

Idiopathic Pulmonary Hemosiderosis*

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IDIOPATHIC pulmonary hemosiderosis was originally described by Virchow in 1851. The first clinical report, by Ceelen, did not appear until eighty years later, in 1931, and was followed by roentgenologic, clinical and pathologic correlations in 1944 by Waldenström.¹ An extensive review of the entire subject was published in 1948 by Wyllie, Sheldon, Bodian and Barlow.² Approximately thirty-three cases have been reported in the English language,¹⁻¹⁶ thirty of which were children and three adults. The present report concerns three adults.

Idiopathic pulmonary hemosiderosis usually starts in childhood, often in children who are sickly from birth. The disease is characterized by recurrent acute episodes of dyspnea, cyanosis, cough with hemoptysis, fever, tachycardia and anemia. Jaundice and abdominal pain occasionally are present. Anemia is often the presenting symptom and may be severe in cases with only minimal hemoptysis. The jaundice may be severe and occurs in those who have been ill for some time. Acute episodes generally last two to three days but occasionally persist for several weeks. During remission there may be apparent complete recovery, but frequently some exertional dyspnea and anemia remain. When the disease has been of long duration, cor pulmonale with congestive circulatory changes may develop. Clubbing of the fingers has been reported.

Examination of the chest during exacerbations may be unremarkable or reveal only scattered areas of dullness and rales, but some patients show signs of lobar consolidation. The appearance of the chest x-ray varies with the stage of the disease. Initially, diffuse homogeneous opacities may be seen, although at times the most dense areas are localized to the mid or central lung fields. A coarse, generalized mottled appearance is often present. As the acute phase subsides a flecked reticular pattern becomes apparent and complete clearing may ultimately occur.

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In exacerbations the sputum contains hemosiderin-filled macrophages. Anemia of the iron deficiency type is present. Reticulocytosis and low serum iron are commonly found, but the response to iron therapy is variable. Serum bilirubin may be elevated, and in a few cases elevated cold agglutinin titers have been demonstrated. Measurements of the clotting mechanisms, iron metabolism and capillary fragility are within normal limits.

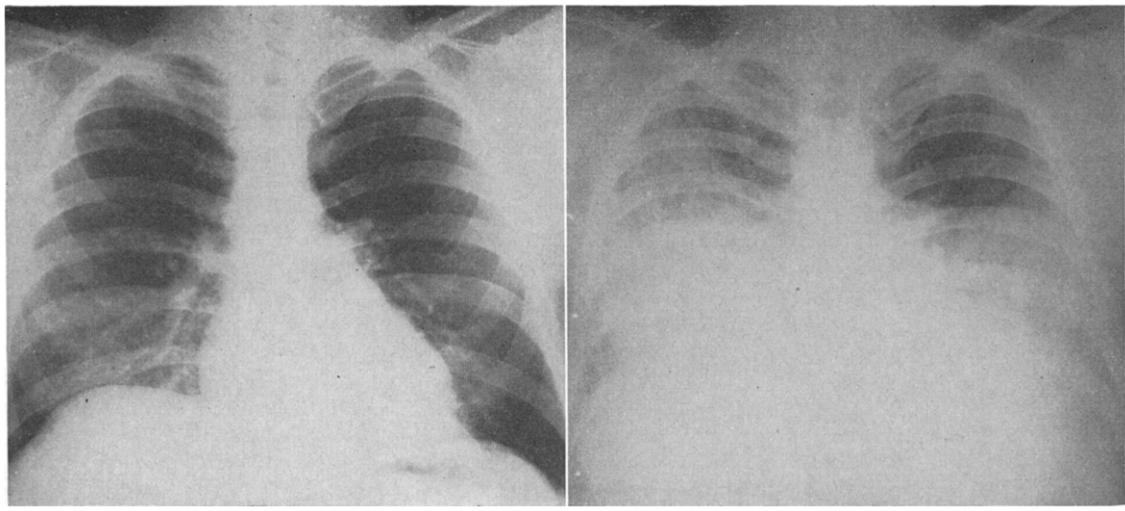
At autopsy the lungs are large, heavy, red-brown and firm. Microscopic examination reveals an increase in reticulum, collagen and muscle, together with decrease or fragmentation of elastic tissue in alveolar walls and small blood vessels. Fresh and old hemorrhages and many hemosiderin-filled macrophages are seen. No evidence of primary heart disease is present. Right-sided dilatation is often present in acute cases and cor pulmonale occurs if the course is protracted. One lung analysed for iron was shown to contain 1.97 mg. per 100 gm. compared to 0.03 mg. per 100 gm. in the lung of a normal control subject.

The most striking difference between the hemosiderosis seen in mitral stenosis and that in this disease is the marked vascular abnormality²⁵ which occurs in the former condition but is not seen associated with idiopathic pulmonary hemosiderosis.

Theories of the etiology of this disease have implicated a primary circulatory defect, a defect in elastic tissue of the blood vessels and lung, primary iron storage in the lung, tissue fragility secondary to iron deficiency and allergy. The presence of a hemolytic element has been suggested. However, the pathogenesis of the entire process has never been established.

CASE REPORTS

CASE I. (Boston Veterans Administration Hospital, No. 8832.) The patient, a twenty-five year old electrician's helper, had been in good health until one year prior to his first admission, at which time slight



1A

1B

FIG. 1. Case 1. A, initial chest film which was interpreted as normal. Increased density in right lower lung field was thought to be due to a heavy pectoral muscle. In retrospect it is believed that it may have been caused by right lower lobe disease. B, film taken at the time of second admission, nineteen days after the first. The disease appears to involve all of the lungs except for part of the left upper lobe.

transient hoarseness and a persistent cough productive of small amounts of light yellow sputum developed. He denied all other symptoms. Three months prior to admission small flecks of blood began to appear in his sputum. A few weeks before admission the cough increased and "mouthfuls" of bright red blood began to appear. For three weeks he noted marked exertional dyspnea and a sensation of tightness at the sternal notch. One week prior to admission he had vomited twice, feeling "that something was stuck in the mid-chest." Hemoptysis had subsided just before admission. He was said to have had four chest x-rays and two examinations of the sputum alone elsewhere during the year, all of which gave "negative" results.

Physical examination revealed his temperature to be 98°F., pulse 72, and blood pressure 140/80. The patient was a well developed, slightly obese, ruddy complexioned young white man in no distress. General physical examination revealed no abnormalities.

Laboratory data revealed the following: hemoglobin, on admission, 11.5 gm.; hematocrit 43 per cent; white blood count 6,500; differential count: neutrophils 54, lymphocytes 32, monocytes 1, eosinophils 3; urinalysis gave negative results; the stool guaiac test gave a negative reaction; the Kahn test gave a negative reaction; the sputum culture revealed alpha hemolytic streptococcus and *Neisseria catarrhalis*; sputum smears were negative for acid-fast bacilli, and the Papanicolaou smear of sputum gave negative results for neoplastic cells. Films and fluoroscopy of the chest revealed no abnormalities (Fig. 1) except for slight prominence of the pulmonary artery.

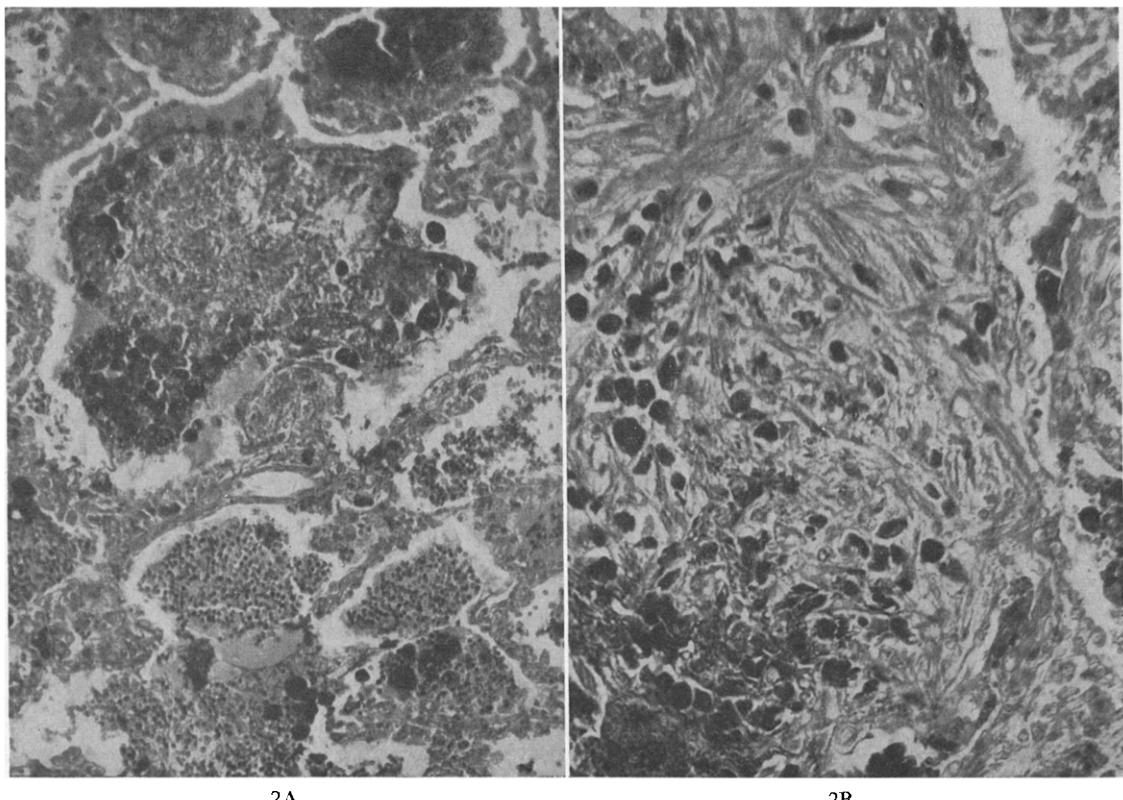
The patient was hospitalized for a total of ten days. During this period he was completely afebrile. A small

amount of sputum was raised which was blood-flecked for the first few days and then the hemoptysis stopped. Laryngoscopy and bronchoscopy were within normal limits. A hemoglobin determination on the sixth hospital day was 13 gm. The patient was discharged to return in six weeks for another x-ray of the chest.

Nineteen days later the patient was readmitted to the hospital. He had been asymptomatic until three days before returning, at which time a severe shaking chill, fever, cough and dyspnea developed. He began coughing up bright red blood, the total volume being estimated at 500 to 700 cc. These symptoms persisted until admission. He received no medication at home.

At physical examination the temperature was found to be 101.6°F., pulse 136, respirations 44 and blood pressure 130/60. The patient appeared acutely ill and was moderately dyspneic. The mucous membranes were cyanotic. There was limited expansion of the chest bilaterally. Breath sounds were generally diminished, and in the region of the right lower lobe dullness, rales and bronchial breathing were noted. Small amounts of bright red blood was noted in the sputum. There were no other remarkable physical findings.

Laboratory data revealed the following: white blood count 19,500; differential count: neutrophils, 85, lymphocytes 14, monocytes 1; hemoglobin 9 gm.; hematocrit 29 per cent; urinalysis gave negative results; bleeding time was one minute; clotting time (Lee-White) eight minutes; prothrombin time 100 per cent; icterus index 10 units; blood smear revealed abundant platelets and red blood cells appeared normal; sputum smear revealed a few gram-negative



2A

2B

FIG. 2. Case 1. A, iron stain of lung showing fresh blood and siderophages (dark) in alveoli and bronchiole; original magnification $\times 100$. B, iron stain of lung showing intra-alveolar plug of fibrous (organized) material containing many siderophages; original magnification $\times 200$.

rods and gram-positive cocci; red blood cells were seen but there were few polymorphonuclears and no macrophages; sputum culture gave negative results for pathogens. Chest film (Fig. 1) showed extensive consolidation of the lower two-thirds of both lung fields, with extension of the process into the apical region on the right. The heart and great vessels were largely obscured but did not appear to be unusual.

Bedrest and oxygen therapy were instituted. Intramuscular aqueous penicillin, 300,000 units every three hours, was given. The patient received morphine and later luminal® to control his apprehension. He continued to have grossly bloody sputum while in the hospital but seemed to improve somewhat overnight. His temperature ranged between 102° to 103°F. The morning following admission the hemoglobin had dropped to 7.5 gm. with a hematocrit of 25 per cent. Through that day he received 1,000 cc. of whole blood intravenously. In the afternoon he became disoriented, then completely unresponsive. He died a few hours later after a sudden marked increase in tachypnea followed by cessation of respirations.

Permission was obtained for posthumous examination of the thorax only.

On gross examination the pleural cavities were essentially normal. The right lung weighed 1,500 gm. and the left 1,225 gm. Both lungs were quite airless and solid without discrete nodulation. Cut surfaces were meaty and dark red with a diffuse brownish tinge. Pressure caused exudation of a considerable amount of blood, but no pus was present. Red blood was present in the bronchi but their mucosa was normal. Pulmonary arteries were not remarkable. A few discrete gray-brown to black hilar lymph nodes were present, the largest being 2.5 cm. in diameter. The heart weighed 300 gm. The right ventricle was 2 to 3 mm. in thickness and the left 1.2 to 1.4 cm. No dilatation of any chamber was noted and all cardiac structures were essentially normal in appearance. The aorta and esophagus were not remarkable.

One section of each lobe of the lungs was microscopically examined. They were all at least seven-eighths airless. (Figs. 2 and 3.) The alveoli were full of red blood cells and masses of macrophages containing hemosiderin. The reaction for iron was strongly positive. The macrophages were often imbedded in plugs of organizing clot. The total area occupied by fresh blood was about twice that of older organizing material. A few neutrophils were present in or along

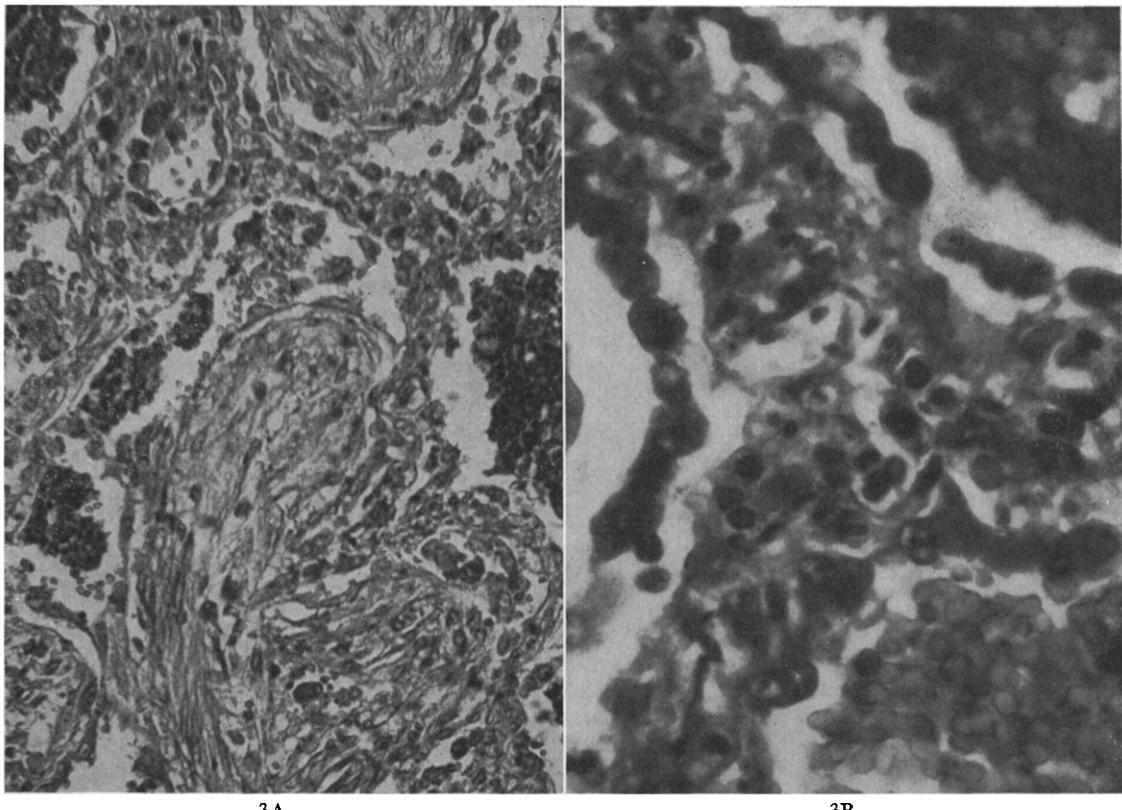


FIG. 3. Case 1. A, intra-alveolar fibrous plugs; only a few of these were present; original magnification $\times 200$. B, Verhoeff's elastic tissue stain of lung; these were the thickest alveolar walls found, showing infiltration by macrophages and lymphocytes, paucity of elastic fibers and swollen epithelial cells lining the blood-filled alveoli; original magnification $\times 450$.

the alveolar walls where hemorrhage was fresh. There was no purulent exudate. The alveolar lining cells were found occasionally to be swollen or cuboidal and there was some thickening of alveolar walls. In a few areas alveoli were completely filled with fibrous plugs. Little hemosiderin occurred in the interstitial tissue. In each section three or four foci were found in which the alveolar walls were thickened as much as two to four times their normal thickness and contained fine collagen fibers and sparsely scattered macrophages and lymphocytes. Verhoeff's stain revealed a marked reduction of the elastic tissue in the thickened walls in some of the foci, but elsewhere the elastic tissue was essentially normal. One or two venules showed reduction or absence of elastic tissue in part of the wall but none appeared ruptured. There was no capillary engorgement as seen in mitral stenosis. Arteriolar necrosis and foreign body giant cells were absent.

The solid fibrous plugs represented old organized lesions, the dense fibrinous exudate full of siderophages was a more recent organizing process, and the fresh blood within fibrin was from very recent hemorrhage.

The hilar lymph nodes showed dilatation of the sinuses, which contained many neutrophils and littoral cells, and small numbers of red blood cells. Iron stain showed intracellular hemosiderin in the sinuses throughout the nodes. The remainder of the intrathoracic organs were not remarkable.

Cultures taken at autopsy yielded alpha hemolytic streptococcus from the heart's blood and coagulase-negative *Staphylococcus aureus* from one lung. Smears of the lungs were devoid of organisms.

Diagnoses were pulmonary hemosiderosis, idiopathic; hemosiderosis of hilar lymph nodes.

CASE II. (West Roxbury Veterans Administration Hospital, No. 25067.) This twenty-two year old white male college student was in good health until the age of eighteen when he coughed up a teaspoonful of bright red blood. X-ray of the chest was taken and he was hospitalized for two weeks with a diagnosis of virus pneumonia. The patient was given "sulfa" and discharged asymptomatic, the subsequent x-ray having appeared clear.

At the age of twenty-two a cough developed and the patient experienced a two-day episode of hemoptysis, bringing up six teaspoonsfuls of bright red blood daily.

One month later hemoptysis recurred and persisted for ten days. Exertional dyspnea developed and the patient felt feverish. At this time he was admitted to another hospital. Positive findings included a temperature of 101.2°F., rales and bronchial breathing in the right mid-chest, and granular infiltration of the lower lung fields near the hilus. The white blood count was normal and the sputum was negative for pathogenic bacteria. The patient received intramuscular penicillin and several blood transfusions. The acute symptoms subsided in about one week. Bronchoscopy at that hospital gave negative results save for the presence of clotted blood in the main bronchi. The patient was discharged on a regimen of oral iron and referred to the West Roxbury Veterans Administration Hospital for further work-up.

On admission the patient was noted to be well developed and nourished, and without noteworthy abnormalities on physical examination.

Laboratory data revealed a hemoglobin of 10.2 gm.; red blood count 3.2 million; white blood count 5,700; differential count: neutrophils 64, lymphocytes 32, monocytes 4; urinalysis gave negative results; stool guaiac test was 1+; sedimentation rate (Wintrobe) was 11 mm. per hour; reticulocyte count 1.4 per cent; platelet count 135,000; bleeding time was four and a half minutes; venous coagulation time five minutes; the Kahn test was negative. Postero-anterior x-rays of the chest showed increased bronchovascular markings at both bases. Bronchogram was normal.

The patient was asymptomatic while in the hospital. He continued to receive oral iron. A repeat hemoglobin on the tenth hospital day was normal and he was discharged on no medication. Two weeks following discharge he noted the sudden onset of shaking chills, weakness, malaise, nausea and vomiting. Twelve hours later he began coughing up large amounts of bright red blood. Shortness of breath developed and he returned to the hospital.

At physical examination his temperature was found to be 103°F., pulse 100, respirations 24 and blood pressure 110/60. The patient appeared acutely ill and continually coughed up dark red blood. Fine rales were heard at the left lung base. Examination was otherwise not remarkable.

Laboratory data revealed a hemoglobin of 11.5 gm.; red blood count 4.1 million; hematocrit 40 per cent; white blood count 13,900; differential count: neutrophils 85; lymphocytes 14; eosinophils 1; sedimentation rate was thirty-four mm. in one hour; prothrombin time 70 per cent; bleeding time was two and a half minutes; venous coagulation time twelve minutes; reticulocyte count was 1.1 per cent; platelets abundant on smear; urinalysis gave negative results save for 1 to 3+ albuminuria; sputum culture revealed *Diplococcus pneumoniae* and *Staphylococcus albus*. Postero-anterior view of the chest showed diffuse infiltration of both lung fields and scattered residual

lipiodol. Interpretation was difficult because of the presence of lipiodol.

The patient rapidly became worse. The temperature remained between 103° and 104°F., and shortness of breath became extreme. He was treated with large doses of penicillin and chloromycetin,[®] then aureomycin, without improvement. The hemoptysis continued and the hemoglobin dropped to 7.5 gm. The patient was transfused with 500 cc. of whole blood. Signs of acute right-sided congestive failure appeared. He was digitalized without improvement and died of respiratory failure on the third hospital day.

Autopsy was performed. On gross examination, the pleural cavities were not remarkable. The right lung weighed 1,500 gm., the left 1,440 gm. Both lungs appeared blackish red and were firm and non-crepitant. The bronchi and trachea were filled with dark blood and postmortem clot but were otherwise not remarkable. The blood vessels were of normal appearance. Cut surfaces of the lung revealed the parenchyma to be markedly engorged with blood and have a fine nodular feeling. The heart weighed 465 gm. The right ventricle was 0.4 cm. in thickness and the left 1.2 cm. The valve measurements were as follows: tricuspid valve 12 cm., pulmonic 6.5 cm., mitral 9 cm. and aortic 6 cm. The valves, musculature and arteries of the heart were entirely normal. There were no other positive gross findings.

Sections taken from all lobes, except the right middle lobe, were examined microscopically. In each the parenchyma was 75 to 90 per cent airless and 50 to 80 per cent filled with fresh blood. There were a few foci 2 to 4 mm. in diameter where alveoli were filled with an exudate consisting of old fibrin, macrophages, siderophages and fibroblasts, with occasional clumps of neutrophils. In these foci the alveolar walls and interstitial tissue were edematous with some infiltration by macrophages, lymphocytes and eosinophils. In two sections small interstitial foci of lymphocytes were noted. Occasional chronic inflammatory cells were seen in proximity to normal venules. Large, round, empty spaces were present which perhaps represented air or the former sites of iodized oil. No completely organized fibrous plugs were seen. Large numbers of siderophages were scattered throughout both the freshly hemorrhagic areas and the relatively bloodless foci, at times filling the entire alveolar space.

The section from the right middle lobe was different from the rest. Here the tissue was well aerated and many alveoli were lined with a thick eosinophilic hyaline membrane containing small hemosiderin granules and a few neutrophils. Siderophages were scattered loosely throughout but filled a few alveoli. Some alveoli contained pale edema fluid. The interstitial tissue appeared edematous with moderate infiltration by macrophages, lymphocytes, neutrophils and hemosiderin-containing macrophages. The bronchioles were empty.

Examination of sections stained with Verhoeff's stain showed elastic tissue to be preserved in hemorrhagic areas but completely lacking in the foci of organizing exudate. There was no giant cell reaction. The sinuses of the carinal lymph nodes were full of reticulo-endothelial cells and hemosiderin. These sinuses also contained lymphocytes and a few neutrophils and eosinophils. Slight vacuolization of cell cytoplasm in the lobule centers of the liver was seen and also a few polymorphonuclear cells in the adjoining sinusoids. The adrenal gland showed some reduction in cortical lipid vacuoles. Culture of heart's blood was positive for *Pseudomonas aeruginosa*. Culture of the left lower lobe was positive for the same organism but there was no growth from the right lower lobe.

Diagnoses were pulmonary hemorrhage and hemosiderosis; slight chronic and subacute focal (organizing) pneumonitis; and hemosiderosis of carinal lymph nodes.

Comments. Both Cases I and II clinically fit the entity of idiopathic pulmonary hemosiderosis. The source, or sources, of hemorrhage was not discovered but there is a hint of chronic pneumonitis in Case I and it is definitely present in Case II. The latter patient received a sulfonamide in 1945 and had eosinophils in the pulmonary lesions at autopsy but no clear evidence of hypersensitivity angiitis was distinguishable; the focal lesions presumably date from his May or June episodes. That posthemorrhagic secondary infection could produce such lesions seems unlikely but must be considered a possibility.

CASE III. (West Roxbury Veterans Administration Hospital, No. 24251; Boston Veterans Administration Hospital, No. 1687.) A twenty-nine year old male office clerk was well until 1946 when he noted the onset of malaise, mild chills and a cough productive of small amounts of light green, mucoid sputum which had some blood streaking. The episode lasted two weeks and cleared spontaneously.

The patient remained asymptomatic until 1947 when symptoms recurred but now with persistent frank hemoptysis accompanied by pallor. Weakness, exertional dyspnea and palpitations were noted. He was seen by a physician, who is said to have found a hemoglobin of 50 per cent and bilateral pulmonary infiltrations on x-ray examination. The patient was hospitalized and kept at bedrest for five weeks, during which time he made a slow recovery. Two months after discharge he felt quite well and returned to his work.

In 1949 his symptoms again recurred, and he was seen at the West Roxbury Veterans Administration Hospital. Physical examination revealed a well developed, slim young male with moderate pallor. A few rales were noted at the left base.

Laboratory data revealed a hemoglobin of 7.5 gm.; peripheral blood smear showed hypochromia and polychromatophilia of red cells, 2 per cent nucleated

red cells and abundant platelets; white blood count was 11,700; differential count: neutrophils 55 per cent, lymphocytes 38 per cent, monocytes 4 per cent, eosinophils 2 per cent, basophils 1 per cent; reticulocyte count was 5 per cent; bleeding time was two minutes; venous coagulation time (Lee-White) three and a half minutes; prothrombin time was 100 per cent; serum bilirubin was 0.2 mg. per cent; urinalysis gave negative results for bile and urobilinogen, otherwise normal; the Kahn test was negative. Postero-anterior x-ray of chest showed diffuse miliary shadows varying from 1 to 3 mm. in diameter (Fig. 4) throughout both lung fields. Gastrointestinal series and barium enema test gave negative results. Bacteriology of the sputum, including studies for acid-fast bacilli, was not remarkable.

A diagnosis of "idiopathic pulmonary hemosiderosis" was made. The patient was given oral iron with symptomatic improvement and was discharged after three weeks. Over the next six months his hemoglobin gradually rose to 12.5 gm. and remained at that level for two and a half years. X-ray of the chest (Fig. 4) showed marked clearing within four months and within eighteen months appeared normal. The patient returned to work and felt quite well, although his friends stated that he lacked his former strength and endurance.

In 1952 a "cold" developed accompanied with a persistent dry cough. After two and a half months the cough became productive of white sputum, which soon became yellow and bloodstreaked. Ten days later chills and sweats developed, with hemoptysis of one to two teaspoonsfuls of frank blood daily. Marked malaise ensued and after four days the patient was admitted to the Boston Veterans Administration Hospital.

The vital signs were normal. The patient was noted to be pale and thin but in no acute distress. General physical examination was within normal limits. X-ray of the chest (Fig. 5) revealed a finely granular infiltration involving most of both lung fields.

Laboratory data revealed a hemoglobin of 8 gm.; peripheral blood smear showed polychromatophilia of red cells and abundant platelets; white blood count was 14,250; differential count: neutrophils 76 per cent, lymphocytes 17 per cent, eosinophils 3 per cent, monocytes 4 per cent; sedimentation rate was 25 mm./hour; bleeding time was one minute; venous coagulation time six minutes; prothrombin time 74 per cent; serum non-protein nitrogen was 41 mg. per cent, total protein 5.7 gm. per cent, albumin 3.3 gm. per cent, globulin 2.4 gm. per cent; serum bilirubin was 1.6 mg. per cent; serum iron 64 gamma per cent (normal 60–130 gamma per cent); cold agglutination test, Coombs test, and stool guaiac test gave negative results; fecal urobilinogen 625 mg. per 100 gm.; (normal 30–300 mg. per 100 gm. of stool); urinalysis was entirely within normal limits; sputum smear revealed hemosiderin-filled macrophages; sputum

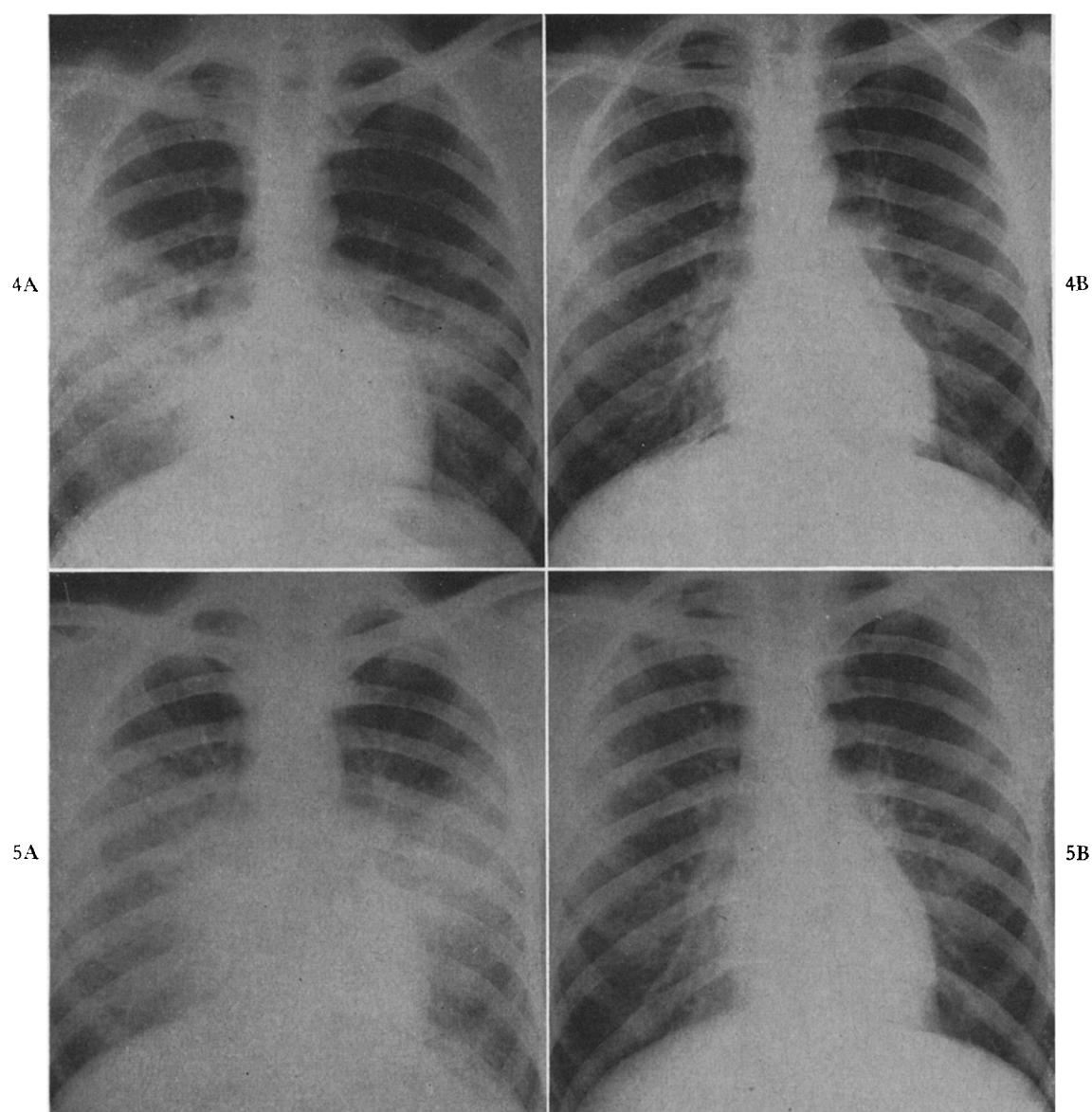


FIG. 4. Case III. A, initial chest film taken on June 1, 1949, showing bilateral pulmonary infiltration. B, chest film taken on October 7, 1949, four months later, showing striking clearing of lung fields.

FIG. 5. Same case. A, chest film taken on July 29, 1952, showing extensive diffuse pulmonary infiltration. B, chest film taken on September 16, 1952, seven weeks later, showing clearing of infiltration following therapy with ACTH and cortisone.

bacteriology was negative for pathogens including tubercle bacilli.

The patient had a low grade fever for the first three hospital days and was then afebrile for three weeks save for occasional temperature elevations to 100°F. During the first six weeks he continued to raise small amounts of blood and his anemia fluctuated in severity, the hemoglobin dropping as low as 5.8 gm. Bronchoscopic examination was not remarkable save

for blood in the bronchi. Malaise and exertional dyspnea persisted. The patient was given four units of whole blood during this period, without reaction. After seven weeks he was started on a regimen of ACTH, 15 mg., intravenously in an eight-hour drip; two weeks later this was changed to cortisone, 100 mg. daily. While on this regimen the hemoptysis stopped; the malaise and shortness of breath diminished; and the chest x-ray (Fig. 5) showed clearing. At the time

of discharge, on his seventieth hospital day, the hemoglobin was up to 9.5 gm. Discharge medications included cortisone, 100 mg. daily, and oral iron.

The patient felt quite well following discharge but when seen one month later scleral icterus was noted and he was re-admitted to the hospital. Physical examination was unchanged except that the liver was now palpable two fingerbreadths beneath the right costal margin. Hemoglobin was 12.3 gm.; reticulocyte count 1.5 per cent. Chemical studies were consistent with serum hepatitis. The patient remained in the hospital for three weeks and the jaundice cleared promptly. X-ray of the chest at this time was normal. He was once more discharged on cortisone, iron and a high protein and carbohydrate diet.

In December he was seen again and was completely asymptomatic. Bronchograms obtained at this time were normal. Hemoglobin was 14.3 gm. All medications were stopped, and he has been asymptomatic and working daily since, but does not try any strenuous activity.

The patient has had extensive studies of his iron metabolism and blood clotting mechanisms performed in the Radioisotope Laboratories at the Boston Veterans Administration Hospital and the Hematology Laboratory at the New England Medical Center. All of the data are within normal limits and will be published at a later date.²⁰

DISCUSSION

The ages of onset of symptoms in these three patients were eighteen, twenty-two and twenty-four, the ages of death in the two fatal cases were twenty-two and twenty-five. One patient is in a complete remission at the age of thirty. Family histories were non-contributory. No history of exposure to toxins was obtained. The initial symptom in each case was cough, with hemoptysis shortly thereafter. Characteristically, the course was one of remissions and exacerbations. There were some remissions to a state of apparent normal health but often exertional dyspnea and easy fatigability were present. Two patients had complete clearing of the x-ray findings during remission, while the third had clearing save for some slight residual streaking.

Acute episodes were characterized by severe cough with massive hemoptysis and shortness of breath. Areas of dullness, rales and bronchial breathing could usually be detected and cyanosis was often present. During severe attacks fever, anemia and abnormal x-rays were constant findings. In none of the patients did jaundice develop but most had reticulocytosis. All studies of bleeding and clotting mechanisms and iron metabolism were normal. There was never

evidence of bleeding apart from the lungs. One patient had *D. pneumoniae* in his sputum but autopsy findings were not those of pneumococcal pneumonia. No structural abnormality of the lungs could be demonstrated.

The two deaths occurred within five days of the onset of symptoms of acute exacerbation and both were due to respiratory failure. The total duration of symptoms during a single exacerbation varied from a few days to several months. Transfusions may have been of some help in one patient in whom the last exacerbation was prolonged and remission finally occurred only after ACTH and cortisone therapy. One can only speculate as to whether or not the remission was due to these drugs. There was no response to other types of therapy.

The diagnosis of idiopathic pulmonary hemosiderosis should be considered in individuals with unexplained diffuse pulmonary infiltrations and anemia. Cough with hemoptysis, dyspnea, fever and reticulocytosis may be regarded as offering further evidence in favor of the diagnosis, while the demonstration of hemosiderin-containing macrophages constitutes a strong confirmatory finding in patients free of heart disease. Aspiration lung puncture with demonstration of siderocytes in the aspirated material has been reported as a successful diagnostic measure.²⁶ If the patient is acutely and severely ill, jaundice may be present. Past history of similar attacks should be sought and, if the disease is of long duration, evidence of right-sided heart failure may be present. The diagnosis can be accepted only after other conditions such as mitral stenosis, infectious processes and blood dyscrasias have been ruled out.

Inasmuch as three cases have been encountered on a 150 bed medical service over a four-year period, the disease probably is not too uncommon but often goes unrecognized. It certainly must occur more frequently in young adults than is indicated in the literature. As yet no conclusive evidence has been found to explain the pathogenesis of this remarkable process. It is hoped that through recognition of the condition and further study of cases as they occur some knowledge of etiology and therapy may be gained.

In Case III there was improvement following therapy with ACTH and cortisone and, although nothing can be concluded from this single observation, these drugs merit further trial.

SUMMARY

1. A brief review of the literature, and the clinical and the pathologic picture of idiopathic pulmonary hemosiderosis is presented.

2. Three instances of this disease are reported, unusual in that they are in adults, albeit in young adults. Two cases ended fatally while the third is in apparent remission.

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