Pulmonary alveolar microlithiasis

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Summary

A case of pulmonary alveolar microlithiasis is reported. This is of especial interest as it is the first case to be reported from Africa South of the Sahara. The clinical presentation of the patient follows a similar pattern as that of cases already described by other workers. The diagnosis in this case was made in life by a combination of radiological examination and a lung biopsy. The clinical presentation and aetiology of the condition are briefly discussed.

Introduction

'Microlithiasis alveolaris pulmonum' was the name given by Ludwig Puhr in 1933 to a remarkable case of pulmonary calcification, in which the lungs were increased four-fold in weight and a great number of the alveoli were filled each by a laminated 'stone', the radiograph having shown opaque lung fields with fine miliary peripheral opacities. This condition was however first described by Friedreich in 1856. The striking feature is the diffuse and extensive radiological appearance in contrast to the minimal or absent symptoms and signs early in the disease. The disease is world-wide and has a tendency to occur in siblings. Sosman *et al.* (1957), Finkbiner, Decker and Cooper (1957) and several other authors (Greenberg, 1957; Chinachoti and Tangchai, 1957; Viswanatham, 1962; Badger, Gottliers and Gaensler, 1955) have described the disease in its varying forms in different races. This disease is extremely rare, and so far as can be ascertained, it has never been reported from a Nigerian. In fact, the only report in the literature of this disease on the African continent was that of two Egyptian Arabs described by Abdel-Hakim *et al.* (1959).

This is a report of the first case diagnosed in an 8-year-old schoolgirl, who presented at the University

College Hospital, Ibadan, with an unrelated medical complaint, but whose lesion was recognized because of a chance radiological finding.

Case report

An 8-year-old schoolgirl presented at the general outpatient clinic of the University College Hospital, Ibadan, with rashes on her limbs, fever, and tiredness for 3 weeks. She denied any history of cough or dyspnoea. There was no history of a previous medical illness. On examination, she was an intelligent and co-operative girl, a little small for her age. She was not dyspnoeic, cyanosed, or anaemic. There were a few discrete lymphatic glands in both axillae. Her pulse rate was 60/min and regular. Blood pressure was 80/40 mmHg. The heart sounds were normal. Examination of the chest revealed dullness to percussion on the right lung base posteriorly. Air entry was reduced in both lung bases. The liver was enlarged 4 cm below the right costal margin. It was smooth and non-tender. There was maculo-papular rash of patchy distribution on both upper limbs. The rash was not itchy. Investigations included a chest X-ray (Fig. 1) which showed diffuse, minute, sand-like nodulation all over the lung fields. This was thought to be unusual for miliary tuberculosis, which is frequently seen in this environment. Packed cell volume was 32%, total white cell count was 3600/mm³. The differential cell count revealed neutrophils 45%; lymphocytes 40%; eosinophils 15%. ESR was 64 mm in 1 hr (Westergren). Faecal examination revealed ova of *Ascaris* and of *Trichuris*. Blood film for microfilaria was negative. The eosinophil count fell to 4% after the patient had been treated with anthelmintics. Sputum examination for acid-fast bacilli was repeatedly negative. The cytology of the sputum did not reveal malignant cells. Mantoux test was also negative. X-rays of the abdomen, skull, hands and feet were normal. The LE cell test was

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negative and the test for rheumatoid factor was negative. The liver function test was normal. The total plasma protein was 7.4 g/100 ml with albumin of 3.1 g/100 ml, and globulin of 4.3 g/100 ml. The serum calcium was 9.8 mg/100 ml, and serum phosphorus was 3.6 mg/100 ml.

FIG. 1. Chest X-ray showing diffuse, minute nodulations all over the lung fields.

FIG. 2. The histopathology of the lung showing the lamellar arrangement of the concretions in the alveoli.

A provisional diagnosis of alveolar microlithiasis was made with reservation as the condition was unheard of in this environment. A lung biopsy, using Steel's high speed drill needle, was performed. The report of the pathologist was as follows:

'The specimen received consisted of few fragments of greyish-white tissue and some blood clots. Histologically, sections of lungs are truly remarkable and, at first sight, recall a very psammomatous meningioma of a mass of prostatic calculi. There is patchy fibrosis in places. In addition, there are spaces (apparently alveoli) occupied by rounded concretions of concentric pattern (microliths or calcospherites) which had evidently receded from the spaces they originally filled, the split occurring so as to leave a fine outer layer adherent to the surrounding alveolar walls. Some of these alveolar walls consist of avascular strands of fibrous tissue containing scanty chronic inflammatory cellular infiltration (Fig. 2). The overall appearances are consistent with lung in microlithiasis alveolaris pulmonum'.

The parents, two brothers and one sister of the girl were accordingly examined. Their radiographic examinations were normal. This girl is being followed-up regularly and, up to the time of writing, remains asymptomatic.

Discussion

The aetiology of pulmonary microlithiasis is unknown. Good accounts of this have, however, been attempted by Sharp and Danino (1953). Sosman *et al.* (1957) and Kent, Gilbert and Meyer (1955). Sosman *et al.* remarked that the only established facts are that this rare condition may occur in young people, with no history or sign of antecedent disease; may lie dormant for 10–25 years, and then reveal its presence by slowly increasing pulmonary insufficiency, and signs of right heart failure. The only aetiological clue so far discovered is the fact that the disease tends to occur in families, which suggests some inherited abnormality or defect in pulmonary metabolism. Kent and colleagues (1955) suggest that the disease is the result of a peculiar exudative response to a variety of insults such as mitral stenosis, recurrent pneumonia and exposure to dusts. However, evidence of these was not found in the present case nor in many of the other cases quoted.

The pattern at presentation of the disease in this case follows that described in many cases in the literature (Sosman *et al.*, 1957; Finkbiner *et al.*, 1957; Greenberg, 1957; Chinachoti and Tangchai, 1957). The patient was 8 years old at presentation. The youngest case ever reported in the literature was 6 years old. This disease is reported to be even rarer below the age of 10, the average age being

between 30 and 50 years. There was no familial tendency in the patient's family.

While cases like this are admittedly rare, there is the probability of misdiagnosis in some areas, where the physicians are not aware of its mode of presentation and where there are no facilities for adequate investigations. Such cases very often can be mistaken for miliary tuberculosis and the patients placed unnecessarily on anti-tuberculosis therapy. However, the exercise tolerance of the patient, the continued absence of tubercle bacilli from the sputum, the discrepancy between the mild clinical symptoms and the high degree of radiographic opacity should exclude severe tuberculosis. The absence of iron from the sections of the lungs in this case, the miliary pattern, which fell into a different category from those of haemosiderotic lungs, described by Laubry and Abbas (1948) and Lendrum, Scott and Park (1950) and the relative freedom of the patient from recurrent attacks of dyspnoea and cyanosis may help to exclude haemosiderosis. The clinical history and the extent and pattern of distribution of the pulmonary opacities are likely to distinguish such diseases as silicosis, histoplasmosis and schistosomiasis.

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Paravertebral and peripheral ligamentous ossification: an unusual association of hypoparathyroidism

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Summary

A 62-year-old man with idiopathic hypoparathyroidism and extensive paravertebral and ligamentous ossification is reported. The clinical and radiological findings of this, and other reported cases, are discussed and compared with other causes of paravertebral ossification.

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Introduction

Paravertebral and ligamentous ossification appears to be an unusual complication of hypoparathyroidism. The association has been reported in idiopathic and post-operative hypoparathyroidism (Büscher, 1948; Salvesen and Böe, 1953; Gibberd, 1965; Chaykin, Frame and Sigler, 1969).

The clinical, biochemical and radiological features of another case are described and the differential diagnosis is discussed.