Pseudotumours and IgG4-related disease: a case report

Paul Tan, Graeme Taylor, Rennae Thiessen, Lutz Beckert

Abstract

We report a case of a patient presenting with abdominal pain, weight loss and CT imaging showing mass lesions in the chest and abdomen associated with lymphadenopathy. He was diagnosed as having IgG4-related disease and responded well to steroid treatment.

Case report

A 77-year-old man with known idiopathic non-specific interstitial pneumonia presented with a 3-week history of epigastric discomfort, anorexia and weight loss of 7 kg.

Clinical examination revealed mild epigastric tenderness without evidence of organomegaly or palpable lymphadenopathy. His full blood count and biochemistry tests were within normal limits except for elevated serum lipase of 324 U/L (normal range 8–78 U/L) and marginally elevated amylase of 58 U/L (normal range 8–53 U/L).

A chest X-ray revealed a right hilar lesion. His CT chest abdomen and pelvis showed chest and abdominal lymphadenopathy and mass lesions in the right lung and both kidneys. These findings were thought to be consistent with disseminated malignancy or lymphoma.

The patient underwent an EBUS guided fine needle aspiration of his subcarinal and right hilar lymph nodes followed by a core biopsy of the abdominal paraaortic lymph node/lesion. No malignant cells were seen in any samples.

Core biopsy histology showed a fibroinflammatory process associated with obliterative phlebitis and >30 IgG4 positive plasma cells per high power field in keeping with IgG4 related disease.

Subsequently his serum IgG4 levels were found to be elevated at 3.03g/L (range 0.03–2.01) and he was started on prednisone therapy, 0.5 mg/kg with a tapering regimen.

He made good symptomatic recovery with radiological improvements on serial CT scanning and reductions in his IgG4 levels to 1.52 g/L.
Figure 1. Serial CT scans showing resolution of abdominal paraaortic lymphadenopathy following prednisone therapy (see arrow)

Figure 2. Histology, left to right: Core biopsies showed fibrosis and lymphoplasmacytic infiltrates (H&E stain, magnification ×40); focally there was obliterative phlebitis (Elastic van Geison stain, magnification ×100) and there were >30 IgG4 positive plasma cells per high-power field (IgG4 immunostain, magnification ×200)

Discussion
IgG4-related disease is a fibroinflammatory entity that was only recognised to have systemic manifestations in the last decade.1,2 Conditions such as Riedel’s thyroiditis and Type 1 autoimmune pancreatitis are now considered to be part of the spectrum of IgG4-related disease.1
Clinical presentations are dependent on the organ involved and patients can present with mass lesions that may be mistaken for malignancy, such as was the case in this instance. Tumefactive lesions have been described in many organs systems including
the kidneys, pancreas, lungs, salivary glands, lymph nodes, thyroid and peri-orbital regions.4,5

IgG4 has also been described to cause a NSIP pattern on CT scanning.3,6 In our case the NSIP pattern on radiology was thought to be independent as it remained stable for a period of 4 years and was uninfluenced by the prednisone course.

The diagnosis of IgG4-related disease is made on tissue biopsies showing lymphoplasmacytic infiltrates, a storiform pattern of fibrosis, obliterative phlebitis and increased numbers of IgG4 plasma cells.7

There is a paucity of data regarding the use of serum IgG4 levels in extrapancreatic manifestations of IgG4 related disease for diagnosis or disease activity monitoring. However in IgG4 related autoimmune pancreatitis, elevated IgG4 levels have been reported to have a diagnostic sensitivity of 76% and specificity of 93%.8

The role of serum IgG4 levels in monitoring for disease activity is unclear. Kamisawa et al9 identified that 30% of patients with elevated serum IgG4 in spite of treatment relapsed versus 10% in patients with normal serum IgG4 levels.

The optimal treatment for this condition has not been established but patients typically respond well to oral glucocorticoid therapy with symptomatic and radiological improvements. Other immunosuppressive drugs such as azathioprine and mycophenolate are used, but have not been well studied, and for the most part the natural history of patients with this condition is not well defined

Competing interests: Nil.

Author information: Paul Tan, Respiratory Physician, Department of Respiratory Medicine, Christchurch Hospital, Christchurch; Rennae Thiessen, Radiologist, Department of Radiology, Christchurch Hospital, Christchurch; Graeme Taylor, Pathologist, Department of Pathology, Nelson Hospital, Nelson; Lutz Beckert, Respiratory Physician, Department of Respiratory Medicine, Christchurch Hospital, Christchurch

Correspondence: Lutz Beckert, Department of Respiratory Medicine, Christchurch Hospital, Christchurch 8011, New Zealand. Fax: +64 (0)3 3640914; email: lutz.beckert@cdhb.health.nz

References:
