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IDIOPATHIC PULMONARY HAEMOSIDEROSIS

BY

G. B. TAIT AND M. CORRIDAN

From the General Hospital, Dewsbury

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Idiopathic pulmonary haemosiderosis (essential brown induration of the lungs) is now well recognized in children. Originally described by Ceelen in 1931, many other authors have added cases (Wyllie, Sheldon, Bodian, and Barlow, 1948).

It manifests itself clinically by attacks of tachycardia, pallor, anaemia, cyanosis, haemoptysis, and sometimes jaundice, together with clinical or radiological signs in the chest. The earlier attacks subside and are often mistaken for haemolytic episodes. Pathologically there is haemorrhagic pulmonary consolidation. The alveoli are filled with haemosiderin-laden phagocytes ("heart failure cells") and these are also present in the alveolar walls.

A similar condition has been described only three times in adults. Belfrage and Waldenstrom described a case which developed at the age of 16 years and ended fatally at 19 years (Wyllie and others, 1948). Walton and Williams (1951) described another case in the same age group. Only one patient whose symptoms started later in life has been described previously, a man aged 38 years, by Borsos-Nachtnebel (Scheidegger and Dreyfus, 1945). We describe here a case occurring in a man aged 31 years.

CLINICAL FEATURES

The patient was a school-teacher aged 31. He had enjoyed good health until two and a half years previously, when he began to suffer from occasional bouts of diarrhoea, sometimes severe but without mucus or blood. He had been investigated fully for this complaint at another hospital, but no evidence of organic disease had been found.

He attended the out-patient department of the General Hospital, Dewsbury, on January 17, 1951, with a history of one week's cough with sputum which had been blood-stained. The blood-staining had ceased at the time of attendance. Otherwise he felt well, but had lost some weight over the past three years. There was nothing relevant in the family history. Physical examination revealed a spare individual of average height. The respiratory and other systems were normal. There was no telangiectasis of the skin. His blood pressure was 110/90. Blood examination showed haemoglobin 109% (16.1 g. per 100 ml.), erythrocyte sedimentation rate (E.S.R.) 1 mm. fall in one hour. A radiograph of the chest showed no abnormality, and the sputum did not contain tubercle bacilli. Examination of stools revealed no evidence of parasites.

The patient was kept under observation as an out-patient. Repeated physical and radiological examinations were negative. Bronchoscopy on May 16, 1951, by Mr. G. H. Wooler showed nothing to account for the bleeding.

From time to time bouts of haemoptysis recurred. He was admitted to hospital on August 12, 1951, after a profuse haemoptysis.

No clinical abnormalities were detected except early clubbing of the fingers. The blood haemoglobin was 77% (11.4 g. per 100 ml.), the E.S.R. was 13 mm. in one hour; otherwise the blood was normal. The stools were re-examined, with negative results; they had a normal fat content. Repeated examination of the sputum showed no acid-fast bacilli. The Wassermann and Kahn tests were negative. A chest radiograph revealed no abnormality. From time to time he complained of vague discomfort behind the sternum and attacks of feeling he might choke. He improved on bed-rest, and the bleeding gradually ceased. He was discharged on August 28, 1951.

He was readmitted on November 1, 1951, in a collapsed state after a severe haemoptysis. He was transfused with blood, but in spite of this deteriorated and died.

NECROPSY

The body was that of a thin, red-haired, freckled man with pale skin and mucous membranes. The lungs were large and voluminous and did not collapse on opening the pleural cavities, which were normal. The pleura covering each lung was strikingly dark purple. The mid zones were particularly firm, and haemorrhagic fluid could be squeezed from the lung parenchyma. The trachea and bronchi contained a large amount of haemorrhagic fluid. A superior bronchopulmonary and an inferior tracheo-bronchial lymph node on the left side, and a superior and an inferior tracheo-bronchial lymph node on the right side, showed moderate enlargement. The cut

surface of these enlarged nodes was greyish-brown. The right side of the heart was slightly dilated but otherwise appeared normal. The liver and spleen were normal in size and appearance. The gastro-intestinal tract and other abdominal organs were normal macroscopically. The pia-arachnoid was congested and the anterior part of the corpus callosum showed several purpuric spots of about 2 to 4 mm. in diameter.

MICROSCOPICAL EXAMINATION.—Many sections from various parts of both lungs were examined, and showed the same picture. Very few alveoli contained air. There were areas of recent haemorrhage, in which the alveoli were stuffed with fresh red blood cells representing the fatal haemoptysis. The number of alveoli in these haemorrhagic areas was quite variable. The older haemoptyses and intra-alveolar haemorrhage were represented by areas in which the alveoli contained numerous haemosiderin-laden phagocytes (siderophages) which gave a brilliant Prussian blue reaction (Fig. 1), and also by areas of casts in the alveolar ducts and related alveoli. The central part of these casts, in the ducts, was composed of red blood cells, while the alveolar or peripheral part showed siderophages and fibrin threads which stained with Mallory's phosphotungstic haematoxylin (Fig. 2). These areas of recent and old haemorrhages alternated irregularly throughout the lungs. The alveolar walls contained siderophages and free granules of haemosiderin.

There was a slight increase of the reticular fibrils of the alveolar walls. The elastic tissue of the alveoli showed neither iron impregnation nor foreign-body giant-cell reaction, and, apart from fractures, was regarded as being within normal limits. The small and medium-sized vessels were normal. The broncho-pulmonary and tracheo-bronchial lymph nodes showed simple hyperplasia and contained a small number of siderophages in the peripheral sinuses. Sections of spleen, bone marrow, and small and large intestine were normal and showed no haemosiderin.

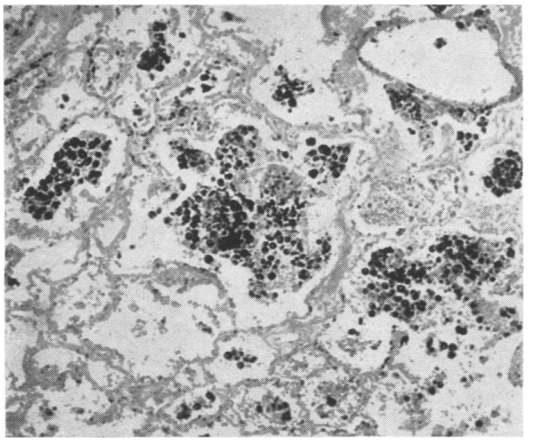

FIG. 1.—Lung alveoli containing haemosiderin-laden phagocytes. (Prussian blue, × 75.)

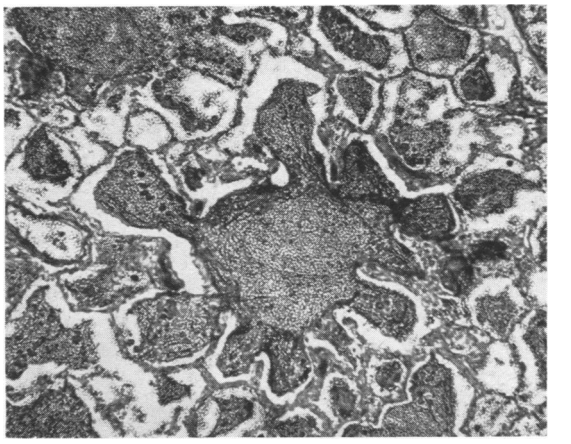

FIG. 2.—Cast in alveolar duct and related alveoli composed of red blood cells, fibrin threads, and haemosiderin-laden phagocytes. (Phosphotungstic acid haematoxylin, × 75.)

DISCUSSION

There are several points of interest about this case. In the first instance it is only the second case reported in an individual over 16 years of age. The reason for the higher incidence in children is unknown.

In children the condition is marked by anaemia, and although clinical signs in the chest are usually absent, radiological appearances are definite. These often superficially resemble miliary tuberculosis. Diffuse mottling is usually present, especially at the hilar areas.

In this case there was no anaemia until seven months after the onset and then it was only of mild degree. In the final stages anaemia was profound.

The episodic nature of the condition in children is similar to the repeated bouts of haemoptysis in this case. The complete absence of radiological signs in the chest is interesting. In children the radiological signs wax and wane with the severity of the attacks and with the resulting anaemia. In this case the degree of bleeding into the lung tissues must have been small until the final attack. This is mirrored by the absence of any marked anaemia until late in the disease. In the last episode, with clinical signs present in the chest, it is almost certain that radiological features would have been

present had radiography been possible. Ellman and Gee (1951), however, suggest that haemorrhage alone does not account for the degree of anaemia in these cases, for this has been marked in cases with minimal radiological signs.

At no time was there any evidence of right heart failure, which frequently supervenes in children. Diagnosis is sometimes helped by examination of the sputum, or material obtained from lung puncture, for "heart failure cells."

Lendrum, Scott, and Park (1950) found histological changes in pulmonary haemosiderosis secondary to mitral stenosis identical with those described in the idiopathic disease. The only difference is that these changes are more widespread throughout the lung in idiopathic pulmonary haemosiderosis. The elastic tissue of the alveoli and pulmonary vessels was regarded as being within normal limits in this case, as in the case described by Nancekievill (1949).

It seems likely, therefore, that the changes in the alveolar walls and pulmonary vessels in idiopathic pulmonary haemosiderosis are secondary to haemorrhage into the lung parenchyma and not the primary cause of the haemorrhage.

We are unable to state whether the source of haemorrhage was the pulmonary capillaries or the broncho-pulmonary anastomosis. The cause of idiopathic pulmonary haemosiderosis remains unexplained.

Summary

A case of idiopathic pulmonary haemosiderosis is described, the second reported in an adult over the age of 16.

The clinical picture was essentially that of recurrent haemoptysis of unknown origin.

At necropsy the lungs were enlarged, voluminous, and on section showed brown induration.

Microscopically, the alveoli contained red blood cells and haemosiderin-laden phagocytes. The elastic tissue of the alveoli and pulmonary vessels showed no significant change.

The source of haemorrhage could not be determined.

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