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IgG4 Multiorgan involvement

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69 years old female patient presented with complains of acute epigastric pain over the last few hours. Acute abdominal series showed large mass-like opacity at left med/lower lung zone. CT abdomen and pelvis with IV contrast showed acute interstitial pancreatitis IgG 4 levels were elevated, patient's pancreatitis likely secondary to autoimmune pancreatitis. Patient started on prednisone 40 mg once daily with improvement in 2 days after initiation. Plan was made to continue for 4-6 weeks followed by taper. Further thyroid and lung biopsies demonstrated abundance of plasma cells and storiform fibroblasts which consolidated the diagnosis of IgG4 related autoimmune disease with multi system involvement. Immunoglobulin G4-related disease (IgG4-RD) is an immune-mediated fibroinflammatory condition that is capable of affecting multiple organs. Common forms of presentation include pancreatitis, sclerosing cholangitis, sclerosing sialadenitis in lacrimal, parotid and submandibular glands (Mikulicz disease), orbital disease, retroperitoneal fibrosis chronic aortitis often involving ureters and causing hydronephrosis. Finding of a serum IgG4 concentration greater than five times the upper limit of normal and , the histopathologic findings of a dense lymphoplasmacytic infiltrate, storiform fibrosis (typified by a cartwheel appearance of the arranged fibroblasts and inflammatory cells), and obliterative phlebitis are critical features for establishing the diagnosis. Generally, the minimum for making the diagnosis for most tissues is from 30 to 50 IgG4-positive cells/HPF. However, in some organs or tissues, including the kidney and othesars, only 10 IgG4-positive plasma cells/HPF may be sufficient. Although histopathology findings are important to the diagnosis of IgG4-RD, such findings are never diagnostic alone of IgG4-RD. Pathology findings must always be interpreted in the context of clinical, serologic, and radiologic data

Biography

Dr. Bipneet Singh studied medicine at DMCH Ludhiana and graduated as MBBS in 2021. He then was selected for residency in internal medicine at Henry Ford Health Jackson in Internal Medicine. Due to keen interest in GI he worked with the chair of GI Dr. Bern, working on publications and poster presentations with most prominent ACG Vancouver where he won outstanding poster presenter. With plans to apply for GI fellowship at the end of residency, he is currently involved with projects with faculty at Henry Ford Jackson and University of Michigan

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