Original Research Paper

Volume-8 | Issue-7 | July-2018 | PRINT ISSN No 2249-555X

Radiodiagnosis

"PULMONARY ALVEOLAR MICROLITHIASIS LEADING TO END STAGE PULMONARY FIBROSIS IN A YOUNG: A CASE REPORT".

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ABSTRACT The aim of my study is to show that Pulmonary alveolar microlithiasis (PAM) can present at the end stage of pulmonary fibrosis even in young population. It can also have rapid progression.

A 20-year-old Girl, hailed from a village in Orissa, India, presented with gradual onset shortness of breath, weight loss and clubbing of fingers. Similar complaints were found in her real sister at home and in family history of grand mother's who died of breathing difficulty.

On clinical examination, pulmonary function test and X-Ray chest, a possibility of PAM was suspected. Further evaluation by CT scan chest confirmed its entity. It was in stage IV of CT classification.

She was treated with steroids and with Alendronate Sodium. The suggestion of surgical option was given. After that, we lost to follow up the patient

KEYWORDS :

DISCUSSION:

PAM inherited autosomal recessive abnormality which was first reported by Norwegian Harbitz in 1918; accordingly, it is also known as Harbitz′ syndrome. It was named as Pulmonary Alveolar Microlithiasis by Ludwig Puhr of Hungary in 1933[2]. More than 1022 cases have been reported worldwide until May 2015; [7] from all the continents of the globe without any particular geographic or racial distribution. Asia and Europe shared nearly one-third cases each, Highest cases observed in Turkey and Italy. In India, it was first reported by Viswanathan R in 1962. [1, 14] Approximately, 80 cases have been reported in India until Dec 2014 from different states: Himachal Pradesh & Jammu and Kashmir (northern); Gujarat, Orissa, and Chhattisgarh (central Indian belt). [7] These Indian states are having either extreme hot weather or else extreme cold weather. Hence, hypothesis of extremely high or low temperature can be made as one of the factors in pathogenesis of PAM.

PAM occurs in both sexes with a slight predominance in males than in females. It is frequently observed from 0 to 40 years of age. The appearance is usually late, after 30 years, until then it remains silent. In India, 43 males, 36 females and 1 unspecified PAM cases were reported until December 2014. [7] Maximum life expectancy noted is 56 years till date in a post-transplant patient. [5]

During my practice in Chhattisgarh, India, Our case was referred from a village in Orissa state for tertiary care. She was 20 years girl, came with gradual onset shortness of breath which increased on exertion. Significant weight loss observed in last few months by 10 KG. She had clubbing of fingers giving a clue of severe pulmonary pathology. Her real sister at hometown has similar complaints on and off with less severity which indicated familial link. Family history of grandmother's death due to breathing difficulty suggesting generation skips of the inheritance.

Usually, PAM is misdiagnosed clinically for allergies, bronchitis, bronchial asthma. [12]

On clinical examination, assisted breathing with mild pallor noted. Loose fitted clothes with tachypnoea observed. No evidence of cyanosis. Ungual clubbing was seen. She was sent for pulmonary function test which revealed typical features of a restrictive pattern of breathing with reduced forced vital capacity (FVC) and elevated forced expiratory volume in 1 s(FEV1) /FVC. Reduced total lung capacity and tidal volume. [8] The lung age was 85 years. DLCO was significantly reduced. These findings were suggesting hypoventilated alveoli with respect to pulmonary capillary blood flow, leading to reduced O2 tension in the mixed arterial blood [8]

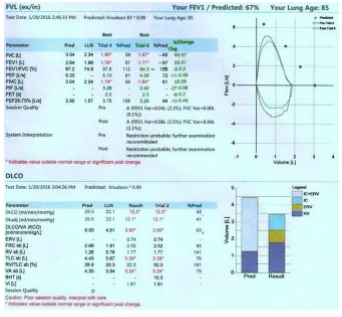

FIGURE 2: pulmonary function test chart with O2 diffusion capacity chart and bar graph showing reduced forced vital capacity (FVC) and elevated forced expiratory volume in 1 s(FEV1) /FVC. Reduced total lung capacity and tidal volume [PAM 2]. The lung age was 85 years. DLCO was significantly reduced.

Chest radiography was ordered. There was striking clinico-radiological dissociation seen. There was 'Snowstorm' / granular appearance of bilateral lung fields, mainly in bases, sparing the apices. Bilateral black pleural lines and white fissural lines and stony lung diagnosed at the bases [Figure 1]. On Radiography, possibilities of calcified Granulomatous pathology, Pneumoconiosis, and PAM were considered.

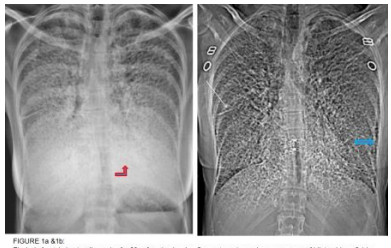

FIGURE 1a &1b:
Fig 1a is frontal chest radiograph of a 20 y. female showing Snow storm / granular appearance of bilateral lung fields, mainly in bases with relative sparing the apices and stony lung at the bases (red arrow).
Fig 1b is frontal CT topogram image showing similar appearance with more pronounced bilateral black pleural lines (blue arrow) and white fissural lines (white arrow).

For differential diagnosis, refer Table below..

Differential Diagnosis table

X-Ray:	X ray	CT:	LUNG BIOPSY
Tuberculosis	Empty Cavitations with asymmetric calcified granulomas	Consolidation, Cavitation, Tree in bud appearance, Miliary Nodules, lymphadenopathy and pleural effusion.	Caseating granulomatous nodules

Calcified miliary histoplasmosis (immunocompromised)	Diffuse calcified granulomas	Extensive calcified miliary Micronodules.	Noncaseating granulomatous nodules, yeast buds
Sarcoidosis	Hilar masses	Miliary nodules, random distribution, nodular septal thickening	Noncaseating granulomatous nodules with exclusion of other causes
Asbestosis	Sheet like pleural calcifications	Upper lung fibrosis, round collapse, pleural calcifications	Demonstration of asbestos particles
Stannosis	Intense metal density nodules	Metal density well-defined interstitial miliary nodules, extensive	Demonstration of tin particles

Further evaluation by CT scan chest was ordered.
CT chest can depict evolution of the disease and can classify PAM in four stages [7]:

a) faint alveolar calcifications by microliths - delineates the early disease stage, generally present in childhood.
b) dense scattered calcific Micronodules giving appearance of 'Sandpaper or Berries', generally present in childhood or young adults.
c) extremely dense calcified nodules with sandstorm appearance, obscuring the cardiac and diaphragmatic margins. Calcification of the pleural lining and interlobular septa which partly masks the micronodules found.
d) diffuse microliths, calcific agglomerates with intense involvement of the pleural lining and interlobular septa. In this phase, calcific fibrosis happens with compensatory paraseptal emphysema and air cysts in the upper lobes, which might result in pneumothorax. This stage is generally seen in older adults and elderly. It represents the advanced form of PAM.

In our case, CT findings were shocking. Almost all the findings of PAM were present. Crazy paving pattern, Alveolar Micronodules, Septal thickening and calcification, sheet-like pleural plaques, obscured margins of pulmonary vessels, predominant calcifications/ ossification of parietal and mediastinal pleura in chest bases. Apices were relatively spared; however, few blebs were seen. Peripheral paraseptal emphysema was striking. So also, rows of variably sized cysts were seen mainly in anterior segments of bilateral upper lobe and right middle lobe suggesting Interstitial fibrosis. Hence, it was labeled as stage IV of evolution.

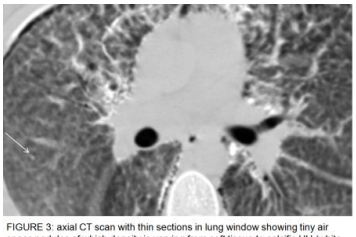

FIGURE 3: axial CT scan with thin sections in lung window showing tiny air space nodules of which density is varying from soft tissue to calcific HU (white arrow).

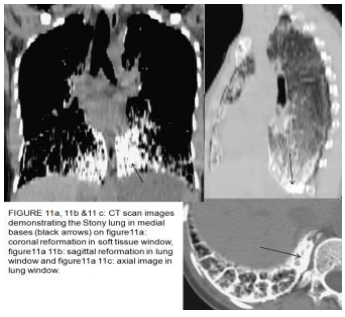

FIGURE 11a, 11b &11 c: CT scan images demonstrating the Stony lung in medial bases (black arrows) on figure11a: coronal reformation in soft tissue window, figure11a 11b: sagittal reformation in lung window and figure11a 11c: axial image in lung window.

Role of BAL is controversial. Tc99m-MDP scintigraphy can demonstrate abnormal pulmonary uptake which can add value to the diagnosis and can avoid unnecessary open lung biopsy. [6] Lung Biopsy can give final histopathological diagnosis demonstrating calcipheres; endobronchial route is preferred. However, imaging is sufficient enough to diagnose the PAM.

She was treated with Oxygen, steroids, and Alendronate or Etidronate Sodium (which prevents as well as reverses the deposition of crystals in alveoli). [10] Corticosteroids are generally considered to be ineffective. However, only a few authors have reported on the effect of corticosteroids. [3] Role of bilateral lung transplant is said to help to prolong the life expectancy. [5] No recurrence of PAM post lung transplant has been observed yet. Counseling of surgical option was given. After discharge, patient did not turn up. We lost to follow up the patient.

The clinical course is also unpredictable. It can remain stable for many years after the diagnosis without any significant progression. [9] It can progress rapidly leading to death due to respiratory failure in few years. [4] In our case, the progression of disease was rapid and reached to the stage of pulmonary fibrosis (stage IV) just at the age of 20 years. Hence, the disease process is still unpredictable. Close follow up of an adequate number of cases should be studied to have clearer picture. There is still a large scope for prospective studies of a good number of PAM cases to know about the demography, progression and treatment options of PAM since it is very rare disease with unpredictable course and no therapy has proved beneficial including whole lung lavage.

Limitation of our study was inability to proceed with Bronchiolo-alveolar lavage and lung Biopsy since we lost to follow up the patient after she was discharged.

Our research focused on advanced disease and its abrupt progression at a very young age, which is usually seen after 40 years of age. Our case had almost all the imaging findings described in literature till date. Also, hypothesis of extremely high or low temperature as one of the contributing factor in the etiology of PAM was raised, which further prospective studies and literature needs review to prove or disprove.

Disclosures:
Funding / Grants / Competing interest- None.

Acknowledgements:
My technical staff to help me in organizing tests in smoother way.

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