IDIOPATHIC PULMONARY HAEMOSIDEROSIS IN AN ADULT

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Idiopathic pulmonary haemosiderosis was first described in an adult patient by Waldenstrom in 1940, since when a further 16 cases in patients aged 15 or over have been described; these were summarized by Wynn - Williams and Young (1956). Whilst many cases progressed to an early fatal termination, there may be, as Kerley (1951) suggests, a mild form of the disease which persists for many years. This paper presents the case of a patient who remains in comparatively good health since symptoms started in 1944; she has been under regular radiological observation since that time. There is evidence that she has a generalized disorder of capillary endothelium.

CASE REPORT

A trained nurse, born in 1925, enjoyed good health until 1943, when she had a slight haemoptysis following a heavy cold. In 1944 she developed collapse and consolidation of the right middle and lower lobes after appendicectomy. Since then she has been subject to a cough productive of red or pink sputum in the morning. This became more frequent over the years, until by 1954 it had become a daily occurrence. The staining of the sputum was more severe at the time of the menses, and was associated with a feeling of tightness in the chest, which was relieved when she coughed up blood. She had also had three major haemoptyses over the last 10 years, each of about 1 pint.

During the same period she has been subject to attacks of fever, wheeziness, and purulent sputum, which have necessitated her staying in bed for a few days at least twice each winter. These attacks were all associated with more severe haemoptysis.

RADIOGRAPHIC APPEARANCES.—The chest was first radiographed in November, 1944, after the appendicectomy (Fig. 1), when there was an extensive opacity in the right lower zone consistent with pneumonia and partial collapse of the middle and lower lobes, the remainder of the lungs appearing clear. In April, 1945, after a haemoptysis of 1 pint (600 ml.) or more, a radiograph showed scattered, ill-defined, blotchy shadows throughout both lung fields (Fig. 2). Four weeks later these shadows had cleared completely. After another small haemoptysis in February, 1948, a chest

radiograph (Fig. 3) showed for the first time scattered minute dense shadows over the lower half of the right lung, and the lower two-thirds of the left lung, in keeping with haemosiderosis. These shadows have gradually progressed since another large haemoptysis in 1950, and by 1955 were very obvious, particularly in overexposed films. A radiograph in March, 1957 (Fig. 4), showed little further change. A bronchogram in May, 1953 (Fig. 5), showed moderate cylindrical dilatation of bronchi in the right middle lobe and the basal segments of the right lower lobe. There was no evidence of bronchial obstruction.

She was admitted to hospital in 1955 after a haemoptysis of about $\frac{1}{2}$ pint (300 ml.) associated with a right-sided pleurisy, fever, and a recent increase of purulent sputum to 3 to 4 oz. (90 to 120 ml.) daily.

Her general condition was fair, with fever up to 102° F. (38.9° C.) and considerable wheeziness at rest. There was extensive purpura over both shoulders, with several large spontaneous bruises over both legs. There was no obvious jaundice. In the chest, the breath sounds were reduced and the percussion note was impaired over the right lower and middle lobes, with abundant moist sounds, and a pleural friction rub over the anterior part of the axilla. There were scanty moist sounds over the left lower lobe with inspiratory and expiratory rhonchi. Finger clubbing was absent. The heart sounds and the blood vessels were normal; blood pressure 125/80 mm. Hg.

There was no abnormality in the nervous system; the liver and spleen were not palpable, and there were no enlarged lymph nodes.

INVESTIGATIONS.—A chest radiograph showed widespread dense stippling in the lower two-thirds of each lung field, somewhat obscured on the right side by superimposed soft, blotchy shadows through the middle and lower zones.

Sputum contained many macrophages heavily laden with haemosiderin, and many red cells, but no eosinophils. On culture, it gave a mixed growth of pneumococci and coliform bacilli.

An electrocardiogram was normal. The urine on routine testing showed no abnormality.

Haemoglobin concentration was 80%. Erythrocytes, numbered 4,320,000 per c.mm., of which reticulocytes were 1%; platelets numbered 250,000, and leucocytes 9,300 per c.mm., with a normal differential

FIG. 1

FIG. 1.—Radiograph of chest, November, 1944.

FIG. 2

FIG. 2.—Radiograph of chest, April, 1945.

FIG. 3.—Radiograph of chest, February, 1948.

FIG. 4.—Radiograph of chest, March, 1957. Lower part of left lung, to show detail of mottling.

FIG. 3

FIG. 4

FIG. 5.—Bronchogram, May, 1953.

count. The red cell fragility was normal (lysis starting at 0.48% and completed at 0.35%), and a Coombs test was negative.

The bleeding time (90 seconds) and clotting time (7 minutes, Lee and White's method) were normal; serum bilirubin was slightly raised at 1.1 mg. per 100 ml. Hess's test for capillary fragility (the sphygmomanometer cuff being held at 100 mm. Hg for five minutes) was very strongly positive, with small and large petechiae at 1 to 2 mm. intervals over the whole forearm, too numerous to count.

A vitamin C saturation test (Gray, 1953) showed a slightly impaired rate of saturation.

EVIDENCE OF ASSOCIATED GENERALIZED CAPILLARY DISORDER.—From 1954 the patient noticed a tendency to spontaneous bruising, particularly over the shins, with crops of purpura over the shoulders, particularly when the haemoptysis was most marked. These observations were confirmed on her admission to hospital in July, 1955, when a Hess test was very strongly positive, indicating an increase in capillary fragility.

In April, 1955, she had an attack of pain in the left loin suggesting renal colic and, at the same time, developed very brisk haematuria which lasted for 18 hours and subsided spontaneously. Subsequent intravenous pyelograms were normal, and there has been no recurrence.

She had spontaneous abortions in 1949, 1951, 1952, and 1953, the last being followed by a sharp haemorrhage requiring transfusion of 2 pints of blood. A healthy child was born in 1950. She has never suffered undue menstrual loss.

TREATMENT AND PROGRESS.—After the patient's admission to hospital in July, 1955, the chest infection

responded normally to treatment with full doses of penicillin. She takes a full mixed diet, and there was no reason to suppose she had scurvy. Nevertheless in the absence of other recognizable haematological changes, she was treated empirically with ascorbic acid, 1,000 mg. daily by mouth and 1,000 mg. intramuscularly twice weekly. After two weeks of this treatment the Hess test was within normal limits.

The ascorbic acid was continued (200 mg. daily by mouth) until February, 1956; during this period of six months there was no spontaneous bruising and no purpura; and the haemoptysis was remarked by the patient as being less severe and less frequent than at any time in the preceding 10 years.

In February, 1956, she noted an increase in purulent sputum, and remained confined to her house because of wheeziness; she was, however, free of haemoptysis. Culture of the sputum produced a mixed flora, with pneumococci and *H. para-influenzae* predominating. Treatment with oxytetracycline, 2 g. daily, reduced the sputum in the course of a week to a trace of mucus in the morning only. Treatment was maintained with oxytetracycline, 1.5 g., and ascorbic acid, 1,000 mg., daily by mouth. The patient enjoyed apparently normal health with no haemoptysis until July, 1956, when ascorbic acid was discontinued, the antibiotic being continued as before. During the next three months she noticed blood-staining of the sputum on three or four mornings each week, but was otherwise well.

In September, 1956, the oxytetracycline was also discontinued; two weeks later she had an acute febrile illness associated with 2 or 3 oz. (30 to 45 ml.) of offensive purulent, heavily blood-stained sputum daily; the Hess test again showed an increase in capillary fragility. She was given ascorbic acid, 1,000 mg., and oxytetracycline, 1.5 g., daily, and both the infection and the bleeding were fully controlled within a week. She has been maintained on this regime to the time of writing (May, 1958) and enjoys complete freedom from symptoms; the Hess test repeatedly shows normal capillary fragility. In October, 1957, she had a sharp attack of clinically typical influenza, associated with bronchospasm, but no haemoptysis and no purulent sputum.

On April 29, 1958, after an uneventful pregnancy, she gave birth to a healthy child after surgical induction of labour at 38 weeks. Post-partum bleeding was normal in amount.

DISCUSSION

Pulmonary haemosiderosis may be the end-result of a number of conditions which have in common capillary haemorrhages in the lungs with the deposition of haemosiderin, usually causing characteristic radiographic changes. Mitral stenosis has an established aetiological relationship with these lung changes. Apart from this the causation remains unknown. The 16 examples of this group of "idiopathic"

pulmonary haemosiderosis reported in patients aged over 14 have been summarized by Wynn-Williams and Young (1956), who add a case of their own. It might be supposed that the recurrent haemoptysis of bronchiectasis could be followed by similar changes; and indeed two cases have been reported by Cameron (1956) which show the association of diffuse haemosiderosis, demonstrated histologically, with generalized bronchiectasis. In one of these pneumoconiosis was also present. Both patients had necropsy evidence of pulmonary hypertension; the characteristic fine dense stippling was absent from the chest radiographs, so that these two patients possibly represent a different syndrome from the "idiopathic" group.

In the present case, localized cylindrical bronchiectasis in the right middle and lower lobes is present, which evidently dates from post-operative collapse-consolidation in 1944; but it is difficult to accept that the very widespread changes throughout both lungs could be the result of bleeding from this localized lesion. Furthermore, there is evidence of generalized increased capillary fragility presenting as spontaneous bruising, spontaneous purpura, haematuria, and possibly repeated spontaneous abortion.

In our opinion the recurrent haemoptyses which have produced the radiographic picture of haemosiderosis are the result of a generalized

capillary disorder affecting the pulmonary as well as the systemic capillaries, and which appears to have been controlled by treatment with ascorbic acid. The bronchiectasis is probably an aggravating factor; continuous antibiotic treatment has eliminated the recurrent febrile respiratory illnesses to the time of writing.

SUMMARY

The case of a woman aged 33 with idiopathic pulmonary haemosiderosis is presented. The first haemoptysis occurred at the age of 18, and typical radiographic changes first developed at the age of 23. There is an associated localized cylindrical bronchiectasis.

Ten years after the first symptom she developed spontaneous bruising and purpura suggesting a generalized capillary disorder.

The bleeding has ceased following the administration of ascorbic acid, and the associated recurring chest infections have been controlled by prolonged administration of oxytetracycline.

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