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Rare case of pulmonary sarcoidosis with cystic bronchiectasis

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Accepted 14 July 2020

DESCRIPTION

A previously well 27-year-old man presented with a 5-day history of photophobia. Chest high-resolution CT (HRCT) scan on admission showed bilateral hilar lymph node enlargement and nodules with irregular boundaries, encircled by a rim of numerous tiny satellite nodules (sarcoid galaxy sign) predominantly in the bilateral upper lobes (figure 1A). He was diagnosed as systemic sarcoidosis associated with uveitis and lung involvement. Therefore, he was received 1g/day of intravenous methylprednisolone for 3 days, followed by oral prednisolone (PSL) at 40 mg/day. After these initial corticosteroid treatments, the chest CT abnormalities immediately resolved with improvements of the photophobia. However, he stopped taking oral PSL on his own judgement. After 3 years, he had a dry cough, dyspnoea on exertion and general fatigue. Chest HRCT revealed marked bronchiectasis,

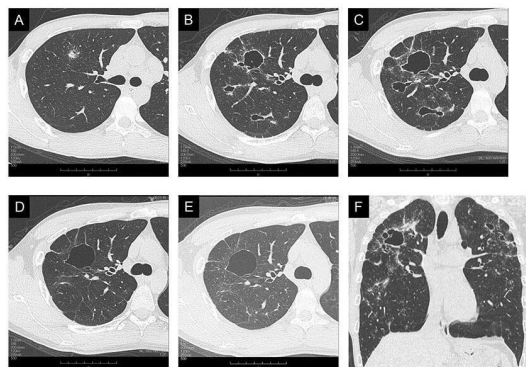


Figure 1 Serial changes in chest high-resolution CT (HRCT) images. (A) On initial visit, chest HRCT revealed bilateral hilar lymph node enlargement and nodules with irregular boundaries, encircled by a rim of numerous tiny satellite nodules (sarcoid galaxy sign) predominantly in the bilateral upper lobes (B) following 3 years, chest HRCT showed marked bronchiectasis, peripheral enlarged thick-walled cysts, adjacent multiple small nodules occurring in the peribronchovascular regions. (C) Following another 1 year, images of chest HRCT showed further deterioration of cystic bronchiectasis and multiple small nodules. (D) At 4 months after oral corticosteroid therapy, thick-walled cystic bronchiectasis and multiple small nodules in both lung fields markedly improved. (E) At 25 months after oral corticosteroid therapy, a part of thin-walled cystic bronchiectasis remained unchanged. (F) Coronal images of chest CT at immediately before administration of oral corticosteroid therapy around the same time (C). Note that cystic bronchiectasis were gradually extended and multiple small nodules deteriorated in the upper and middle zones predominance.

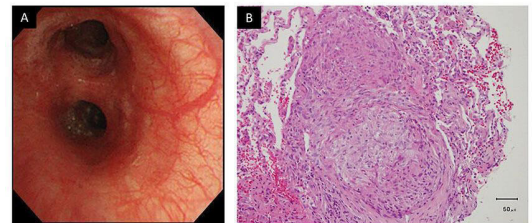


Figure 2 (A) Bronchoscopy findings were consistent with vascular network vessels at airway lumens. (B) Photomicrograph of transbronchial lung biopsy specimen demonstrated a non-caseating epithelioid cell granuloma in the alveoli and pleura (H&E stain) (scale bar=50 µm).

peripheral enlarged thick-walled cysts, adjacent multiple small nodules occurring in the peribronchovascular regions (figure 1B). The laboratory data revealed normal value of serum angiotensin-converting enzyme (20.5 U/L) and increased lysozyme (12.2 µg/mL). The pulmonary function test showed a normal range of spirometry with normal diffusion capacity. Bronchoscopy findings were consistent with vascular network vessels at airway lumens (figure 2A). Examination of bronchoalveolar lavage fluid revealed a normal percentage of lymphocytes of 8% and an increased CD4/CD8 ratio of 3.8. Photomicrograph of transbronchial lung biopsy specimen demonstrated non-caseating epithelioid cell granulomas in the alveoli and pleura, confirming the diagnosis of pulmonary sarcoidosis (figure 2B). After another 1 year, images of chest HRCT showed further deterioration (figure 1C,F). In addition, the value of forced expiratory volume in 1 s decreased 4.34–3.91 L. Eventually, he was treated with oral PSL at 30 mg/day. His clinical condition and chest imaging abnormalities markedly improved at 4 months after PSL therapy (figure 1D). In particular, some cystic bronchiectasis disappeared. Subsequently, the dose of PSL was gradually reduced to 5 mg/day over 12 months. Although no exacerbation has been observed in the



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To cite: Sugino K, Kurosaki A, Homma S, *et al.* *BMJ Case Rep* 2020;**13**:e237863. doi:10.1136/bcr-2020-237863

Learning points

- ▶ Cystic bronchiectasis are rare in non-fibrotic pulmonary sarcoidosis.
- ▶ The check-valve mechanism due to stenosis of bronchi with peribronchial fibrosis or accumulation of granulomas may result in peripheral cyst formation in pulmonary sarcoidosis.

subsequent 25 months, a part of thin-walled cystic bronchiectasis remains unchanged (figure 1E).

Patients with extensive fibrotic pulmonary sarcoidosis often can develop honeycomb-like pattern or clustered cysts.^{1 2} Recently, Sawahata *et al* speculated that honeycomb-like pattern may result from traction bronchiectasis in patients with fibrotic pulmonary sarcoidosis.^{3 4} However, in this case, there has been seen not extensive fibrosis but numerous small nodules with cystic bronchiectasis. The check-valve mechanism due to stenosis of bronchi with peribronchial fibrosis or accumulation of granulomas may result in peripheral cyst formation. In fact, these cysts were connected with distal bronchiectasis and resolved with PSL therapy. On the other hand, anti-granulomatous therapy may prevent a possible sarcoidosis antigen from being cleared, resulting in relapse when the anti-granulomatous therapy is withdrawn.⁵ Indeed, Gottlieb *et al* reported that patients with sarcoidosis receiving corticosteroid therapy had a higher rate of relapse than those who are observed without treatments.⁶ Therefore, we speculate that cystic bronchiectasis formation might not develop over time, if this patient did not have treatment with a high dose of corticosteroid at the initial visit.

Contributors All the authors have read the manuscript, had acknowledging responsibility for the work and approved this submission. KS performed patient data collection, KS and AK involved in data analysis and KS, AK, SH and KK contributed in manuscript preparation and review.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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REFERENCES

- 1 Hennebicque A-S, Nunes H, Brillet P-Y, *et al*. Ct findings in severe thoracic sarcoidosis. *Eur Radiol* 2005;15:23–30.
- 2 Jeong YJ, Lee KS, Chung MP, *et al*. Chronic hypersensitivity pneumonitis and pulmonary sarcoidosis: differentiation from usual interstitial pneumonia using high-resolution computed tomography. *Semin Ultrasound CT MR* 2014;35:47–58.
- 3 Sawahata M, Shijubo N, Johkoh T, *et al*. Honeycomb lung-like structures resulting from clustering of traction bronchiectasis distally in sarcoidosis. *Respirol Case Rep* 2020;8:e00539.
- 4 Sawahata M, Johkoh T, Kawanobe T, *et al*. Computed tomography images of fibrotic pulmonary sarcoidosis leading to chronic respiratory failure. *J Clin Med* 2020;9:142.
- 5 Judson MA. Corticosteroids in sarcoidosis. *Rheum Dis Clin N Am* 2016;42:119–35.
- 6 Gottlieb JE, Israel HL, Steiner RM, *et al*. Outcome in sarcoidosis. The relationship of relapse to corticosteroid therapy. *Chest* 1997;111:623–31.

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