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Pulmonary Alveolar and Pancreatic Microlithiasis: An Autopsy Case

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Pulmonary Alveolar Microlithiasis (PAM) is an autosomal recessive disorder characterised by a mutation in the Solute Carrier Family 34, Member2 (SLC34A2 gene [1,2]. The disease involves the deposition of calcispherites (calcium phosphate salts) within the alveoli rather than in the interstitium or along the blood vessels, and it occurs without any disturbance of calcium metabolism [3]. It is most commonly observed in young adults in their third or fourth decades of life, with an almost equal gender distribution [4]. Similar types of calcification may also occur in extrapulmonary sites such as the sympathetic ganglia, testes, seminal vesicles, and epididymis [4,5].

Symptoms are usually mild in the early stages, and some patients may remain asymptomatic. The incidence of PAM in India is approximately 0.06 [6]. Herein, authors present an autopsy case demonstrating the presence of calcispherites in both the lungs and pancreas.

A 35-year-old female was brought to the Casualty by her relatives, complaining of abdominal pain and severe dyspnoea. These symptoms had been present for about one week, initially mild but worsening significantly 24 hours before arrival. The patient succumbed to death within a few minutes before hospital admission.

The deceased was non alcoholic and a non smoker. According to her spouse, she had no significant medical history, including respiratory disorders (such as tuberculosis), hypertension, diabetes mellitus, blunt trauma, myocardial infarction, or any other infectious diseases.

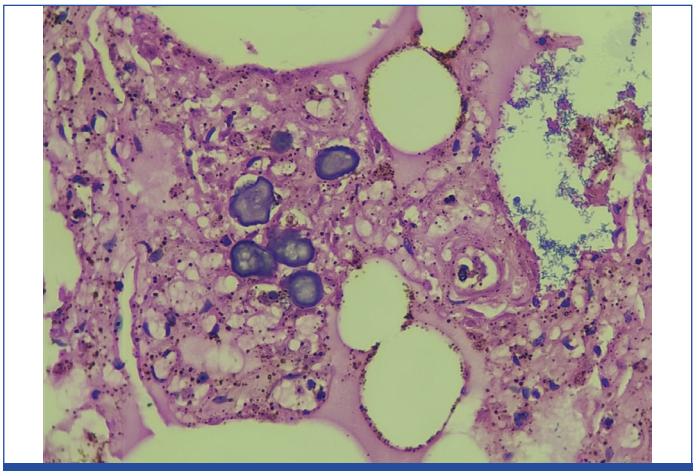
A detailed autopsy revealed abnormalities in both the lungs and pancreas. The pleura was adherent to the chest wall, and pleural effusion was noted. Both lungs appeared congested and oedematous, with patchy areas of consolidation involving both sides [Table/Fig-1]. On cut section, intrapulmonary haemorrhage was observed bilaterally. The pancreas was grossly intact but showed areas of congestion and partial autolysis on section [Table/Fig-1].

[Table/Fig-1]: Gross images of lungs and pancreas microlithiasis.

No other significant findings were noted in the remaining viscera.

As mentioned earlier, the most commonly affected age group is the third to fourth decade of life, with both genders equally involved. In the present case, the patient was 35 years old, consistent with the report published by Sarkar M et al., [5]. Gayathri Devi HJ et al., reported a case involving a 15-year-old boy [3].

Histopathological examination {Haematoxylin and Eosin (H&E)} of the autopsy specimens revealed numerous calcospherites distributed throughout the alveoli [Table/Fig-2]. These deposits were

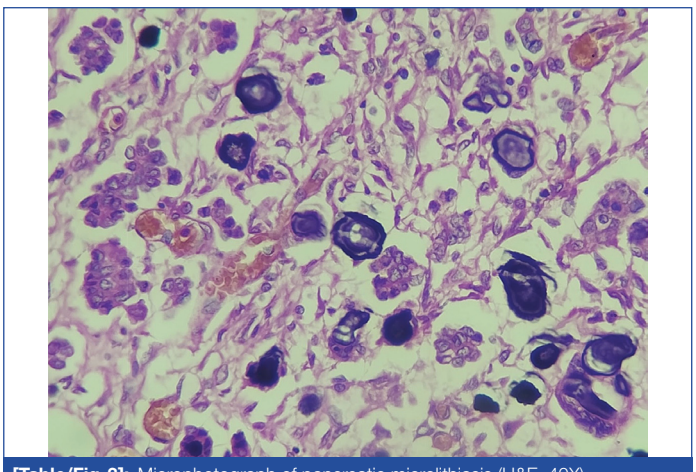


[Table/Fig-2]: Microphotograph of pulmonary alveolar microlithiasis (H&E, 40X).

not confined to the lungs but were also identified in the pancreatic tissue [Table/Fig-3].

The PAM is known to exhibit a significant clinicoradiological dissociation [3]. Plain chest radiographs in such patients typically show the pathognomonic “sandstorm-like” diffuse bilateral calcific micronodules, predominantly involving the middle and lower lobes [1,2,4].

In the present case, the patient’s symptoms were initially mild but later escalated, resulting in severe breathlessness. A chest X-ray could not be performed in the patient, nor had it been done prior



[Table/Fig-3]: Microphotograph of pancreatic microlithiasis (H&E, 40X).

to death. In the case reported by Gayathri Devi HJ et al., the patient was asymptomatic and had no respiratory difficulties, which contrasts with the case described by Sarkar M et al., where the patient had experienced respiratory symptoms for three years, with worsening in the last four months [3,5]. The chest X-ray in that case revealed numerous micronodules in both lungs. The GeneXpert test was positive for *Mycobacterium tuberculosis* complex. A lung biopsy additionally revealed numerous lamellated calcispherites [5], and gene mutation studies confirmed the SLC34A2 mutation. The severe symptoms in that case were therefore attributed to coexisting tuberculosis.

There is limited literature confirming the involvement of the pancreas in PAM, as observed in the present case [5]. Extrapulmonary involvement occurs because the SLC34A2 gene is also expressed in other tissues such as the liver, pancreas, kidneys, and male genitourinary system [5]. Previous studies have suggested a plausible association between acute pancreatitis and acute lung injury [7,8]. In such cases, pancreatitis may be induced by microlithiasis or dysfunction of the sphincter of Oddi, leading to lung injury through inflammatory cytokine-mediated mechanisms [9,10].

In the present case, abdominal pain could be attributed to pancreatitis, possibly secondary to microlithiasis in the pancreatic parenchyma. The cause of death was respiratory failure resulting from PAM.

The differential diagnosis to be considered include miliary tuberculosis, silicosis, amyloidosis, metastatic calcification due to hyperparathyroidism, and end-stage renal disease. It is noteworthy that the cases reported by Gayathri Devi HJ et al., and Sarkar M et al., did not demonstrate extrapulmonary involvement [3,5].

In the present case, there was no evidence of granulomatous inflammation or amyloid deposition, as confirmed by negative Ziehl-Neelsen and Congo red stains, respectively. There was also no history suggestive of renal dysfunction, and histopathological examination of the remaining viscera did not reveal any significant findings.

The definitive treatment for PAM is lung transplantation. Medical management includes bisphosphonates, inhaled and systemic corticosteroids, and a low-phosphate diet [3].

It was not possible to confirm the diagnosis through molecular studies for the SLC34A2 gene. However, available literature indicates that genetic testing is not mandatory for diagnosis [5]. Given the autosomal recessive inheritance pattern, an open lung biopsy can be performed when molecular testing is not feasible, enabling early diagnosis and potentially preventing catastrophic events in siblings.

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