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Precalcific phase of pulmonary alveolar microlithiasis

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DESCRIPTION

A teenage boy, with a history of bronchial asthma since early childhood, was referred to our respiratory service with an incidental finding of diffuse micronodular opacities on chest imaging ordered following a road traffic accident (figure 1A–D).

His history was negative for respiratory, systemic and connective tissue disease symptoms, and was unremarkable for exposure to smoke, debris and animals. There was no previous COVID-19 infection and his only medication was a salbutamol inhaler as needed for mild intermittent bronchial asthma. His parents are first cousins with a negative family history of respiratory disease.

A chest X-ray was done 9 years prior to presentation for a suspected lung infection that revealed the same abnormality (figure 1E). The initial differential diagnosis included hypersensitivity pneumonitis, respiratory bronchiolitis, pulmonary haemosiderosis and infectious process. Pulmonary function tests (spirometry, lung volumes and diffusion capacity) and laboratory investigations including workup for connective tissue disease were within normal limits.

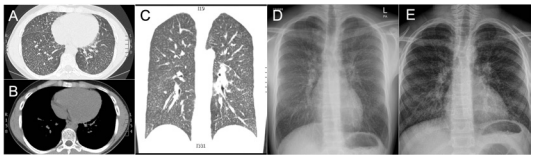

Figure 1 (A and C) High-resolution CT shows extensive bilateral poorly defined centrilobular ground-glass nodules. (B) No calcification is evident on the soft tissue window. (D) Chest X-Ray showing diffuse bilateral pulmonary nodules. (E) An old chest X-ray taken 9 years prior to presentation does not demonstrate appreciable changes over the years.

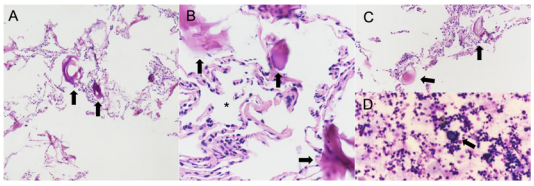

Figure 2 (A) Low-power magnification of alveolar spaces with calcification inside some of them (arrows). (B) Medium-power magnification of the lung parenchyma showing intra-alveolar concentric calcification (arrows) in contrast to the empty alveolar space (asterisk). (C) High-power magnification showing centrally lamellated calcified bodies (arrows). (D) Bronchoalveolar lavage showing numerous macrophages and lymphocytes with a calcific body (arrow) (Giemsa stain 200× magnification).

Learning points

- Pulmonary alveolar microlithiasis (PAM) typically presents radiologically with diffuse calcified pulmonary nodules giving rise to the characteristic sandstorm appearance.
- Early in the course of the disease, PAM may be unsuspected due to the non-calcified pulmonary nodules in chest imaging.
- Demonstration of microliths from lung biopsies or bronchoalveolar lavage can establish the diagnosis of PAM.

A bronchoscopy with bronchoalveolar lavage (BAL) and transbronchial lung biopsy was performed. BAL cell count and microbiology were unremarkable. BAL cytology and transbronchial lung biopsies were diagnostic for pulmonary alveolar microlithiasis, showing calcified bodies and intra-alveolar microliths (figure 2).

Pulmonary alveolar microlithiasis (PAM) is a rare autosomal recessive disorder involving the SLC34A2 gene which encodes type IIb sodium phosphate cotransporter. This gene is expressed in the lungs by alveolar type II cells and its dysfunction may reduce the clearance of phosphate from the alveolar space leading to widespread formation of intra-alveolar calculi called microliths.[1]

PAM is typically diagnosed during the second and third decades of life.[2] Clinical presentation is variable, and cases are usually discovered incidentally by chest imaging. Radiological classification of severity comprises four phases. In the first phase, the precalcific/minimally calcified phase, microliths are few and the calcification grade is low, making it difficult to recognise. Only three authors have reported the discovery of cases in this phase, all in asymptomatic children.[2] Our patient presented in this phase with imaging showing widespread non-calcified centrilobular pulmonary nodules. Subsequent phases are easier to diagnose due to characteristic radiological findings with an increase in the size and grade of calcification of the pulmonary nodules and involvement of the lung interstitium and pleura.[2]

Definitive diagnosis of PAM can be made by histopathological demonstration of microliths from BAL or a lung biopsy (transbronchial or surgical). Microliths are composed of concentric layers of hydroxyapatite crystals surrounding an amorphous nucleus.[3] This disease is progressive in nature and there is still no approved medical treatment which may alter its course. Multiple treatments were studied for PAM, including glucocorticoids, calcium-binding agents and whole lung

lavage, and proven to be ineffective. Lung transplant, which is reserved for advanced cases, is the only effective treatment to date.[3]

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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REFERENCES

1 Huqun IS, Izumi S, Miyazawa H, Ishii K, *et al*. Mutations in the SLC34A2 gene are associated with pulmonary alveolar microlithiasis. *Am J Respir Crit Care Med* 2007;175:263–8.
2 Castellana G, Castellana G, Gentile M, *et al*. Pulmonary alveolar microlithiasis: review of the 1022 cases reported worldwide. *Eur Respir Rev* 2015;24:607–20.
3 Bendstrup E, Jönsson Åsa Lina M, . Pulmonary alveolar microlithiasis: no longer in the stone age. *ERJ Open Res* 2020;6:00289-2020–20.