MILIARY APPEARANCES IN THE LUNGS IN MITRAL STENOSIS: ENDOGENOUS PULMONARY HAEMOSIDEROSIS

BY

T. E. GUMPERT, M.B., Ch.B., F.R.C.P.

Physician, Sheffield Royal Hospital

[WITH PHOTOGRAPHIC PLATE]

Aggregations of so-called heart-failure cells in the alveolar spaces and interstitial tissues of the lungs have been known to pathologists and physicians for many years, but their recognition by means of radiographs in certain cases of mitral stenosis is a relatively new achievement and correspondingly little known. The following is a clinical record of such a case and also a consideration of the factors which may have been responsible for certain remarkable radiological appearances closely simulating miliary tuberculosis.

Case History and Clinical Details

The patient, a spare man of average height aged 36, was admitted to the Sheffield Royal Hospital on Sept. 2, 1946, with a partial left hemiparesis of sudden onset which cleared up very quickly. He had a history of several attacks of rheumatic fever between the ages of 7 and 13. His heart was alleged to have been affected in the first attack, and he was known to have had mitral stenosis and auricular fibrillation ten years before, when he was last admitted to the hospital. He had had pneumonia at the age of 23, and haemoptysis on four occasions during the past ten years.

Condition on Admission.—He was a thin man, sitting up in bed with orthopnoea. The respiration rate was 36. There was no cyanosis, oedema, or engorgement of cervical veins, but increased carotid pulsation. No hepatic enlargement or tenderness was present, the spleen could not be felt, and there was no ascites. He had slight clubbing of the fingers, but no capillary pulsation in the nails or lips. The pulse (92) was completely irregular, with a tendency to the water-hammer type. The cardiac impulse was 6 in. (15 cm.) from the midline in the sixth interspace. Percussion revealed about 1 in. (2.5 cm.) enlargement to the right. There was a systolic thrill at the apex, coinciding with a squeaking systolic murmur transmitted into the left axilla; and a mid-diastolic rumble at the apex, accentuated by exercise. A constant early aortic diastolic regurgitant murmur was present, loudest in the third and fourth left interspaces close to the sternum. The blood pressure was 140/60. No abnormal lung signs or adventitious were noted, and no further abnormalities other than partial left hemiparesis with increased tendon reflexes, slight facial weakness, and up-going toe, all on the left side.

Electrocardiograms showed typical auricular fibrillation: the fibrillatory rate was about 500 a minute and the ventricular response approximately 101 a minute before digitalization. There was marked right axis deviation. A radiograph of the chest (Plate, Fig. 1) revealed the typical mitral configuration of the heart with a prominent pulmonary artery shadow, a small aortic knuckle, a well-rounded and enlarged right auricular shadow, and a hypertrophied left ventricle with a straightish left border. Both lung fields were uniformly stippled from apex to base and closely resembled in appearance the punctate mottling of miliary tuberculosis or a finely disseminated pneumoconiosis. The pulmonary artery shadows were prominent, but there was no gross evidence of pulmonary congestion. A radiograph taken ten years before (Plate, Fig. 2) showed a typical mitral heart with increased vascular shadows but no nodulation.

The patient's progress was excellent. He made a rapid recovery from the hemiparesis, and the ventricular rate was well controlled with "digoxin." He was discharged from hospital on Oct. 23, 1946.

Discussion

So far as the heart was concerned the case was perfectly straightforward. It was an old-standing case of mitral stenosis with a small aortic leak and auricular fibrillation. The arresting finding was the radiological appearance of the lung parenchyma, which bore an extremely close resemblance to that of miliary tuberculosis or a fine discrete nodular pneumoconiosis. This was exactly the sort of case in which our forefathers first described heart-failure cells in brown induration of the lung.

The mechanism would appear to be as follows: The mitral obstruction causes an increase of pressure in the left auricle and its ultimate tributaries the alveolar capillaries. These become engorged to such an extent that repeated intra-alveolar haemorrhages occur with or without clinical haemoptysis. The intra-alveolar haemoglobin disintegrates and is converted into relatively soluble iron-free haematoidin and relatively insoluble particles of iron-containing haemosiderin. The latter act as foreign bodies and are treated as such by the phagocytic alveolar histiocytes, which engulf them just as though they were inhaled particles of a fine dust. Their transport, however, via the lymphatics of the alveolar wall, the peribronchial, perivascular, and subpleural lymphatics to local aggregations of lymphoid tissue scattered throughout the lung substance and finally to the bronchial lymph glands is probably not so readily achieved as in a true pneumoconiosis because of the chronic pulmonary venous congestion and consequent obstruction to the lymphatic drainage, with the result that most of the phagocytic haemosiderin-laden heart-failure cells remain in the alveoli, and, being radio-opaque, give the characteristic radiological alveolar stippling we see in this case. Larger aggregations in the lymphatic apparatus of the lung are thus prevented and a coarse nodulation such as is seen in a typical pneumoconiosis (silicosis) does not occur.

This view is supported by the resemblance of the stippling to the familiar radiological appearance of intra-alveolar lipiodol and still more to that of miliary tuberculosis, where lesions of a similar size and distribution occur. Moreover, if the individual nodules are examined closely with a magnifying glass, certain of them (Plate, Fig. 3) seen end-on appear to be ring-like in structure with the form of a rosette—the central hole being the terminal respiratory bronchiolus beset with the rosette-like iron-containing acini.

Review of the Literature

Similar radiological appearances have been described in other cases of mitral stenosis by Ryder and Reineke (1945), by Hanson (1945), and by Pendergrass and Leopold (1945). Kerley (1938), writing about passive hyperaemia of the lungs in mitral stenosis, states that "occasionally the combination of swollen end-on vessels and alveoli filled with heart failure cells causes a miliary appearance in both lungs." Hanson's case was proved post mortem, and the radiological appearances were those of a diffuse fibrosis of the lung resembling silicosis; but there was no nodulation, and, in spite of an industrial hazard of sandblasting for five years, microscopically and histologically the lung showed no silicosis—merely a fine fibrosis together with passive congestion. Pendergrass and Leopold refer to pseudo-nodulation limited to the upper two-thirds of both lung fields occurring in a case of mitral disease within a year of an attack of acute rheumatism with rheumatic pneumonitis. They consider the pseudo-nodulation to be due to shadows cast by iron pigment—the end-result of the haemorrhagic perivascular infiltration of rheumatic

fever—and refer to a personal communication received from Sostman, who had observed several patients with long-standing passive pulmonary congestion from mitral stenosis in whom a finely nodular radiographic appearance simulating the nodulation stage of silicosis had developed. At necropsy the individual nodules proved to be fibrotic and to contain much blood pigment, which took the specific stain for iron. In the discussion which follows their communication Enzer refers to the pseudo-nodulation which he has observed in a few cases of mitral stenosis. He considers that the lesions may be due to the organization of macrophages filled with haemosiderin. And, finally, there is the excellent recent clinical, radiological, and pathological account by Scott, Park, and Lendrum (1947) of eight cases examined post mortem. They state that “in rare cases of mitral stenosis the radiological picture is one of miliary opacities in both lungs. These opacities do not alter in appearance over long periods of time, in contrast with the changes produced by pulmonary oedema. Pathological examination shows that the opacities are due to focal deposits of haemosiderin. A number of these cases have been reported in the literature, but the changes are frequently attributed to congestion of the lungs.”

Similar appearances are also described by Pilcher and Eitzen (1944) and Reye (1945) in severe (haemolytic) anaemias of childhood, where the haemosiderosis is due to repeated intra-alveolar haemorrhages.

Clinical Investigations

I was not aware at the time of our patient's admission that mitral stenosis with chronic pulmonary venous congestion could produce radiological signs of this description and therefore did not associate the two conditions. In fact, the radiograph of the chest was so arresting and unusual that it dominated and confused the issue. A dual pathology seemed to be the only solution to the problem and miliary tuberculosis was seriously considered. Acute miliary tuberculosis was clearly out of the question: there was no pyrexia, no sweating, no hectic flush of the face, no splenic enlargement, and none of the cachexia invariably associated with this condition. The Mantoux test was negative and the cerebrospinal fluid normal in all respects. Chronic miliary tuberculosis appeared to be the answer, *faute de mieux*, but the association of mitral disease and tuberculosis is very rare and the two conditions are usually considered to be mutually antagonistic. Pulmonary sarcoidosis had to be considered, but there were no other lesions of this condition—no involvement of the skin, bones, or uveal tract. The lack of any occupational hazard negated the pneumoconioses. The yeast and mould infections of the lungs, moniliasis, aspergillosis, and coccidiosis—all of them capable of giving appearances similar to those of miliary tuberculosis—were next considered, but none of the characteristic yeasts or moulds could be seen or cultured from the sputum. There were none of the suffocative phenomena of bronchiolitis, nor did the patient seem ill enough for this condition to be seriously entertained. Similarly untenable was a diagnosis of miliary carcinomatosis of the lungs. It was not, in fact, until Dr. Grout had given me his opinion of the x-ray appearances that I was able to correlate the miliary appearances of the lungs with the mitral stenosis. It was then a simple matter for the pathologist to stain the sputum for the Prussian-blue reaction and to discover heart-failure cells in the expectoration.

Conclusion

Miliary appearances in the lungs in mitral stenosis occur from time to time. They are considered to be due to

an endogenous pulmonary haemosiderosis. It might be objected that mitral disease with chronic pulmonary venous congestion is so common in medical practice that the radiological appearances under discussion would be widely known, but this is a facile argument and of no validity. In fact, Scott, Park, and Lendrum, having become interested in the condition in 1943, have been able to collect eight cases of mitral disease showing post-mortem evidence of haemosiderosis. Only two of them, it is true, showed the typical miliary x-ray appearance during life, but one with equivocal radiological signs showed quite definite radiological (and pathological) evidence of haemosiderosis in the isolated lung after death. Conditions must obviously be peculiarly favourable to permit of this orderly deposition of pigment throughout the lung substance, and it is only by further observation and investigation that one is likely to discover the precise set of circumstances leading to a diffuse punctate pulmonary haemosiderosis in mitral disease.

Summary

A case of mitral disease with a small aortic leak and auricular fibrillation is described in which the radiological appearances of the lungs simulated miliary tuberculosis.

The case is considered to be one of endogenous pulmonary haemosiderosis due to the deposition of haemosiderin in the lung substance.

Heart-failure cells rich in haemosiderin were recovered from the sputum.

Allusion is made to strictly comparable cases and to cases in which haemosiderin is similarly deposited in certain anaemias of childhood.

The radiological appearances are reproduced and a previous radiograph, taken ten years earlier, is also shown by way of contrast.

ADDENDUM.—A not dissimilar condition is described by Elkeles and Glynn (1946) in the literature recently of a disseminated parenchymatous ossification in the lungs in association with mitral stenosis, but the radiological appearance is different in texture and distribution (in that the apices are spared) and the pathology likewise differs.

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Among the many books intended to instruct the mother in bringing up her children *The Pocket Book of Baby and Child Care*, by Benjamin Spock, M.D., is outstanding for the common sense that its author displays. It is obtainable from Messrs. Delisle, Ltd., 122, City Road, London, E.C.1, at the remarkably low price of 2s. 6d. (or 2s. 10d. including postage). Dr. Spock does not describe an ideal baby, or even a “normal” one, and imply that all should be like that. He points out that a baby can be perfectly healthy and yet sleep very little, or have a bad temper, or cry a lot. He also gives many practical instructions for understanding the minor ailments of infancy and childhood, including a brief account of mental development and disturbances. The book is enlivened by some delightful sketches, one of them depicting how to perform that difficult operation—getting a teat on to a feeding-bottle.

**TUBERCULOSIS OF THE GREAT TROCHANTER:
BARRY McMURRAY**

FIG. 1.—Erosion of great trochanter following tuberculosis of the bursa with secondary infection. Typical peripheral erosion with some sclerosis.

FIG. 2.—Long-standing tuberculous erosion: sclerosis and new bone formation due to secondary infection.

FIG. 3.—Tuberculosis of great trochanter arising in the bone. Extensive destruction of centre of trochanter but no involvement of soft tissues.

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FIG. 1.—Radiograph of the chest.

FIG. 2.—Radiograph ten years before Fig. 1.

FIG. 3.—Infraclavicular area magnified.