

Unveiling the Link: IgG4-Related Disease (IgG4-RD) as the Unifying Diagnosis for Autoimmune Pancreatitis, Obstructive Uropathy, and Recurrent Pericardial Effusion

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Introduction:

IgG4-related (IgG4-RD) disease is an autoimmune disorder marked by IgG4-positive plasma cell infiltration, fibrosis, and obliterative phlebitis, commonly affecting the pancreas, salivary glands, and retroperitoneum, with less frequent involvement of the heart, kidneys, and lungs. This case highlights a rare IgG4-RD presentation with **recurrent pericardial effusion**, preceded by **pancreatitis** and **obstructive uropathy**.

Case presentation:

A 62-year-old Malay female with poorly controlled diabetes, hypertension, and chronic kidney disease presented with **obstructive uropathy** requiring ureteric stenting. Imaging revealed pancreatic head calcifications and dilated pancreatic ducts, suggestive of **autoimmune pancreatitis**. An incidental **pericardial effusion** was noted, which recurred despite prior pericardiocentesis. She underwent video-assisted thoracoscopic surgery (VATS) with creation of a pericardial window. Histopathology confirmed IgG4-related disease, showing dense **lymphoplasmacytic infiltrates**, **storiform fibrosis**, and **numerous IgG4-positive plasma cells (Figure 1)**. Immunological workup revealed **positive ANA**, **monoclonal gammopathy**, and **elevated serum IgG**. Pericardial fluid analysis showed **lymphocyte** and **plasma cell** predominance (**Figure 2**), supporting the diagnosis. The patient was treated with **corticosteroids** and **methotrexate**, leading to resolution of the effusion on serial echocardiograms and improved renal function over 6 months.

Conclusion:

This case demonstrates the diagnostic complexity of IgG4-RD and emphasizes the value of a multidisciplinary approach. Multisystem involvement allowed for early diagnosis and effective immunosuppressive therapy, resulting in a favorable outcome.

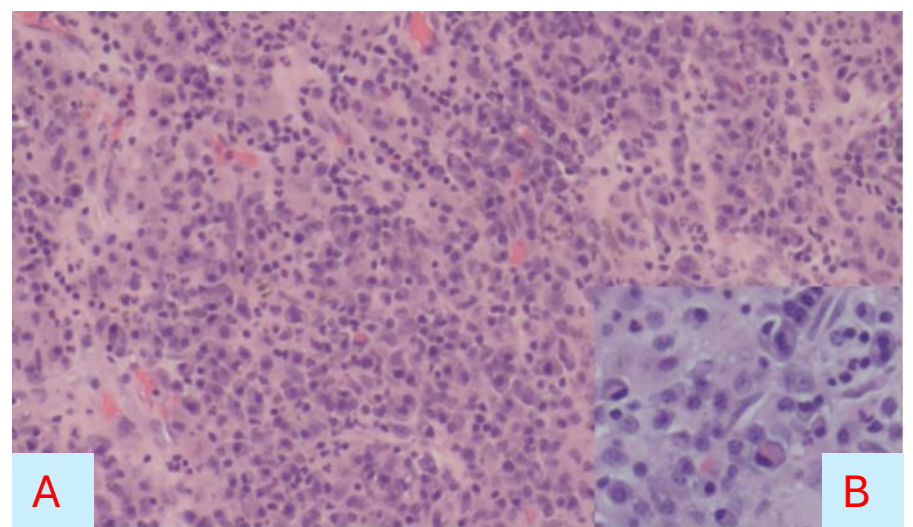


Figure 1: (A) shows the tissue that is densely infiltrated by abundant plasma cells. The inset **(B)** shows binucleated plasma cells and one with pinkish immunoglobulin in the cytoplasm. [Hematoxylin & Eosin stain, original magnification (A) x10, (B) x 20]

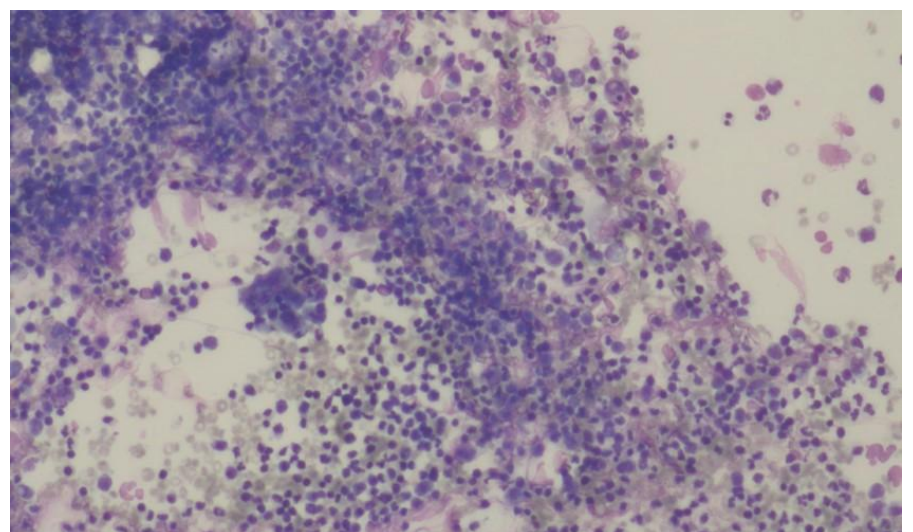


Figure 2: Pericardial fluid FEME shows abundant mixed inflammatory cells, predominantly plasma cells. [May-Grunwald Giemsa stain, original magnification x10]