Case Report

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TANAFFOS

Pediatric Pulmonary Alveolar Microlithiasis: A Case Report

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Pulmonary alveolar microlithiasis is a rare infiltrative pulmonary disease characterized by deposition of microliths in the alveoli. In this case report, we present a case of a 5 year-old girl with complaints of common cold and loss of appetite. Chest radiograph showed innumerable small, dense nodules, diffusely involving both lungs. High-resolution CT scan illustrated widespread micro-nodular infiltration, diffuse ground-glass attenuation areas predominantly in upper anterior regions and septal thickening. Trans-bronchial biopsy confirmed the diagnosis.

Key words: Microlithiasis, Pulmonary, Pediatric

INTRODUCTION

Pulmonary alveolar microlithiasis (PAM) is a rare idiopathic condition characterized by widespread intra-alveolar deposition of spherical calcium phosphate microliths .The exact etiology and pathogenesis have yet to be clearly identified (1-4). It has a familial incidence with an autosomal recessive inheritance pattern (1,3,4). There is no known calcium metabolism disorder. Microliths range from 0.01 to 3 mm in size and are composed of calcium phosphate (1,4,5). It typically demonstrates sand like calcification distributed throughout the lungs; distribution is bilateral with middle to lower zone predilection. CXR usually shows diffuse tiny nodules in milliary pattern. Black pleural lines may also be evident on CXR (1). High-resolution CT scan usually illustrates widespread micronodules, diffuse ground-glass attenuation areas, septal thickening, and in some cases black pleural lines-predominantly in the basal regions (6). The diagnosis is usually confirmed by transbronchial biopsy (5).

CASE SUMMARIES

A 5 year-old girl with common cold consulted a pediatric physician. On her CXR bilateral reticulonodular opacities were noted (Figure 1). She was treated with the diagnosis of pneumonia. She came back after 6 months for further evaluation but there was no change on her CXR. She was referred to a university hospital because of abnormal CXR. During this period she only had loss of appetite. On her physical examination: there was no evidence of clubbing or cyanosis in extremities and no

hepatosplenomegaly. On auscultation bilateral fine crackles were present. She was diagnosed and treated for TB in the hospital with no evidence of improvement.

Her family history was negative for TB and alveolar microlithiasis.

Vital Signs: RR: 20 PR:100 T:37˚C BP:95/65 mmHg

Laboratory data:

VBG: PH: 7.3 PCO2:48 HCO3:23 PaO2:55

CBC: Hb: 13.6 WBC: 6.99 (N: 39%, L: 59%) PLT: 406000

ESR: 12 RP: Negative LDH: 560 HIV: Negative

PPD: 5mm; after 3 gastric lavages the result was negative for TB.

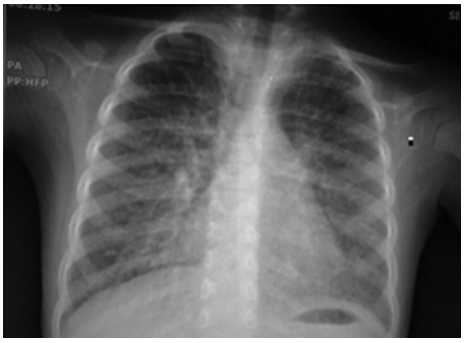

Figure 1. Chest X-ray showing bilateral diffuse high density micro-nodular opacities.

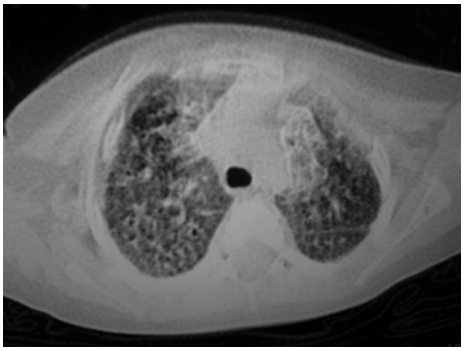

Figure 2. Micro-nodular infiltration and bilateral ground glass opacities in upper lobes. Anterior dense reticulation especially on the left side.

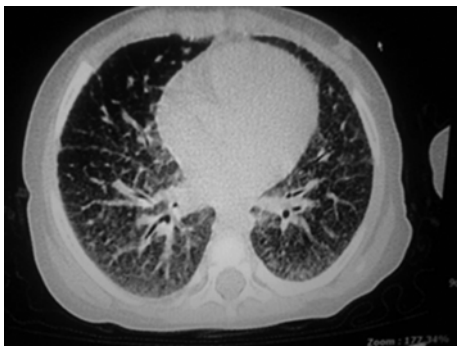

Figure 3. HRCT of thorax showing bilateral diffuse micro-nodular opacities

DISCUSSION

Pulmonary alveolar microlithiasis is a rare disease (1) and its radiological picture is very characteristic. Bilateral sand-like micro-nodules of calcific density are mostly seen in the middle and lower zones near the apices. Radiological findings are diagnostic; but they can mimic miliary tuberculosis, berylliosis, hemosiderosis, silicosis, fungal infections, sarcoidosis and carcinomatosis. Despite extensive radiological changes, most patients remain asymptomatic (1,4) and the condition is usually diagnosed incidentally on CXR taken for other reasons. CT findings are innumerable tiny sand-like calcified micronodules spread throughout both lungs (3).

Mikhaylov (1, 4) was the first to report the familial occurrence of pulmonary alveolar microlithiasis. More than 50% of reported cases of alveolar microlithiasis have a familial incidence occurring in siblings. A few cases of parent and child involvement have also been reported. It seems that a genetic factor is responsible for this familial pattern. In genetic conditions, older patients are expected to manifest more severe pathological changes (1,4,5). But, it was not apparent in our patients.

An apparent feature of alveolar microlithiasis is the lack of significant symptoms despite extensive radiographic changes (1). Patients may become symptomatic between third and fourth decades; the disease may progress slowly and lead to progressive dyspnea with or without cough. It will finally end in respiratory insufficiency and cor pulmonale (1, 4, 5).

Complications due to this disease are rare and consist of respiratory insufficiency, clubbing, cyanosis, and pulmonary hypertension (1, 4).

No effective medical treatment is available to reduce or stop the progression of disease. There are some palliative treatments such as systemic corticosteroids, calcium-chelating agents and serial bronchopulmonary lavages which have been shown to be ineffective. Diphosphonate is used to reduce calcium phosphate precipitation in pulmonary alveoli (1,2).

Based on previous studies lung transplantation is the only possible treatment for end-stage cases (3, 4, 6).

CONCLUSION

In this article we presented a 5 year-old girl with alveolar microlithiasis which is rare in children. As discussed, the pattern of involvement is different from the usual cases. The distribution of microlithiasis is usually more prominent in inferior and posterior regions, but here we reported a case of microlithiasis which was more severe anteriorly. Also, its occurrence in childhood is unusually rare.

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