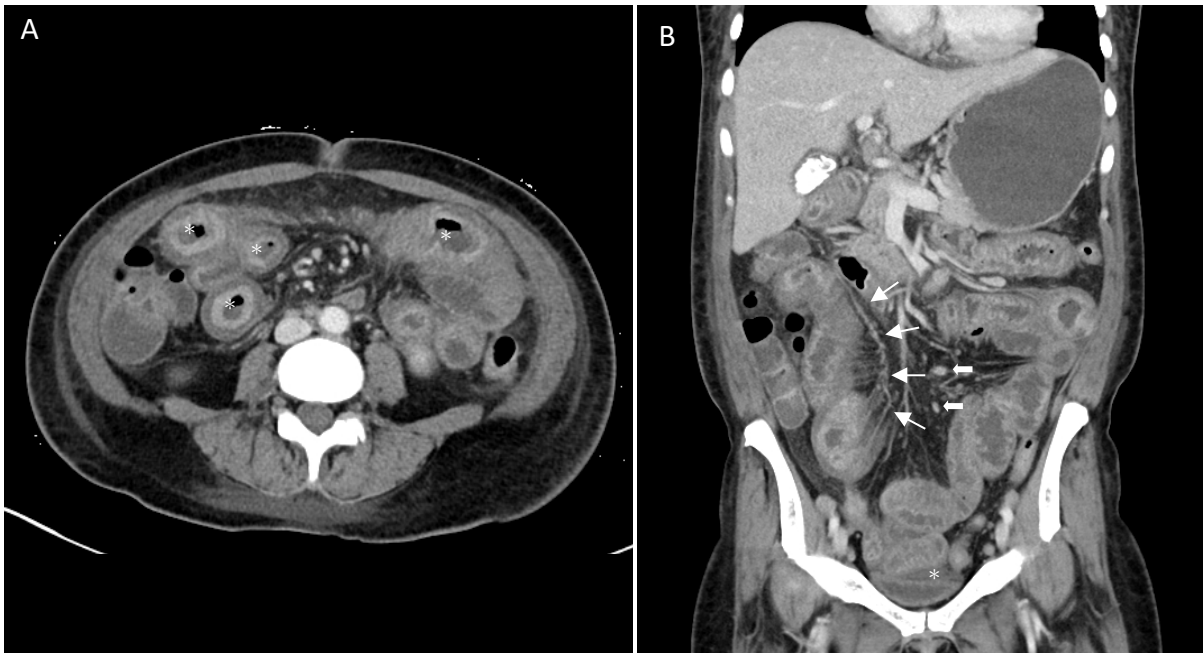


影像競圖獎

Clinical Images:

Lupus Mesenteric Vasculitis: CT Imaging Revealing Diffuse Small Bowel Wall Thickening, Lymphadenopathy, and Ascites



A 52-year-old woman was presented with abdominal pain and bloody stool for one day. Additionally, she had experienced poor appetite, unintentional weight loss and arthralgia for the past year. She had no known past medical history. Laboratory tests revealed leukocytosis, anemia and elevated inflammatory markers. The initial abdominal computed tomography (CT) demonstrated edematous changes in the proximal jejunum. A duodenal ulcer was noticed by a panendoscopy and no obvious lesion was noticed by colonoscopy. The patient was discharged after a seven-day course of antibiotic treatment.

However, she developed severe abdominal tenderness the day after discharge and subsequently returned to the emergency department. A follow-up abdominal CT revealed diffuse circumferential wall thickening with submucosal edema extending from the gastric body to the ileum (Panel A, asterisk), accompanying with hypervascularity of the mesentery (Panel B, thin arrow). Furthermore, ascites (Panel B, asterisk) and multiple lymphadenopathy (Panel B, thick arrow) were observed. Further evaluation showed a positive anti-nuclear antibody (ANA) test with a 1:320 homogeneous pattern, hypocomplementemia, proteinuria and a positive direct Coomb's test.

A diagnosis of lupus mesenteric vasculitis was made. The patient was treated with intravenous methylprednisolone pulse therapy. She is currently receiving monthly cyclophosphamide therapy.

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A Challenging Case of Multicentric Lymphadenopathy: Idiopathic Multicentric Castleman Disease Mimicking IgG4-Related Disease

We present a 56-year-old man with a three-year history of systemic symptoms and generalized lymphadenopathy, ultimately diagnosed with idiopathic multicentric Castleman disease—not otherwise specified (iMCD-NOS), following an initial misdiagnosis of Immunoglobulin G4-related disease (IgG4-RD). He initially presented with submandibular lymphadenopathy, fever, night sweats, and weight loss. Laboratory data revealed polyclonal hypergammaglobulinemia and an elevated serum IgG4 to IgG ratio. Lymph node biopsy revealed dense plasma cell infiltration, abundant IgG4-positive plasma cells, and an elevated IgG4/IgG ratio. Given overlapping histopathologic features, a definitive diagnosis was not possible, and IgG4-RD-directed therapy was initiated with limited response. Despite rituximab treatment, inflammatory markers remained elevated.

Follow-up evaluation revealed persistent hypergammaglobulinemia, elevated interleukin-6 and C-reactive protein (CRP) levels, thrombocytosis, and widespread hypermetabolic lymphadenopathy on positron emission tomography/computed tomography (PET/CT). A repeat biopsy showed sheets of polytypic plasma cells, a lower IgG4/IgG ratio, and absence of defining features of IgG4-RD. A final diagnosis of iMCD-NOS, plasmacytic variant, was made based on international consensus criteria. This case may represent the idiopathic plasmacytic lymphadenopathy (IPL)-type of iMCD—a proposed but unvalidated subtype increasingly recognized in recent literature.

Figure 1 FDG PET/CT Findings in iMCD

Fluorodeoxyglucose (FDG) PET/CT demonstrates diffusely hypermetabolic lymphadenopathy involving cervical, supraclavicular, axillary, mediastinal, gastrohepatic, retroperitoneal, iliac, and inguinal regions. These findings are consistent with a systemic lymphoproliferative disorder, as seen in idiopathic multicentric Castleman disease (iMCD).

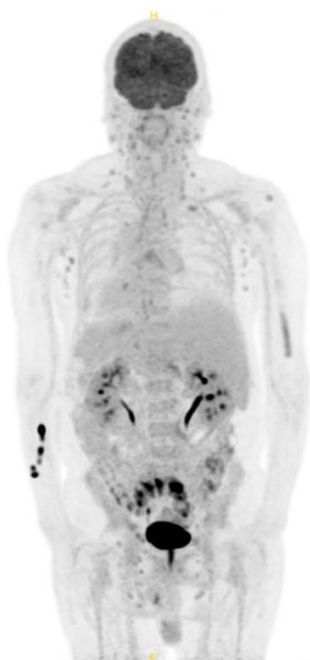
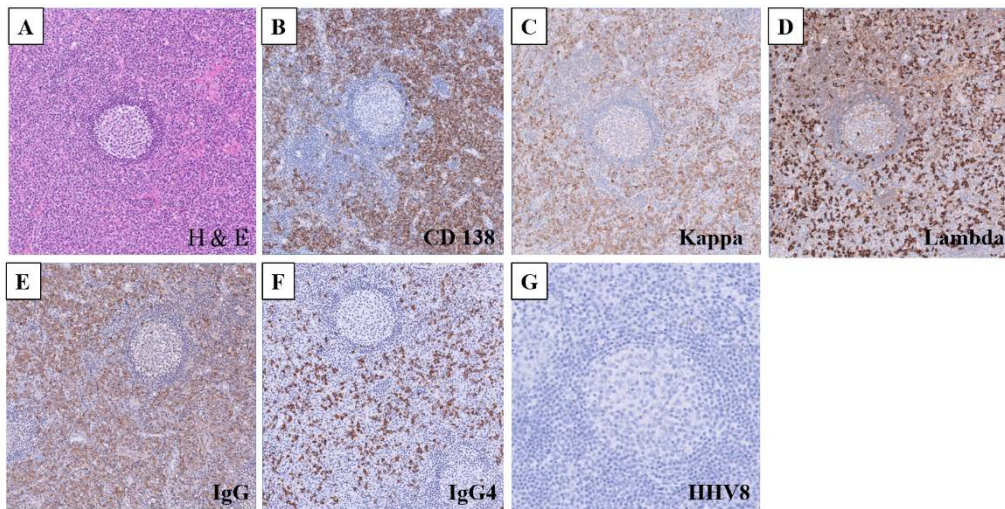


Figure 2 Histopathological and Immunohistochemical Features of Lymph Node Biopsy in iMCD

(A) Hematoxylin and eosin (H&E) staining shows sheets of grade 3 polyclonal plasma cell proliferation without regressed germinal centers, prominent follicular dendritic cells (FDCs), or increased vascularity. (10x, H&E)
(B–D) Immunohistochemistry(IHC) highlights dense CD138-positive plasma cells with a Kappa/Lambda ratio of approximately 2:1, supporting polyclonal proliferation. (10x, IHC)
(E–F) IgG4 immunostaining reveals up to 200 IgG4-positive plasma cells per high-power field (HPF) and an IgG4/IgG-positive cell ratio of 20%. (10x, IHC)
(G) Human herpesvirus 8 (HHV-8) staining is negative. (20x, IHC)



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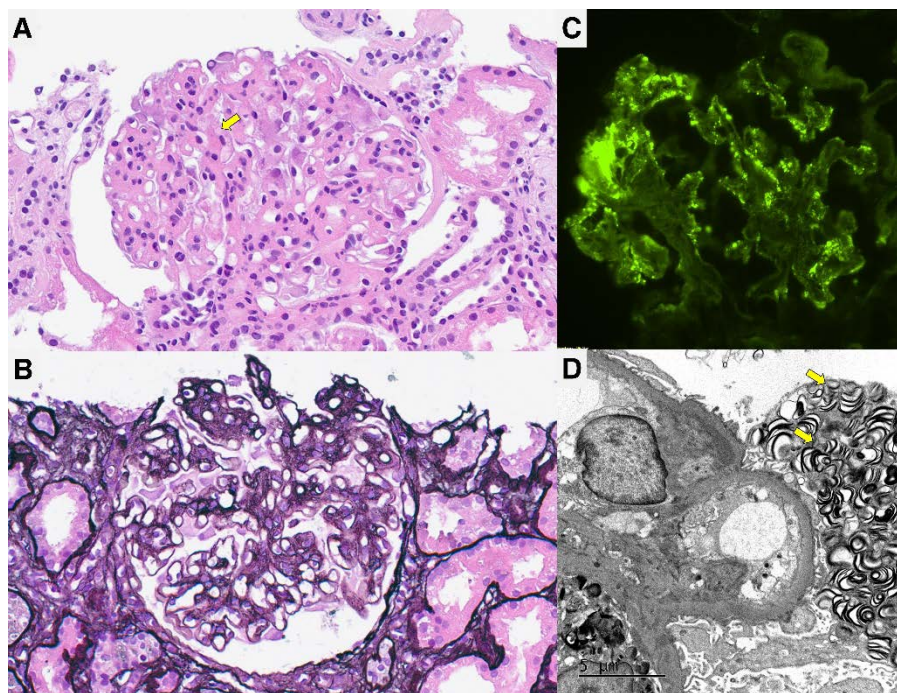
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Clinical Images: Coexisting Fabry Disease in a 47-Year-Old Woman with Systemic Lupus Erythematosus



A 47-year-old woman with a long-standing history of systemic lupus erythematosus (SLE), diagnosed at age 26, presented with cutaneous manifestations and lupus nephritis. She had been treated with mycophenolate mofetil (MMF) at a dose of two tablets twice daily, hydroxychloroquine (HCQ), and oral glucocorticoids. In February 2022, she developed progressive proteinuria. Ciclosporin was initiated but subsequently discontinued at the patient's request.

During follow-up, her proteinuria worsened, and renal biopsy was done in January 2025. Immunofluorescent revealed focal proliferative lupus nephritis (ISN/RPS Class III) with a modified NIH activity/chronicity index of 1/4, accompanied by immune complex deposition (**Figure 1A-1C**). Additionally, electron microscopy revealed myelin body accumulation in podocytes, mesangial cells, endothelial cells, parietal epithelial cells, and tubular epithelial cells (**Figure 1D**). These myelin body findings are suggestive of Fabry disease but may also be seen in certain drug toxicities (e.g., chloroquine). She was referred to a pediatric genetic specialist. Genetic analysis revealed a heterozygous pathogenic variant in the GLA gene: c.1208T>G, p.(Leu403Ter), confirming a diagnosis of Fabry disease. She was diagnosed with Fabry disease coexisting with SLE. For her lupus nephritis, HCQ was discontinued, and MMF was titrated to four tablets twice daily.

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