Association of IgG4-related disease and sarcoidosis

Autoimmune pancreatitis (AIP) is a syndrome characterised by an enlarged pancreas with an irregular narrowing of the main pancreatic duct, a high serum IgG4 concentration and IgG4-positive plasma cell tissue infiltration.

A wide variety of lesions have been associated with AIP, including pulmonary lesions. Consequently, the terms “IgG4-positive multi-organ lymphoproliferative syndrome (IgG4+ MOLPS)” seem more appropriate.

To our knowledge, sarcoidosis has never been reported in association with AIP. We report the case of an association of AIP with sarcoidosis in an elderly woman.

An 80-year-old woman presented with a 1-year history of chronic diarrhoea and a weight loss of 12 kg during the previous 6 months. Physical examination was normal. Chest and abdominal CT scan disclosed an increased volume of the pancreas, coeliac and hilar lymphadenopathy and pulmonary lesions (figure 1). Pancreas biopsies revealed chronic pancreatitis. Accessory salivary gland and coeliac adenopathy biopsies showed non-caseating giant-cell epithelioid granuloma. The tuberculin purified protein derivative test and the Quantiferon assay were both negative. Laboratory analysis revealed a polyclonal hypergammaglobulinemia with IgG level at 35 g/l; serum IgG4 level was increased at 6.8 g/l (normal <0.8 g/l), white blood cell count revealed a lymphopenia (1000 mm3/l) and ACE was within the normal range.

Because of the histological picture of non-tuberculous granulomas, and mediastinal lymph nodes with pulmonary involvement on chest CT, sarcoidosis associated with an IgG4+ MOLPS was diagnosed. Corticosteroids (1 mg/kg/day) led to a dramatic improvement in the general and digestive manifestations within a 1-year follow-up.

AIP is a form of chronic pancreatitis characterised by a high serum IgG4 concentration and abundant IgG4-bearing plasma cell infiltration in the pancreatic lesion. This entity has been reported to be associated with a variety of extrapancreatic lesions. It is generally accepted that this form of pancreatitis is a part of a multi-systemic clinical syndrome, and this disease was redefined as ‘IgG4-positive multi-organ lymphoproliferative syndrome’.2

Recently, Tsushima et al3 compared the clinicopathological features of pulmonary lesions in 19 patients with AIP and 8 patients with sarcoidosis; 17 of the 19 patients with AIP showed bilateral hilar lymphadenopathy, while 8 showed pulmonary nodules. IgG4-positive plasma cells were identified in the pulmonary lesions of patients with AIP. Our patient presented an authentic chronic pancreatitis with a significant increase in serum IgG4 level. She fulfilled the revised diagnostic criteria for AIP.4 Because of the presence of pulmonary lesions and hilar lymphadenopathies, salivary gland and coeliac adenopathies were biopsied, and they both revealed non-caseating epithelioid cell granulomas. Although sarcoidosis is uncommon in the elderly, the presence of disseminated granulomatous lesions led us to suspect sarcoidosis. However, it is difficult to determine whether our 80-year-old patient has an IgG4-related disease with systemic granulomatous lesions or an association of AIP with true sarcoidosis.

To our knowledge, such an association of AIP with granulomatous lesions mimicking sarcoidosis has never been reported previously in the literature, and this enlarges the spectrum of IgG4-related disease.

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Figure 1 Contrast-enhanced coronal reformatted CT view: hilar and mediastinal adenomegaly, diffuse homogeneous hypootenuating enlargement of the pancreas with minimal peripancreatic stranding.
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