First report of pulmonary alveolar microlithiasis diagnosed by cryobiopsy

Sir,

Pulmonary alveolar microlithiasis (PAM) is characterized by the widespread intra-alveolar accumulation of innumerable minute calculi called microliths. PAM is caused by inactivation of the gene SLC34A2, due to which the alveolar type II cells are unable to clean up the phosphorus ion from the alveolar space resulting in its accumulation forming microliths rich in calcium phosphate. Till date, more than 1000 cases have been reported worldwide and possibly 30 from India.[1-4] We describe herein a case in whom we performed cryobiopsy of the lung to confirm the diagnosis of PAM.

BK, a 24-year-old man, presented with the only complaint of progressive dyspnea on effort for 4 years. He was a smoker but denied any comorbid condition. He had exposure to the dusty environment being a soldier. His general and systemic physical examination was unremarkable. The routine blood investigations including serum calcium, phosphorus, and parathyroid hormone were within the normal range. The high-resolution computed tomography of the chest showed diffuse bilateral distribution of numerous calcific micronodules and circumferential pleural calcification appearing as a pencil-thin sharp dense white linear radiolucency all along the lung periphery [Figure 1]. Pulmonary function tests revealed postbronchodilator forced vital capacity (FVC) – 72%, forced expiratory volume-one second (FEV1) – 76%, FEV1/FVC – 90%, and the diffusion capacity – 65% of their respective predicted values. A video bronchoscopy was done, and transbronchial lung
Cryobiopsy (TBLC) was performed in the right middle and lower lobes to get 3 large specimens of lung tissue, largest measuring 7 mm in the maximum diameter. The histopathology examination confirmed the diagnosis of PAM in this case [Figure 2].

PAM is a rare lung disease which is a classic example of clinical and radiological dissociation. The diagnosis should be considered in an individual who has significant radiographic changes with no or very few symptoms and physical findings as was in this case who had only a mild restrictive dysfunction. This case presented with the typical radiological picture of PAM such as the sandstorm lung sign, circumferential pleural calcification, ground-glass opacities, and crazy paving pattern. The sandstorm lung sign is characterized by bilateral diffuse clearly outlined and bright calcific micronodules, distributed throughout the lungs, although with a greater concentration in the middle and inferior regions.

Chest radiograph and computed tomography scan remain the most important tool for the diagnosis of PAM. Although lung biopsy is the most definitive diagnostic procedure, it is not frequently performed. In a meta-analysis, Castellana et al.1 reported transbronchial biopsy (n = 175; 28.0%), open lung biopsy (n = 159; 25.4%), or autopsy (n = 20; 3.2%) in only limited number of cases of PAM.1 We report here the first case of PAM where diagnosis was confirmed by TBLC.

There is no effective medical treatment available for PAM. The use of disodium etidronate which reduces calcium phosphate precipitation remains controversial.5 Bilateral lung transplant in advanced cases is the only definitive treatment.

We have described here a case of PAM in whom the diagnosis was confirmed by TBLC which could be a safer and viable option to open lung biopsy in diffuse lung diseases.

Figure 1: High-resolution computed tomography chest (a) lung window showing numerous diffusely distributed sand-like calcific micronodules and bilateral interlobular septal thickening, (b) Mediastinal window showing bilateral circumferential pleural calcification

Figure 2: Lung tissue specimens (a) obtained by transbronchial lung cryobiopsy and a specimen (b) measuring 7 mm in maximum diameter. H and E-stained section of lung biopsy (c) low power view (×40) showing numerous microliths within the alveolar spaces and (d) oil immersion (×1000) showing lamellar concretion

Declarations of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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REFERENCES
1. Castellana G, Castellana G, Gentile M, Castellana R, Resta O. Pulmonary alveolar microlithiasis: Review of the 1022 cases reported worldwide. Eur Respir Rev 2015;24:607-20.
2. Kashyap S, Mohapatra PR. Pulmonary alveolar microlithiasis. Lung India
3. Gayathri Devi HJ, Mohan Rao KN, Prathima KM, Das JK. Pulmonary alveolar microlithiasis. Lung India 2011;28:139-41.
4. Malhotra B, Sabharwal R, Singh M, Singh A. Pulmonary alveolar microlithiasis with calcified pleural plaques. Lung India 2010;27:250-2.
5. Borrelli R, Fossi A, Volterrani L, Voltolini L. Right single-lung transplantation for pulmonary alveolar microlithiasis. Eur J Cardiothorac Surg 2014;45:e40.

Sir,

Bronchoalveolar lavage (BAL) is known to be a safe and useful diagnostic procedure to obtain samples from the lower respiratory tract. Rare complications attributable to BAL are hypoxia, aspiration, fever, and bleeding.\[1\] Fiberoptic bronchoscopy (FOB)-induced pneumothorax is reported in 0.1% of cases, whereas pneumothorax after BAL has been reported in seven cases where the biopsy was taken through FOB.\[2,3\] The incidence of bronchopleural fistula (BPF) varies from 4.5% to 20% after pneumonectomy and 0.5% after lobectomy, but BPF after pneumothorax has not been reported.\[4\] We report a rare case of BPF secondary to BAL-induced pneumothorax.

A 65-year-old male known case of carcinoma hypopharynx underwent total laryngectomy, esophagectomy, and tracheostomy under general anesthesia. Preoperative chest X-ray revealed a cavity lesion in the right lower zone with no air–fluid level. Pulmonary function test suggested mild restrictive disease, and echocardiography was normal. After prolonged surgery, the patient was shifted to the surgical intensive care unit (ICU) for postoperative mechanical ventilation. On day 3, the patient was maintaining oxygen saturation (SpO\textsubscript{2}) of 90% on T-piece, but on auscultation, there was reduced air entry in the left side. Suspecting a mucous plug after chest X-ray, a BAL was performed in the left lung with 2 aliquots of 20 mL normal saline using a gentle suction of 50–60 mmHg. Few hours after the procedure, the patient had tachypnea, respiratory distress, and dip in saturation to 85%. Chest radiograph confirmed the presence of a right lung pneumothorax [Figure 1]. Immediately, a 24-F intercostal drain (ICD) tube was placed in the fourth intercostal space. Gush of air was heard and bubbles started appearing in under water drain. Slowly the vitals of the patient improved to SpO\textsubscript{2} of 96%–97%. The patient was maintaining saturation on T-piece of 90%–95% on day 4 and day 5, but underwater seal showed bubbling and chest X-ray showed residual pneumothorax. The patient found difficult to wean from the ventilator showing bubbling in underwater drain with each positive-pressure ventilatory cycle, which raised the differential diagnosis of BPF. A computer tomography scan of the chest revealed residual pneumothorax, pus-filled cavity in the right lower zone, and ICD in situ.

Pneumothorax post BAL in this case could be the sequelae to air trapping or instilled normal saline, which resulted in rupture of an air-filled cavity. BPF during the procedure possibly results from a snugly wedged FOB into the bronchus. To avoid similar cases in future, a high level of vigilance toward procedural safety and a structured...