Case Report

Pulmonary alveolar microlithiasis

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ABSTRACT

We report a patient with pulmonary alveolar microlithiasis who was admitted to King Abdul-Aziz Hospital, Makkah, Kingdom of Saudi Arabia with chest pain, shortness of breath, dry cough and swelling of lower limbs. The patient underwent chest radiographs and CT scan showing multiple diffuse, almost symmetrical bilateral micronodular opacities of calcific density. The diagnosis was confirmed after percutaneous lung biopsy from the patient. Cardiokinetics, diuretics and oxygen were administered with slight improvement.

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Pulmonary alveolar microlithiasis (PAM) is a rare pulmonary disorder. The etiopathogenesis is still unknown. Many reported cases showed a familial occurrence, which suggest that it could be a genetic disorder. Chest roentgenograms showed a characteristic "sandstorm" picture with masking of cardiac and mediastinal shadows and bilateral micro opacities of calcific density. Therapeutic measure is only partially successful including broncho alveolar lavages. The only valid treatment is lung transplant.

Case Report. A 45-year-old Bangladeshi lady came to the emergency room in King Abdul-Aziz Hospital, Makkah, Kingdom of Saudi Arabia with complaints of breathlessness and chest pain for the past 2-years, associated with dry cough and swelling of both lower limbs. She had recurrent attacks of dyspnea and chest pain with frequent admissions to different local hospitals for the past 2-years. She was being treated at various hospitals, as a case of tuberculosis and chronic obstructive pulmonary disease. She had been working in a cotton cap stitching factory for the past 5-years.

On examination, she was febrile with a temperature of 38.5°C, tachypnoeic (25 breath/min) and had moderately pitting edema at the ankles. She was not cyanosed and had no clubbing or lymphadenopathy. There was tachycardia (pulse 110/min) and bilateral basal crackles. Abdomen was distended with evidence of free fluid. The rest of the examination was normal. After a chest x-ray, she was transferred to the intensive care unit (ICU) for close monitoring with provisional diagnosis of congestive heart failure. She was further worked up in ICU and her investigations revealed a complete blood cell count with white blood cells 8.3 x 10^9/L, hemoglobin 11.1 gm/dl and platelets of 316 x 10^9/L. Her blood chemistry was within normal limits except for hypokalemia of (Boltzmann constant = 2.6 milliequivalent/L). The arterial blood gas showed hypoxemia with partial arterial oxygen tension of 46.7 mm Hg, partial arterial pressure of carbon dioxide in arterial 39.4 mm Hg, pH 7.38, oxygen saturation of 89.7%. Chest x-ray showed both lung fields diffusely occupied by a discrete pinpoint high density calcific opacities resembling grains of sand and each measuring not more than one mm (**Figures 1a, b & c**). The lesion being predominantly basal, which the anatomic land marks had become completely obscured, showed a "white out" lung. The computed tomography (CT) scan confirmed the above mentioned calcific

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Figure 1 a

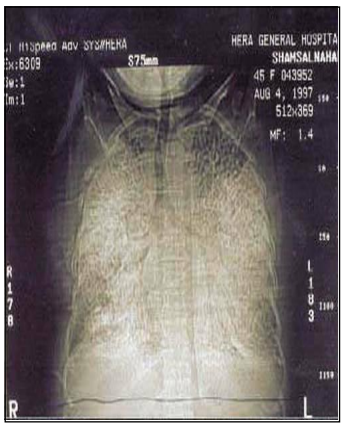

Figure 1 c

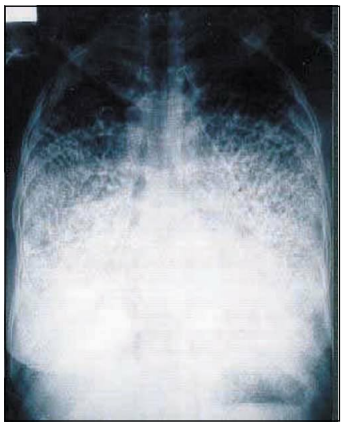

Figure 1 b

Figure 1 - Postero anterior radiograph of a middle aged female patient showing **(a, b & c)** A wide spread uniform sized miliary calcific nodulations. Heart is seen as negative shadow.

density, mainly in basal and posterior areas. A peripheral curvilinear density was demonstrated which likely represents sub pleural fibrosis rather than pleural calcification (**Figures 2a & b**). Soft thickening was noted between the chest wall and calcified lung, which was seen on the chest x-ray film as transradiant peel to the lung. Multiple blebs were also noted. Lung function test showed a restrictive pattern. Her forced vital capacity was 1.07 L (55% of predicted value), forced expiratory volume in one second was 1.00 L (60% of predicted value), electrocardiogram showed a pulmonale. Echocardiography had a normal functioning left ventricle, dilated right atrium and right ventricle. On fiberoptic bronchoscopy, there was a wide spread bronchial mucosal redness. Bronchial washing was negative for acid fast bacillus and microbial culture and the cytology showed no malignant cells. Percutaneous lung biopsy was carried out by Trucut biopsy needle from left mid axillary line through the fifth intercostal space. Histopathological report revealed lung tissue consisting of pulmonary alveoli containing microliths or calcospherites. The interstitium showed black pigmentation in focal areas (anthracosis). Hence, patients diagnosis was confirmed to be pulmonary alveolar microlithiasis.

The patient stayed in the hospital for 2-weeks. During the duration, she was given diuretics, bronchodilators and oxygen. She had marginal symptomatic improvement. After leaving the hospital she traveled back to her home country and hence was lost for follow up.

Discussion. Pulmonary alveolar microlithiasis is a rare disease, often with familial distribution.[1] It suggests that an inherited trait may be involved. Most probably it is an autosomal recessive gene.[2,3] Ucan et al[4] revisited 52 cases of PAM in Turkish

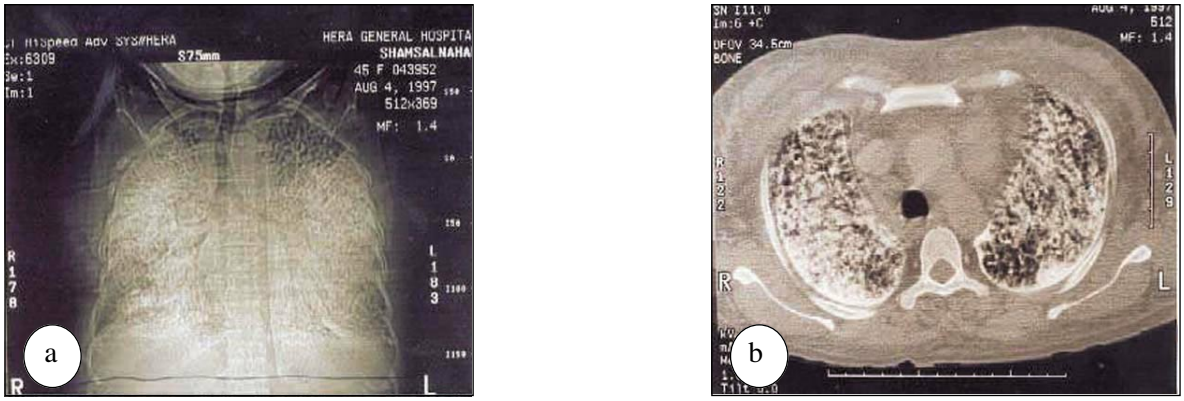

Figure 2 - High resolution computed tomography scan through the lung of one millimeter thickness showing **(a & b)** Clear accentuation of reticular structures with dense miliary sized nodules clearly identifiable, causing nodular thickening of lobular septa.

literature from 1962-1993 and found familial occurrence in 25% of patients. They made an assumption that the gene is most common in Turkish people, which constitutes 23% of all the cases reported. Some physicians suggest that PAM can develop within alveolar spaces either due to calcium salt sedimentation, an increased endoalveolar pH or after alveolar cell insult.[2,5]

The incidence of disease in males and females is equal. The average age of onset is between 30-40-years,[6] but the range is larger with disorder being described from infancy to an 80-year-old person.[2,4,6] Approximately 25% of patients in the literature was presented at less than 10-years of age and in Japan peak presentation is in childhood, a fact that has been ascribed to the greater use of screening radiology.[3] The patient with PAM usually presents with cough and progressive dyspnea. The disease slowly develops into pulmonary fibrosis and leads to chronic respiratory failure and corpulmonale.[2,6] Pulmonary function tests are often normal or near normal even with extensive radiographic changes with the progression of disease, however, a restrictive pattern of lung volume develops as in our patient. The impairment of gas transfer with disturbed ventilation perfusion ratios have been reported.

These patients are often discovered on a routine radiography at an asymptomatic stage in spite of gross changes on the chest x-rays.[7] Radiological features shows both lung fields being diffusely occupied by discrete pin point high density calcific opacities resembling grains of sand. The lesions are predominantly basal and in the upper and middle zones.[2] The micronodular opacities in PAM usually appears from the onset itself as calcific density, sometimes chest x-rays also highlights visible subpleural blebs, which may burst and worsen the clinical picture by inducing pneumothorax.[8] The radiological findings strongly favors the diagnosis which may be confirmed by histopathological examination. The tissue specimen are taken by transbronchial, percutaneous or open lung biopsies. In our case, the initial diagnosis was made clinically and radiologically and confirmation of diagnosis was carried out by a percutaneous lung biopsy. The microscopical result showed the lung tissue consisting of pulmonary alveoli containing microliths or calcospherites.

In one of the studies for microlith chemical composition, Le Charpentier et al[9] found calcium, phosphorus, potassium, sodium, magnesium and carbon were present. Therapeutic broncho alveolar lavage (BAL) has been successfully performed in some conditions like alveolar proteinosis but it has been shown to be ineffective in PAM. Only a small amount of calcospherites may be removed by BAL, due to the large diameter than that of bronchioli.[11] Mascie-Taylor et al[10] washed the lung of a patient under block anesthesia with 2 litres of saline solution, withdrawing 14.5 gm of microliths. The patient's clinical condition, however, did not improve in spite of this attempt. Treatment with sodium etidronate may be helpful in Paget's disease and in progressive ossifying myositis, as it prevents the precipitation of hydroxy apatite crystal. In recent years, sodium etidronate was also employed in PAM patients, however, the results are not yet satisfactory. Prognosis is variable in many patients who remains asymptomatic with stable chest radiographs for many years,[12] others after long period of stability, still experience pulmonary fibrosis or corpulmonale and they ultimately die of the disease.[13,14] Respiratory functions in the end stage of PAM are severely impaired and long term oxygen therapy or mechanical ventilation may be required.[15] Stomitis et al[16] believed that bilateral lung transplantation is the only recognize therapy and the excellent results are seen in a transplanted patient after 18-months.

In conclusion, the treatment of PAM has not been clearly defined.

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