Autoimmune Pancreatitis

TO THE EDITOR: Finkelberg et al. (Dec. 21 issue) review the pathogenesis, diagnosis, and management of autoimmune pancreatitis. We treated a 67-year-old man with a diagnosis of autoimmune pancreatitis made on the basis of computed tomography (CT), endoscopic ultrasonography and biopsy, and elevated IgG levels (including IgG4). The patient had worsening fatigue and generalized weakness. Laboratory studies revealed low levels of thyroid-stimulating hormone, free thyroxine, corticotropin, cortisol, follicle-stimulating hormone, luteinizing hormone, growth hormone, insulin-like growth factor 1, and free testosterone, with a normal prolactin level. Magnetic resonance imaging (MRI) of the pituitary, performed because of the patient’s panhypopituitarism, revealed thickening and masslike enlargement of the infundibulum, consistent with lymphocytic hypophysitis (Fig. 1). Prednisone (40 mg daily) was initiated, with a dramatic improvement in symptoms. Later, levothyroxine and desmopressin were started for diabetes insipidus. Although repeat MRI of the pituitary and CT of the pancreas showed marked improvement, the patient continues to require low levels of hormone replacement.

Lymphocytic hypophysitis, characterized by lymphocytic infiltration of the pituitary gland, is rare but can occur with other autoimmune diseases.1-4 Finkelberg et al. mention the involvement of extrapancreatic organs in autoimmune pancreatitis. This case shows that pituitary-gland involvement also occurs.

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TO THE EDITOR: In response to the review of autoimmune pancreatitis by Finkelberg et al., we report on a 64-year-old man who presented with rapid-onset jaundice and weight loss without pain. Abdominal MRI and endoscopic retrograde cholangiopancreatography showed a hilar bile-duct stricture with wall thickening. Hilar cholangiocarcinoma was strongly suspected, and the patient underwent hilar resection with excision of the caudate lobe. During surgery, the pancreatic segment of bile duct was seen to be sheathed in a pancreatic heterogeneous mass, and a cephalic duodenopancreatectomy was performed. Pathological analysis of the pancreatic and bile ducts did not show neoplastic lesions but did reveal periductal lymphoplasmacytic infiltration, with diffuse fibrosis suggestive of autoimmune pancreatitis. The serum IgG4 level was greatly elevated.

Stenosis of the bile duct frequently occurs with autoimmune pancreatitis. The stenotic portion is usually the lower bile duct; however, strictures of the hilar bile duct or intrahepatic areas are sometimes found.1 Both the bile-duct lesions and pancreatic lesions can improve after corticosteroid therapy.2 When the presentation is indistinguishable...
able from a cancer, a trial of corticosteroids may make it possible to avoid unnecessary surgery.

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TO THE EDITOR: Readers of the review of autoimmune pancreatitis by Finkelberg et al. may be under the impression that 11% of patients with chronic pancreatitis in the United States have autoimmune pancreatitis, which is an overestimation of the disease’s prevalence. The statistic was based on the examination of pancreatic resections for benign disease at the Mayo Clinic, rather than on typical clinical cases of chronic pancreatitis. All 245 cases were forms of chronic pancreatitis, and 27 cases (11%) represented autoimmune pancreatitis with a “tumefactive” presentation. Although some pancreatic resections for suspected cancer are found on histologic analysis to be benign cases of chronic pancreatitis, most patients with typical chronic pancreatitis do not undergo surgery. Conversely, autoimmune pancreatitis may present without tumefaction, so not all patients with autoimmune pancreatitis would be represented in this study. Thus, the actual prevalence of autoimmune pancreatitis in the United States is unknown. In Japan, South Korea, and Italy, prevalences of autoimmune pancreatitis among patients with chronic pancreatitis range from 4.6 to 8.4%. The prevalence of autoimmune pancreatitis in our own experience is even lower.

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THE AUTHOR REPLIES: Ralli et al. remark on the extrapancreatic manifestations of autoimmune pancreatitis. Autoimmune pancreatitis is likely to be a systemic autoimmune disease associated with tissue infiltration by IgG4-positive plasma cells. Although renal, pulmonary, hepatic, and gastro-duodenal lesions have been reported, I was not aware of the possibility of infiltration of the central nervous system. The clinical features of the patient described by Ralli et al. are nearly diagnostic of autoimmune pancreatitis because of the findings on MRI in combination with the elevated serum IgG4 level. The patient’s response to corticosteroids is also consistent with autoimmune pancreatitis. Therefore, I agree that the clinical picture is most consistent with autoimmune pancreatitis with involvement of the hypophysis.

Leclercq et al. comment on bile-duct stenosis as a result of autoimmune pancreatitis. Bile-duct stenosis and its clinical manifestations, such as jaundice and pruritus, are relatively common presentations of autoimmune pancreatitis. Leclercq et al. note one type of stenosis, resulting from extrinsic compression by the involved pancreas. In addition, one must be aware of the possibility of lymphocytic sclerosing cholangitis, which may include diffuse involvement of the bile duct, in contrast to the distal bile-duct stenosis described in their patient. These manifestations are important to recognize because of their responsiveness to corticosteroid therapy.

Raina et al. address the question of the true prevalence of autoimmune pancreatitis. Certainly, the review of surgical pathological specimens in one center is not an ideal method for determining the prevalence of a disease. Nevertheless, the prevalence figures at the Mayo Clinic do not differ substantially from the estimates in Europe and Asia. The prevalence of autoimmune pancreatitis in association with other clinical findings — such as obstructive jaundice, focal pancreatic strictures, and recurrent pancreatitis — varies tremendously. The importance of autoimmune pancreatitis lies not in its modest prevalence but in its excellent prognosis and responsiveness to therapy.

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