



Nephrobronchial fistula: a diagnostic challenge in a patient with IgG4-related disease

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TO THE EDITOR:

IgG4-related disease is an immune-mediated condition characterized by the infiltration of several organs, including the lungs, pancreas, bile ducts, kidneys, thyroid, and central nervous system. One of the main features of the disease is the abundance of IgG4 in the affected tissues, which initially present with an inflammatory phase and progress to subsequent local fibrosis.⁽¹⁾ Thoracic involvement and significant symptoms may occur in 10% of cases, with no specific signs and symptoms. Several changes can be found on chest CT, such as nodules, ground-glass opacities, and peribronchovascular infiltrate.⁽²⁾

The kidneys can be affected in up to 20% of cases, and tubulointerstitial nephritis is the most common manifestation.⁽³⁾ Nephrobronchial fistulas are rare complications of kidney disease, occurring mainly in

association with perinephric abscesses after infectious episodes.⁽⁴⁾ We describe the case of a nephrobronchial fistula in a patient with IgG4-related disease. Free and informed consent was obtained from the patient.

A 51-year-old female presented with a 1-year history right thoracic pain and hemoptysis. Chest CT revealed a nephrobronchial fistula in the thoracoabdominal transition with migrated calculi in the right inferior lobe, associated with a 2.3-cm discontinuity defect in the right diaphragm and atrophic right kidney, along with diffuse urothelial thickening of the ureter (Figures 1A-1C). During investigation, a transbronchial biopsy showed a lymphoplasmacytic inflammatory reaction with a predominance of IgG4. After multidisciplinary discussion, lung segmentectomy, right nephrectomy, and partial hepatectomy were performed to confirm diagnosis and

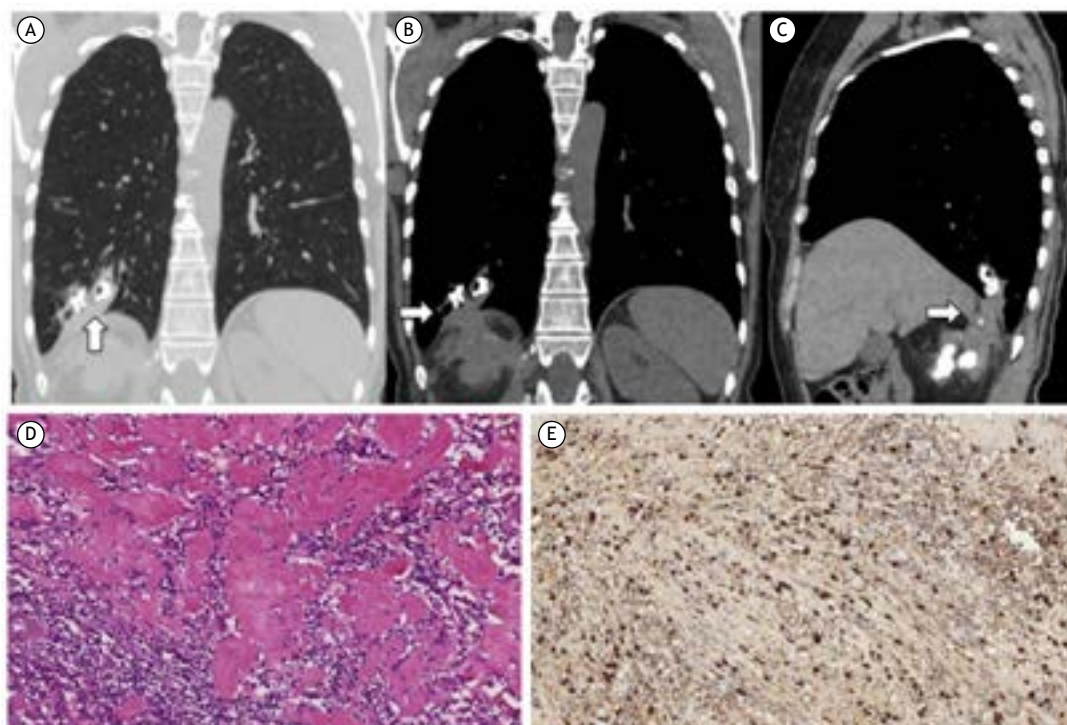


Figure 1. In A, a CT scan showing migrated calculi in the inferior right lobe (arrow). In B and C, coronal and sagittal reconstructions of CT scans showing diaphragmatic discontinuity and a nephrobronchial fistula with nephrolithiasis originated from the right kidney (arrows). Histopathological images showing the nephrobronchial fistula (H&E) with an intense area of fibrosis (in D) and positive IgG4 cells (in E).

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treat symptoms. Local excision of the fistula revealed intense lymphoplasmacytic infiltration, fibrosis, and vascular proliferation, which were present in the renal parenchyma as well. Specific IgG4 immunostaining showed > 100 positive cells per high-power field (Figures 1D and 1E). Nephrolithiasis was found in the fistulous tract with no signs of obstructive nephropathy. Autoantibodies were negative, serum IgG levels were unremarkable, and PET-CT scanning excluded signs of systemic activity.

A nephrobronchial fistula results from kidney inflammation that progresses to the respiratory system, most cases being mainly reported because of pyelonephritis.⁽⁴⁻⁶⁾ We report the first case of a nephrobronchial fistula with calculi in a patient with histopathological results suggesting IgG4-related disease. The pathophysiology is uncertain, and we believe that local IgG4 inflammation could lead to the development of posterior fistulas in adjacent organs

such as the diaphragm and kidneys, although there are no other systemic manifestations related to the disease.⁽⁷⁾ After surgical treatment of the patient, symptoms improved with no signs of disease activity during follow-up. No additional treatment was necessary.

AUTHOR CONTRIBUTIONS

GCMP, GPB, and ECTN: data acquisition, analysis, and interpretation; drafting and revision of the manuscript. RCC, BGB, and FEA: study design; data analysis and interpretation; and revision of the manuscript. RAK: study design; data analysis and interpretation; drafting and revision of the manuscript. All authors read and approved the final version of the manuscript.

CONFLICTS OF INTEREST

None declared.

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