Human Pathology (2012) 43, 2084–2087

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Case study

IgG4–related paratesticular pseudotumor in a patient with autoimmune pancreatitis and retroperitoneal fibrosis: an extrapancreatic manifestation of IgG4–related disease☆,☆☆

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Received 12 March 2012; revised 18 April 2012; accepted 14 May 2012

Keywords:
Paratesticular;
Pseudotumor;
IgG4-related disease;
Autoimmune pancreatitis

Summary In this report, we describe the first case of a patient with an IgG4–related paratesticular pseudotumor. He had histologically proven autoimmune pancreatitis, then later developed a scrotal mass. The orchiectomy specimen revealed that this was a paratesticular pseudotumor with histopathologic and immunohistochemistry findings characteristic of IgG4–related disease. Paratesticular pseudotumors are uncommon causes of intrascrotal masses and have an unexplained pathogenesis. A variety of genitourinary manifestations of IgG4–related disease including IgG4–related tubulointerstitial nephritis, IgG4–related ureteral pseudotumors, and IgG4–related prostatitis has been previously reported. The current case highlights the need to have a high index of suspicion for IgG4 –tissue infiltration in patients with known autoimmune pancreatitis, particularly those with a pseudotumor.

1. Introduction

Paratesticular fibrous pseudotumors are an uncommon cause of scrotal masses [1-3]. These generally have a benign natural history, and to date, no definite etiologic association has been described. We report the case of a patient with known IgG4-related disease (IgG4-RD), who was later found to have a paratesticular pseudotumor with identical histology to that in IgG4-RD. This is the first case of a patient with a paratesticular, or testicular, manifestation of IgG4-RD.

2. Case presentation

A 67-year-old man presented to the gastroenterology clinic in 2005 with weight loss and uncontrolled diabetes mellitus, which required escalation to insulin therapy despite the concurrent weight loss. With the exception of hyperlipidemia requiring treatment, he had no other medical comorbidities. A computed tomographic scan of the

☆ Grant support: None.
☆☆ Conflict of interest: None.
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http://dx.doi.org/10.1016/j.humpath.2012.05.017

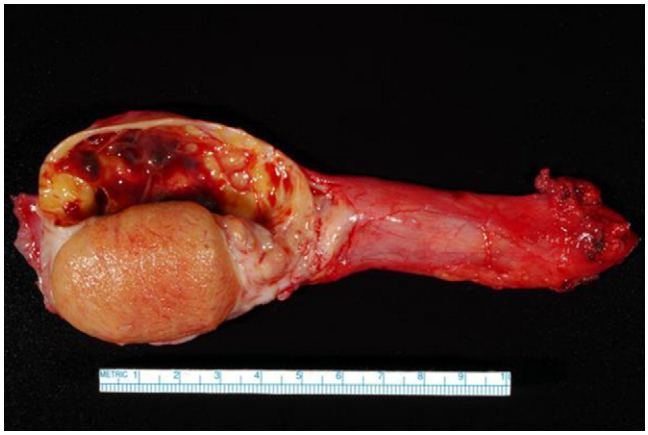

Fig. 1 Transected gross specimen from radical orchiectomy shows a multicystic paratesticular mass.

abdomen revealed diffuse pancreatic enlargement; his serum IgG4 level was elevated at 391 mg/dL (reference <140), and core pancreatic biopsies showed lymphoplasmacytic pancreatitis with many IgG4-positive cells consistent with a diagnosis of autoimmune pancreatitis (AIP). He had retroperitoneal fibrosis causing left hydronephrosis, bilateral renal cortical involvement on imaging, and renal failure (creatinine 1.6 mg/dL and pyuria on urine sediment). He was treated with a 3-month course of prednisone and ureteral stenting, with resolution of the pancreas swelling; improvement in his diabetes control, hydronephrosis, and retroperitoneal fibrosis; and normalization of renal function. Aside from a pancreatic relapse with concurrent acute renal failure requiring an additional course of prednisone, he had no other major medical events.

In fall of 2011, he presented for evaluation of a painless right scrotal mass that had been gradually increasing in size over the preceding 3 months. He denied any lower urinary tract symptoms. An ultrasound with Doppler evaluation showed a multiseptated fluid-filled mass measuring 5.1 × 5.0 × 5.0 cm within the right scrotum; however, it was unclear if this arose from the epididymis or testicle. There was a tiny hydrocele on the left, but otherwise, both testes were normal. Owing to the potential risk of malignancy, albeit small, he underwent right inguinal radical orchiectomy. Intraoperatively, the mass was noted to arise from the right testicular region.

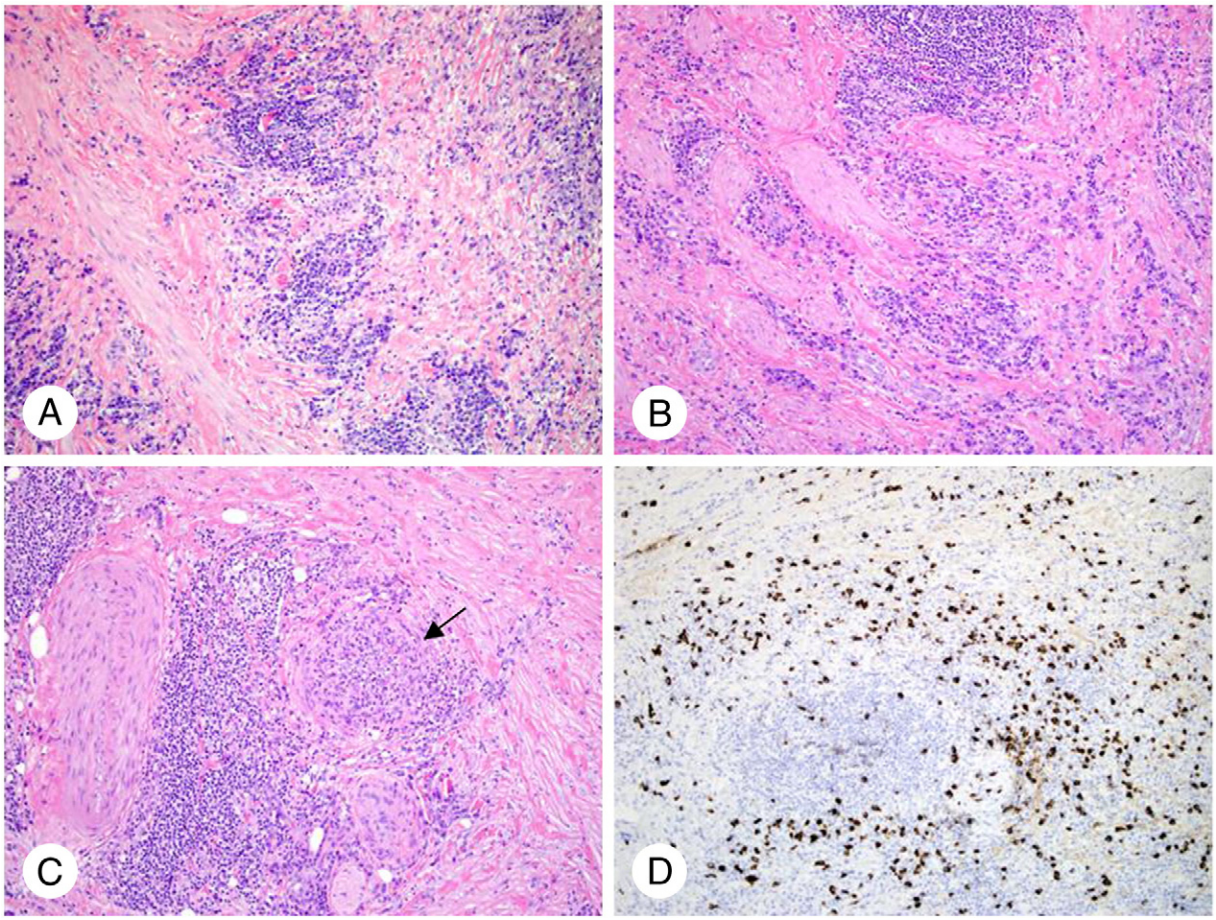

Fig. 2 Low-power views of hematoxylin and eosin stain from paratesticular specimen showing concentrated lymphoplasmacytic infiltrate adjacent to a large band of fibrosis (A), the whorling, or "storiform" pattern of fibrosis (B), and venous obliteration (indicated by the black arrow) (C). Panel D shows marked (>50 per high-power field) IgG4-positive staining plasma cells with IgG4 immunostaining.