A Rare Cause of Undiagnosed Exudative Pleural Effusion

A Case Report

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There are various causes of exudative pleural effusions with up to 20% remain undiagnosed despite a diagnostic workup. IgG-4-related disease is a rare fibroinflammatory disorder that can affect various organs and can be the cause of exudative pleural effusion. We reported a case of IgG4-related pleuritis without other systemic manifestations.

CASE REPORT

A 50-year-old man with a history of giant cell tumor of the left distal femur without evidence of distant metastasis 10 years ago. In 2018, he presented with asymptomatic right pleural effusion. The pleural fluid was a turbid straw color with lymphocytic predominance. Pleural LDH was 2,594 U/L, protein 9.5 g/dL, glucose <4 mg/dL, cholesterol 144 mg/dL, and triglyceride 18 mg/dL. The microbiological studies were all negative. **Right Pleuroscopy revealed diffuse pleural thickening with yellowish plaques (Figure 1). The histopathology showed xanthogranulomatous pleuritis and pleural fibrosis.** Computed tomography (CT) of the chest was performed later which showed consolidation and round atelectasis of the right lower lobe with loculated right pleural effusion **(Figure 2).** Transbronchial lung biopsy was done with a non-diagnostic result. There was no specific treatment and the cause of pleuritis cannot be identified. The patient had no respiratory symptoms with stable minimal right pleural effusion during a 2-year follow-up. Two years later, he developed a new left pleural effusion without systemic symptoms. CT chest revealed no change of loculated right pleural effusion and round atelectasis of the right lower lobe **(Figure 2). The profile of the left pleural fluid and pleuroscopic findings were similar to the previous examination on the right side (Figure 1).** The histopathological results showed dense infiltration by plasma cells with occasional eosinophils and multifocal fibrotic changes. **The immunostaining showed polyclonal plasma cells with CD138+, Kappa to Lambda 1:1, predominance of IgG over IgA and IgM with IgG4 80-90% of total IgG that was compatible with IgG-4-related disease (Figure 3).**

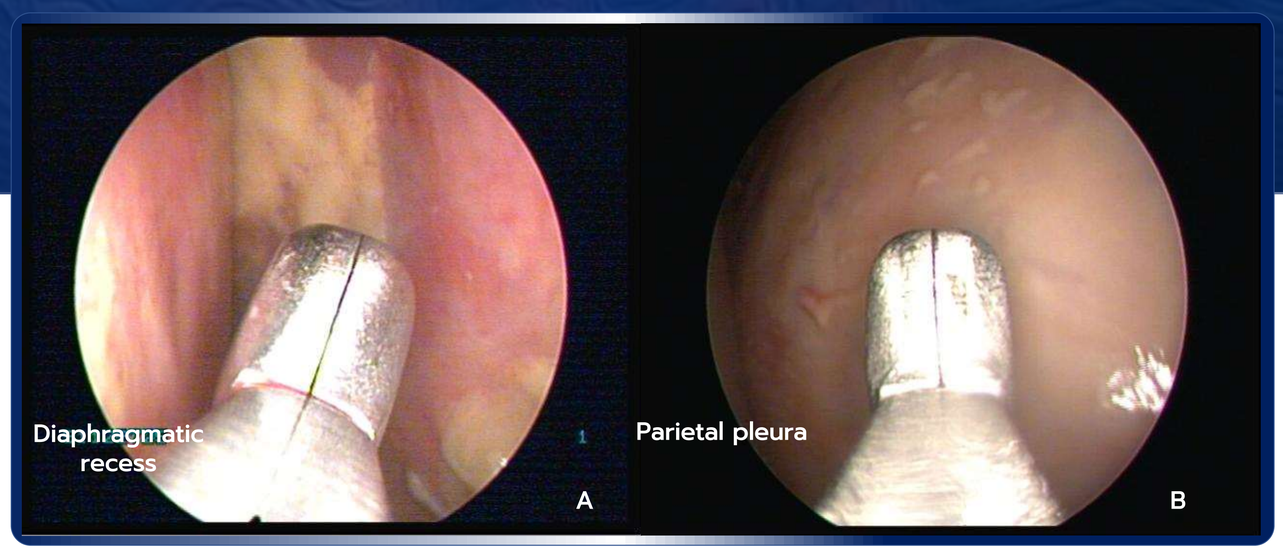

Figure 1: A. Right Pleuroscopy revealed diffuse pleural thickening with yellowish plaques, B. Left Pleuroscopy revealed diffuse pleural thickening with yellowish plaques.

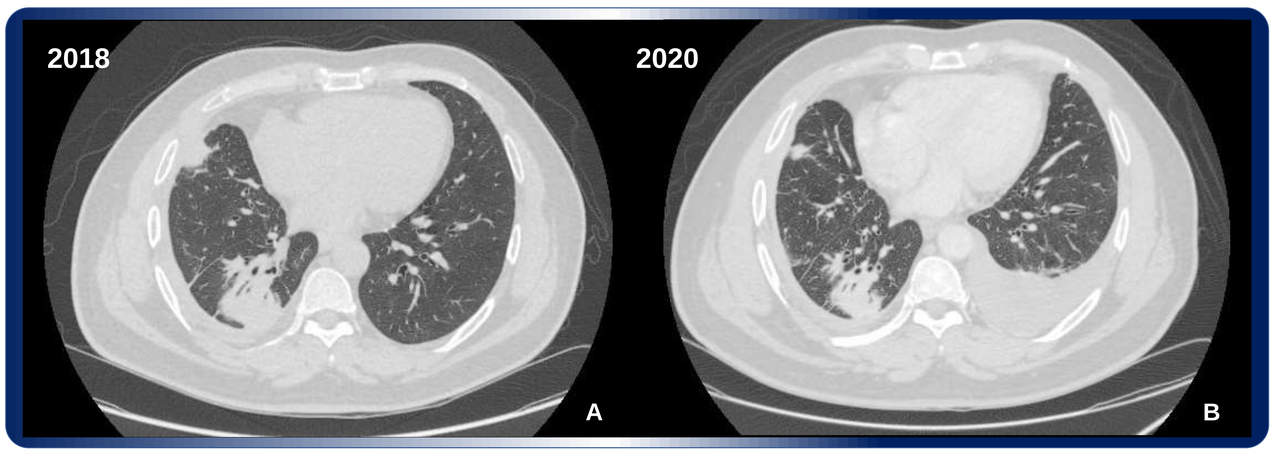

Figure 2: A. Chest CT showed consolidation and round atelectasis of right lower lobe with loculated right pleural effusion, B. Chest CT showed no changed of consolidation and round atelectasis of right lower lobe with new left pleural effusion.

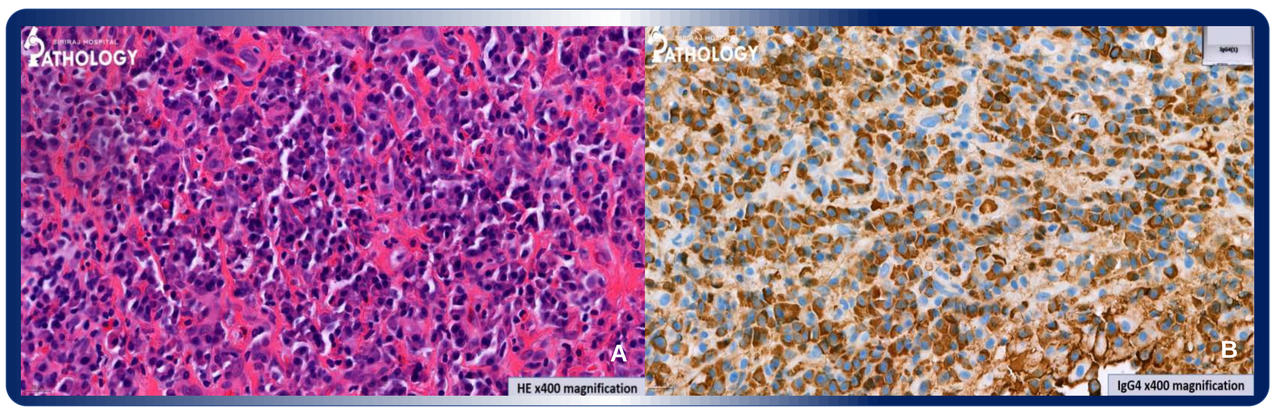

Figure 3: A. H&E Staining showed dense infiltration by plasma cells with eosinophils And multifocal fibrotic changes. B. The immunostaining showed polyclonal plasma cells with IgG4 80-90% of total IgG (Courtesy of Assist. Prof. Panthep Suttinont and Assist. Prof. Ruchira Ruangchira-urai Department of Pathology, Faculty of Medicine Siriraj Hospital, Mahidol University).

DISCUSSION

Thoracic involvement can be seen in approximately 40% of IgG4-RD patients[1]. Pleural manifestation of IgG4-RD include pleural effusion, pleuritis with fibrosis, and pleural mass that can be found in 4.6-16.1% of patients[1,2]. Although it is frequently presented with other organ involvement such as pancreatitis and sialadenitis[1]. IgG4-RD limited to pleura are extremely rare[1,2]. Glucocorticoids are recognized as first-line treatment and shown to improve pleural fluid and symptoms in most reported cases of pleural involvement[1,2].

Conclusion: The exudative pleural effusion with lymphocytic predominance with high levels of LDH and protein and low glucose without other explainable cause, the IgG-4-related disease should be considered as a differential diagnosis.

[1] Murata Y, Aoe K, Mimura Y. Pleural effusion related to IgG4. Curr Opin Pulm Med. 2019 Jul;25(4):384-390.
[2] Campbell SN, Rubio E, Loschner AL. Clinical review of pulmonary manifestations of IgG4-related disease. Ann Am Thorac Soc. 2014 Nov;11(9):1466-75.

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