecular, found in the interstitial tissue as a senile change, and (*b*) nodular circumscribed, observed in younger persons suffering from mitral disease associated in most cases with chronic passive congestion. The nodular form is extremely rare, fewer than 40 cases having been recorded in the literature. A further case is here described.

Case Record

The patient, a single woman aged 39, was admitted to the West Middlesex Hospital on March 3, 1947. For nine years she had suffered from gradually increasing effort dyspnoea, with occasional swelling of the ankles. During the month before admission she had had a cough productive of greyish-white sputum. In the last four days the cough and dyspnoea had greatly increased. She had had rheumatic fever at the age of 20. There was no previous history of pneumonia.

On examination the patient was cyanosed and mildly dyspnoeic. The temperature was 101.6° F. (38.7° C.), the pulse rate 120, and the respiration rate 32. Considerable cardiac enlargement was found, the apex beat being situated in the sixth space 5 in. (12.7 cm.) from the midline. Mitral presystolic and early diastolic murmurs were heard, and there were occasional extrasystoles. There was no evidence of an aortic lesion. The blood pressure was 125/80. No peripheral oedema was seen, but the jugular venous pressure was 3 cm. above the sternal angle with the patient tilted 30 degrees from the horizontal. Examination of the lungs showed signs of consolidation at the right base, with a pleural rub in the right axilla. Scattered rales were heard over the left lung. In the abdomen slight tenderness was found in the right hypochondrium, but the viscera were not palpable. No abnormality of the central nervous system was detected.

It was considered that the patient was suffering from right basal pneumonia and early cardiac failure due to mitral stenosis, and that most of her dyspnoea was caused by the former.

A blood examination on admission showed: haemoglobin, 94%; leucocytes, 12,000 (polymorphonuclear cells 85%). The sputum yielded mixed organisms. A Mantoux test was negative. Radiographic findings are discussed below.

On admission the patient was treated with sulphamezathine (30 g. in four days), oxygen, sedatives, and a linctus. Two days later a course of intramuscular penicillin was begun, 2,400,000 units being given in eight days. In spite of chemotherapy the temperature never settled, but varied from 97 to 100° F. (36.1 to 37.8° C.), rising terminally to 103° F. (39.4° C.). Dyspnoea was at first greatly relieved.

On the tenth day after admission numerous rales and scattered areas of tubular breathing were heard on both sides of the chest, and the sputum had become blood-stained. The liver was palpable 1 in. (2.5 cm.) below the costal margin. Dyspnoea was not in evidence while the patient was at rest, but the respiration rate varied from 24 to 32 a minute. The possibility of infarction of the lungs was considered. Three weeks after admission a generalized scarlatiniform rash appeared, ascribed to drug sensitivity. It faded in three days. The general condition showed little change.

On the twenty-eighth day after admission the patient passed a comfortable day, but at 11 p.m. she suddenly collapsed, dying within a few minutes.

Radiological Reports.—March 6:—Both middle zones showed impairment of a congestive type, more pronounced on the right side. Both lung fields were infiltrated with generalized miliary shadowing, more intense in the basal zones (Plate, Fig. 1). The nodules were discrete and averaged 0.5 to 2 mm. in diameter. No pleural effusions were seen. The heart was enlarged in its transverse diameter and had a contour suggestive of mitral stenosis. *March 26:*—An effusion had developed in the right costo-phrenic angle. The miliary opacities remained unchanged, showing no tendency to resolution or confluence. The tentative diagnosis of haemosiderosis was made.

Necropsy

Macroscopic Appearance.—The subject was well nourished and showed slight oedema of the lower extremities. About 300 ml. of clear straw-coloured fluid was found in each side of the thorax. The heart weighed 570 g. and exhibited gross right ventricular hypertrophy. Both auricles were hypertrophied and dilated, a large ball thrombus being found in the left auricle. The tricuspid valve admitted four fingers. Thickening of the cusps of the aortic valve was present, their edges being adherent and giving rise to some stenosis. The mitral cusps were thickened and shortened, with adherent edges leading to stenosis of the funnel type. Thickening of the chordae tendineae was pronounced. The lungs showed cyanotic induration. Numerous large and small infarcts were evident, especially at the bases. Some were suppurative. The pleurae were studded with miliary calcified plaques 1 to 2 mm. in diameter. Nodules of bone in the lung parenchyma, described below, were neither seen nor felt. The spleen was enlarged, hard, and dark red, and there was moderately severe chronic congestion of the liver. There was also congestion of the mucosa of the stomach and of the renal parenchyma. The brain was not examined.

Microscopical Appearance.—Well-marked chronic congestion was seen in the lung substance. The interstitial tissue carrying the vessels showed some fibrous increase, with large numbers of pigment-carrying cells. Throughout the lung substance were seen numerous rounded nodules of bone with irregularly crenated outlines, many showing a concentric lamellar appearance (Fig. 2). The bone was mostly well calcified, but many nodules had a calcified centre with peripheral additions of osteoid tissue. This was well seen in the subpleural plaques, which showed an inner calcified and an outer osteoid zone. Vessels were present in the bone, but no medullary cavities were seen. The size of the nodules was several times that of an alveolus. A striking feature was that most of these nodules had no fibrous surround and under the microscope gave the appearance of lying free in the lung substance. In some areas, however, the bone was embedded in fibrous tissue containing pigment-laden phagocytes. The vessels in the interstitial tissue of the lungs showed no abnormality.

Discussion

The aetiology of the condition is obscure, and is likely to remain so until more cases are discovered. Past rheumatic infection with advanced mitral stenosis is invariable. Cases which have come to necropsy have all shown some degree of chronic passive congestion of the lungs. The question arises whether this alone can give rise to disseminated ossification or whether another factor—e.g., rheumatic pneumonia—is necessary to produce foci of necrosed or poorly vascularized tissue in which calcification and ossification can take place.

In the above case there is not sufficient histological evidence to warrant the presence of any other aetiological condition than chronic passive congestion. It is suggested that the bone may arise from organization of congestive haemorrhages or of intra-alveolar collections of pigment-bearing phagocytes. It should be appreciated that rheumatic pneumonia as an entity is not universally accepted.

My thanks are due to Dr. M. M. Deane for permission to publish the case, to Dr. A. C. Counsell for the pathological report and for advice and criticism, to Dr. D. G. Arthur for the radiograph, and to Mr. D. A. Vinten for the photograph.

PULMONARY LESIONS IN RHEUMATOID ARTHRITIS

BY

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AND

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[WITH PHOTOGRAVURE PLATE]

The paper by Ellman and Ball (1948) on "rheumatoid disease" with lung lesions will no doubt provoke many observers to report similar experiences. We have so far regarded as unique a case of juvenile rheumatoid arthritis in which a transient diffuse pulmonary lesion formed an intimate part of the disease, which was severe and progressive. We have not made an exhaustive study of the formidable literature, but the inquiries we have made have not produced any serious study directed to this aspect of the disease.

The case mentioned was that of a boy aged 6, weighing 34 lb. 12 oz. (15.76 kg.), in whom the pulmonary episode was heralded by a mild tonsillitis but was symptomless except for a slight increase in malaise. Intermittent pyrexia over eight days increased to 101.8° F. (38.8° C.) and subsided by lysis over a similar period. There was no leucocytosis or eosinophilia. The blood sedimentation rate, which had been 80 to 100 (Westergren) over many months, fell to 15 on one occasion at the height of the fever, rising again almost immediately afterwards. This boy was in hospital throughout in a separate cubicle, and his Mantoux reaction was negative before and after the incident. The radiographs show the condition of his lung in November, 1947, four months before the onset of the lung condition: on March 3, 1948, at the height of it; and on April 6, when it was resolving (Plate, Figs. 1, 2, and 3). Pulmonary congestion from left heart failure, possibly precipitated by a blood transfusion given three weeks before, was considered as a possible explanation of the attack, but this idea was rejected because of the absence of any evidence of heart disease and the rapid recovery without treatment. During this transfusion, the latest of several, an extensive urticarial rash appeared. In an earlier transfusion there had been rather more severe allergic symptoms. The further history of the case is as follows

Case History

A boy aged 6 was admitted to hospital on Dec. 12, 1946. He was the youngest of six children, and his parents and sibs were all healthy. In November, 1945, he had scarlet fever, followed by otitis media. He then remained well until August, 1946, when swelling of various joints occurred, each lasting for only a few days. There was pain on movement, but no tenderness. From time to time all the joints of his body, except those of his hands, feet, and spine, were affected. He was febrile during the attacks of pain, he lost weight, and his energy and appetite decreased.

At the time of admission there was swelling of the right ankle and right knee, but little interference of movement and no tenderness. He was afebrile. There was slight generalized adenopathy, but no apparent enlargement of the spleen then or subsequently. The liver, however, was always palpable. A diagnosis of juvenile rheumatoid arthritis was made, and he was given two courses of gold treatment with a maximal single dose of 25 mg. (total, "myocrisin" 212 and 210 mg.). During the next year he remained in hospital, and the disease was steadily progressive. Profound muscle-wasting occurred, and there was some limitation of movement at the wrists and severe osteoporosis with a tendency to periostitis in the small bones. Anaemia was a persistent feature and was only slightly improved by treatment,

LESLIE NANCEKIEVILL: ACUTE IDIOPATHIC PULMONARY HAEMOSIDEROSIS

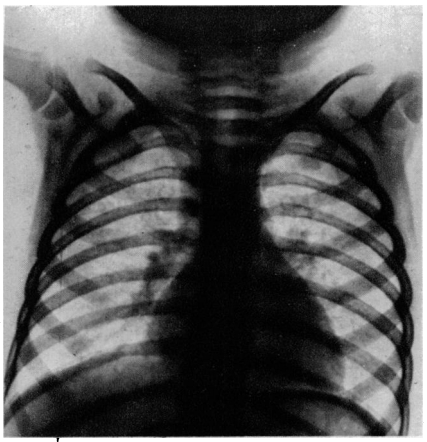

FIG. 1.—Slight mottling of both lungs in early phase.

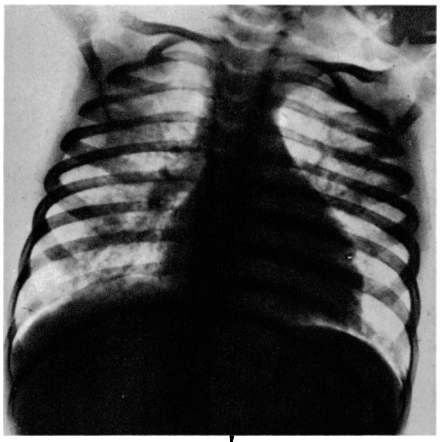

FIG. 2.—Diffuse gross reticulation in late phase.

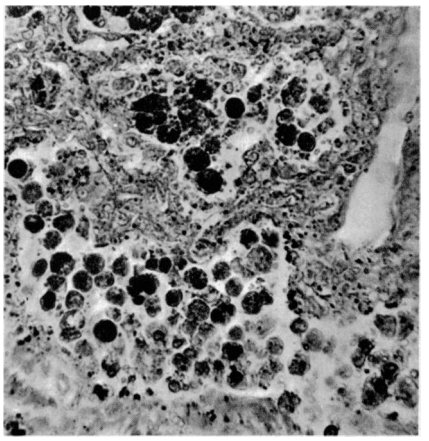

FIG. 3.—Iron-containing cells in alveoli. Prussian blue. (× 100.)

ETHEL BROWNING: BLOOD CHANGES IN LUMINIZERS

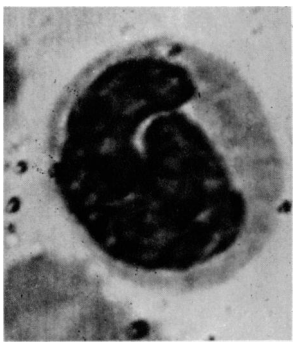

FIG. 1.—Normal monocyte of diameter 13·5μ. (× 2270.)

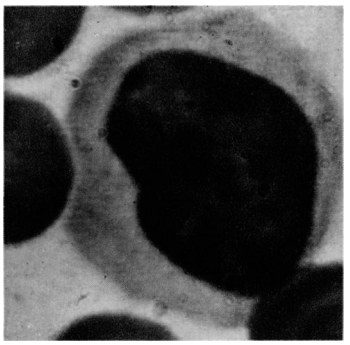

FIG. 2.—Normal monocyte of about 15μ diameter. (× 2270.)

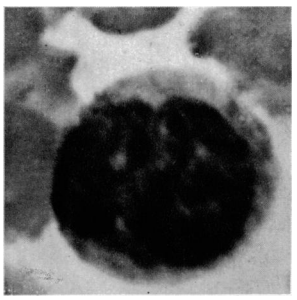

FIG. 3. — Lymphoidocytic monocyte of diameter 11·5μ. (× 2270.)

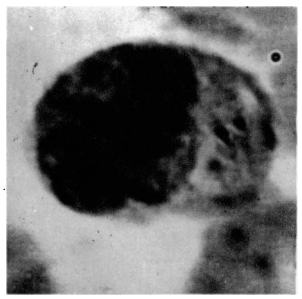

FIG. 4.—Abnormal (intermediate type) lymphoidocytic monocyte, 13·5 by 11μ.

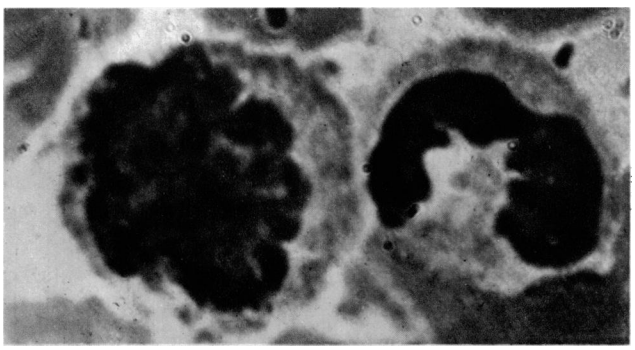

FIG. 5.—Abnormal large monocyte (18·5μ) resulting from initial stimulation of reticulo-endothelial system by ingested radioactive material. Some granularity in cytoplasm. (× 2270.)

H. M. LAWSON: DISSEMINATED OSSIFICATION OF THE LUNGS

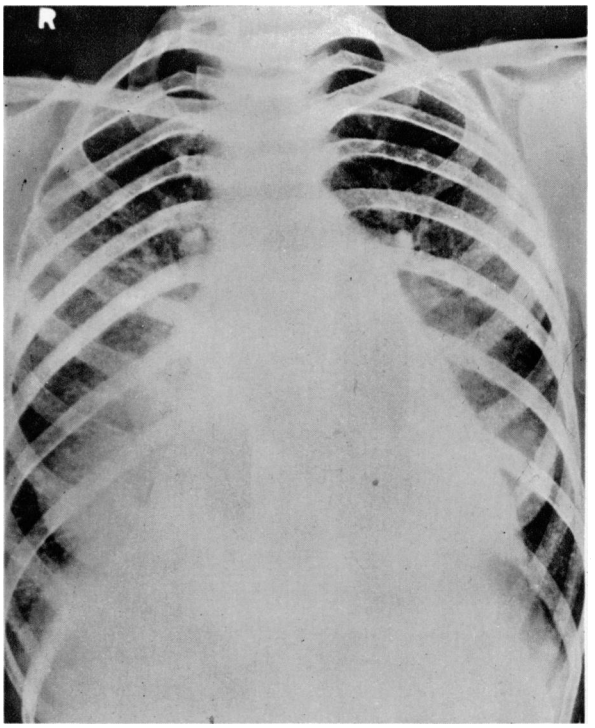

FIG. 1.—Miliary shadowing of both lungs. The heart contour is typical of mitral stenosis.

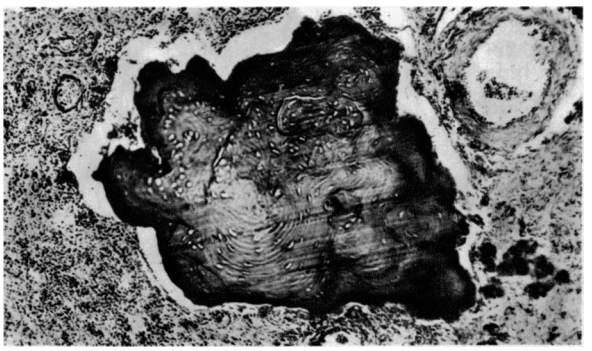

FIG. 2.—Section of lung showing irregular nodule of bone with laminations and included osteoblasts. (× 100.)