

SCLEROSING MESENTERITIS – IgG4 RELATED MESENTERIC PSEUDOTUMOR or IDIOPATHIC SCLEROSING MESENTERITIS?

Jovan Popovich MD, Anusha Sunkara MD, Ramona Mihi MD, Jae Ro MD/PHD, Sandra Sessoms MD

Houston Methodist Hospital, Houston, TX

Background:

IgG4 related disease (IgG4-RD) represents an autoimmune fibro-inflammatory condition that can affect multiple organs and can present itself in multiple different forms. We here describe a case of a patient with a mesenteric abdominal mass with histological aspect of IgG4-RD.

Our patient was a 23-year-old African American female presenting with progressively worsening right lower quadrant abdominal pain radiating to her back, of 2 weeks duration. She exhibited significant right lower quadrant tenderness on examination; MRI of the abdomen revealed a 9.9 x 7.6 x 9.6 cm complex solid-cystic right adnexal mass. Exploratory laparotomy was pursued and the mass, that was originating from the mesentery of the right colon and ileum, was removed. Pathology showed extensive plasma cell infiltration, obliterative phlebitis, scattered areas of spindle cells and reactive lymphoid tissue with numerous germinal centers.

Immunohistochemical staining revealed plasma cells positive for MUM1 and marked increase in the Ig G and IgG4 cells (> 50/hpf) with an IgG4:IgG ratio of approximately 60%. Serum concentrations of IgG and IgG4 were normal. Flow cytometry was negative for monoclonal B cells or aberrant T cell expression. The findings were consistent with the diagnosis of IgG4-related fibro-inflammatory mass. The patient had complete resolution of symptoms after the resection and experienced no recurrence for the past 3 years.

Discussion:

IgG4-RD comprises a wide array of manifestations such as: type 1 autoimmune pancreatitis, Riedel's thyroiditis, sialadenitis, dacryoadenitis, interstitial lung disease, pulmonary nodules, bronchial stenosis, sclerosing cholangitis, cholecystitis, mesenteric fibrosis, periaortitis, aortic aneurysm, tubulointerstitial nephritis. IgG4-RD rarely affects the mesentery. According to a recent review of literature by Avincsal et al. only 14 cases of IgG4-RD sclerosing mesenteritis have been reported. Consensus on considering a previously unrecognized site as IgG4-RD requires fulfillment of four criteria – elevated serum IgG4 levels, elevated tissue IgG4 levels, multi-organ involvement and response to glucocorticoid therapy. Our case does not meet all these criteria: the histology showed typical features of storiform fibrosis, obliterative phlebitis and lymphoplasmacytic infiltrate, with tissue IgG4 >50/hpf and IgG4:IgG ratio of 60%, however there was no elevation of the IgG4 levels nor other organ involvement. It is unclear if this is an incomplete form of IgG4-RD versus an IgG4-RD mimic. Questions are yet to be answered and further research is needed to decipher the real extent of IgG4-RD.