IMMUNOGLOBULIN G4-RELATED DISEASE -WHAT A CLINICIAN NEEDS TO KNOW

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Immunoglobulin G4-related disease (IgG4-RD) is an immune-mediated fibroinflammatory condition that can affect multiple organs, including salivary glands, pancreas, biliary tree, lymph nodes, orbital and periorbital tissues, retroperitoneal tissues, blood vessels, thyroid, lung, kidney, and some other structures. In the late 19th century, the first accounts of organ manifestations associated with IgG4-related disease were published. Johann von Mikulicz first described a patient with an inflammatory disease of the salivary glands (Mikulicz' syndrome). IgG4-RD should be suspected in individuals with any of the following: pancreatitis of unknown origin, sclerosing cholangitis, bilateral salivary and/or lacrimal gland enlargement, retroperitoneal fibrosis, orbital pseudotumor/proptosis, and development of an otherwise unexplained mass lesion in any of the following organs: pancreas, biliary tree, orbits, lungs, kidneys, major salivary gland, or lacrimal gland. The combination of biliary tract and pancreatic disease, or isolated involvement of the bilateral submandibular glands, is highly suggestive of IgG4-RD. Serum IgG4 levels should be measured in all patients with suspected IgG4-RD. In roughly two-thirds of patients with IgG4-RD, serum IgG4 levels are above the upper limit of normal. A CT scan or ultrasound may be used to image the involved organs. A biopsy of the affected organ is needed to confirm IgG4-RD. The biopsy may reveal lymphoplasmacytic tissue infiltration and storiform fibrosis. Glucocorticoid therapy is recommended for all patients with symptomatic disease or asymptomatic disease affecting a vital organ (e.g., aorta, retroperitoneal fibrosis causing hydronephrosis). In refractory cases, either rituximab, azathioprine, methotrexate, or mycophenolatemofetil can used. Most patients respond well to glucocorticoid at first, although relapses are common after stopping therapy. Uncontrolled, progressive inflammatory and fibrotic changes in affeced tissues can cause significant organ dysfunction. Early diagnosis and treatment improve the prognosis of IgG4-RD.

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