Tanaffos (2007) 6(3), 75-78

A 28-Year-Old Man with Chronic Cough and Dyspnea

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WHAT IS YOUR DIAGNOSIS?

A 28 year - old man presented with productive cough and mild dyspnea for the past seven years, which have been aggravated in recent months. He was single and lives with his parents (who are first maternal cousins) and his brother and three sisters .He does not have any major medical or surgical history. He does not have any history of transfusion, allergy, or drug abuse. He had smoked one pack of cigarette per 2.5 years but quit three years ago. His work was crafting gold jewelry and also building construction since the age of twenty. There was no important family history. Clinical exam was entirely normal and the results of pulmonary function tests (PFT) and chest plethysmoghraphy (performed twice) were normal.

Blood leukocyte count, differential, hemoglobin, platelet, biochemistry, electrolytes, renal and liver function tests were all in normal limits .PPD was negative. Echocardiography was normal. CT of the paranasal sinuses was normal. Chest x- ray (Figure 1) and lung HRCT (Figure 2) were performed. ***(Tanaffos 2007; 6(3): 75-78)***

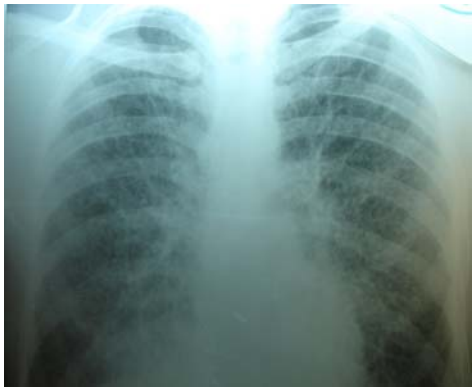

Figure 1. CXR of patient

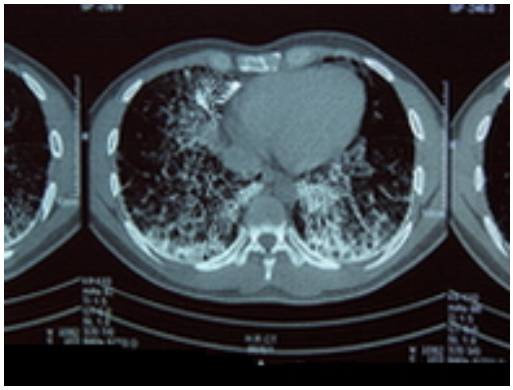

Figure 2. CT- Scan of patient

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Diagnosis: Pulmonary Alveolar Microlithiasis

The chest X- ray revealed bilateral reticular infiltration of both lungs (Figure 1).

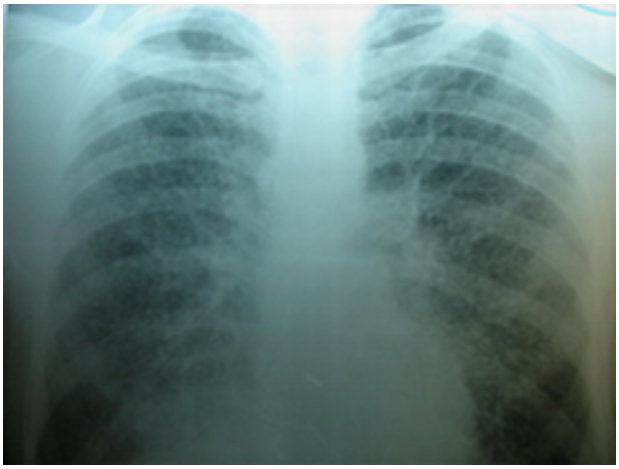


Figure 1. Bilateral reticular infiltration.

HRCT of the lung demonstrated diffuse bilateral, symmetrical, interstitial thickening of the lungs and pleura along with profuse interalveolar, septal and parenchymal nodules associated with prominent calcification in the middle and lower zones of both lungs, suggesting alveolar microlithiasis (Figure2).

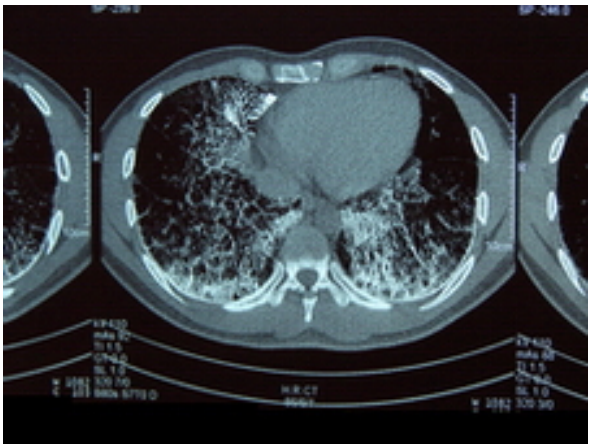
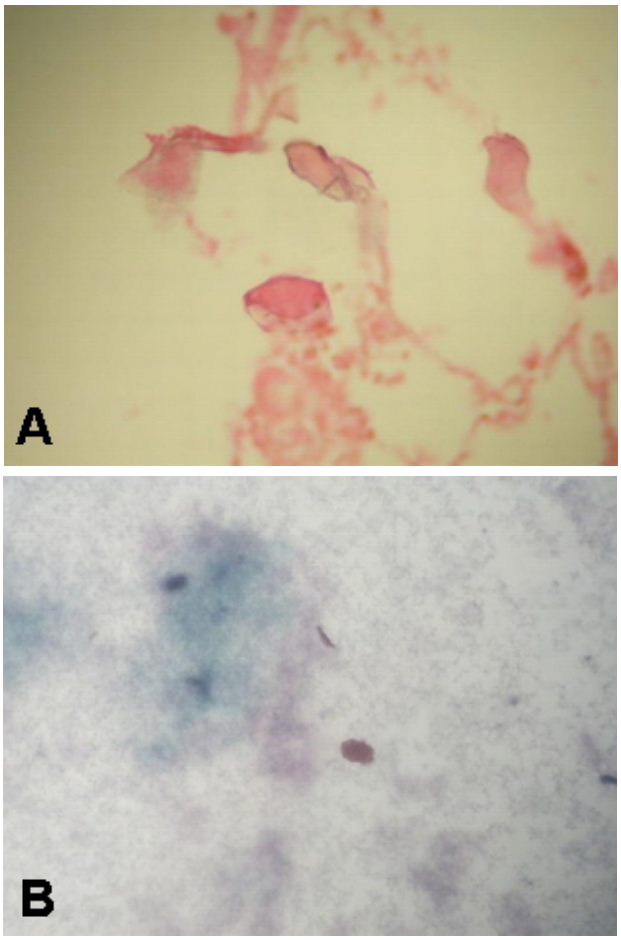


Figure 2. CT- scan of patient.

A transbronchial lung biopsy (TBLB) was performed which revealed bronchiolar mucosa infiltrated by inflammatory cells, consisting of lymphocytes, plasma cells and goblet cells.

The lung parenchyma was characterized by thin – layered alveolar spaces, laminated with basophilic calcifications, encircled by areas of mild inflammation (Figure 3).

Based on histopathologic findings and pulmonary function tests, the patient was treated symptomatically and an influenza virus vaccination was injected.



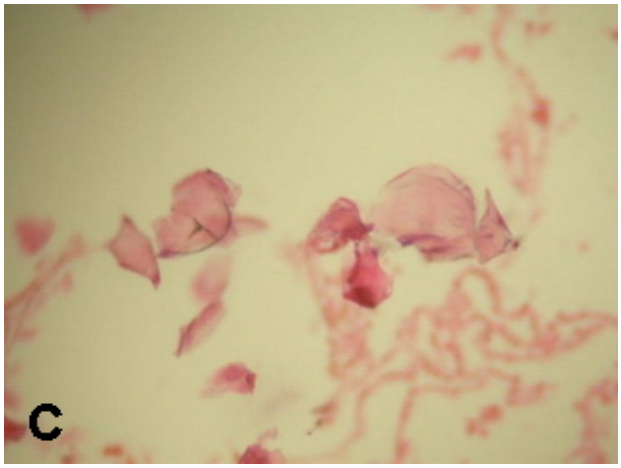


Figure 3. (A, B, C) Microscopic examination.

He is currently under observation on a regular out – patient basis.

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Pulmonary alveolar microlithiasis (PAM) is a rare idiopathic disease with unknown etiology affecting the lungs typified by deposition of calcium phosphate microliths within alveoli (1).

It affects people of all ages and the transmission mechanism is thought to have an autosomal recessive pattern. Recently, the cause of PAM has been determined to be a gene mutation in the sequence governing the type 2b sodium phosphate co-transporters (SLC34A2), expressed by alveolar epithelial cells. (2)

A total of 424 cases have been reported worldwide up to the end of 2001, 269 of them were sporadic and showed a predominance in males and 155 of them were familial cases with females more affected (3).

The highest number of cases have been reported from Europe and Asia, while the countries with the great numbers of reported cases were Turkey, Italy and the USA (4).

Most patients are asymptomatic for several years or even decades and generally, the disease is found incidentally during clinical investigations unrelated to the disorder.

In some cases it can be progressive leading to severe respiratory failure and death. (1)

The onset of this potentially lethal disease varies from the neonatal period to older ages. (5)

In reviewing over 300 individuals reported with pulmonary alveolar microlithiasis, the age of clinical onset was highly variable (5-41 years), and there was a great discrepancy between radiological features and clinical symptoms. The age range at the time of diagnosis is varied, but the diagnosis is usually made between the ages of 20 and 30 years. The condition usually evolves over 10 to 20 years and is sometimes very indolent (6, 3).

A sandstorm-appearing pattern on chest roentgenogram is the typical diagnostic finding (7).

There is no known treatment for PAM but lung transplantation has been performed in severe, progressive cases.

In conclusion, this report demonstrates the importance of radiological findings in diagnosis of PAM which may be confirmed with pathologic examination of the lungs as was done in this case.

REFERENCES

1. Murray and Nadel .Textbook of Respiratory Medicine. –June 4th ed. 2005.p. 1749-50.
2. Corut A, Senyigit A, Ugur SA, Altin S, Ozcelik U, Calisir H, et al. Mutations in SLC34A2 cause pulmonary alveolar microlithiasis and are possibly associated with testicular microlithiasis. ***Am J Hum Genet*** 2006; 79 (4): 650- 6.
3. Castellana G, Gentile M, Castellana R, Fiorente P,

Lamorgese V. Pulmonary alveolar microlithiasis: clinical features, evolution of the phenotype, and review of the literature. ***Am J Med Genet*** 2002; 111 (2): 220- 4.

4. Ucan ES, Keyf AI, Aydilek R, Yalcin Z, Sebit S, Kudu M, et al. Pulmonary alveolar microlithiasis: review of Turkish reports. ***Thorax*** 1993; 48 (2): 171- 3.
5. Esguerra Gomez G, Lochtem-Berger E, Santanaria A, Carvajal L, Jimenez-Penuela B, Saaibi E, et al. Familial pulmonary alveolar microlithiasis: four cases from Colombia, S. A.; is microlithiasis also environmental disease. ***Radiology*** 1959; 72 (4): 550- 61.
6. Cale WF, Petsonk EL, Boyd CB. Transbronchial biopsy of pulmonary alveolar microlithiasis. ***Arch Intern Med*** 1983; 143 (2): 358- 9.
7. Balikian JP, Fuleihan FJ, Nucho CN. Pulmonary alveolar microlithiasis. Report of five cases with special reference to roentgen manifestations. ***Am J Roentgenol Radium Ther Nucl Med*** 1968; 103 (3): 509- 18.