

Images in Clinical Urology

Testicular Inflammation as a New Manifestation of IgG4-associated Disease

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IgG4-related disease has properties of a systemic disorder but simultaneously is associated with a growing list of organ-specific manifestations including autoimmune pancreatitis, IgG4-associated cholangitis, IgG4-related kidney disease, and IgG4-associated prostatitis. In this study, we present, to the best of our knowledge, the first case of a patient with multiorgan IgG4-related disease who lost his testes because of IgG4-related testicular inflammation. We postulate that IgG4-related disease in the urogenital tract is not restricted to IgG4-related kidney disease and prostatitis, but that this rare disorder may also affect the testis. *UROLOGY* 82: e15–e16, 2013. © 2013 Elsevier Inc.

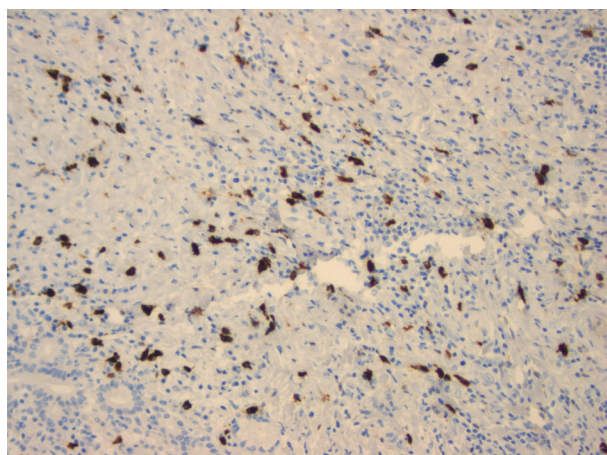


Figure 1. Revision of the surgical specimen of pancreatic tissue collected 5 years earlier during the Whipple procedure. Immunohistochemistry for IgG4 revealed a maximum of 50 IgG4-positive plasma cells per high powered field, supporting the diagnosis of IgG4-related disease.

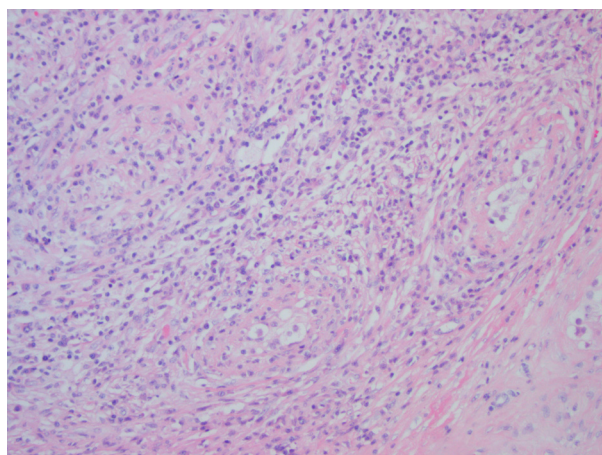


Figure 2. Hematoxylin-eosin staining of the tissue of the right testis showing a plasma cell-rich infiltrate and myofibroblastic spindle cell proliferation around the seminiferous tubules. These changes were distributed unevenly over the tissue and affected the largest part of the testis.

Our patient had undergone Whipple surgery for suspected pancreatic malignancy at the age of 57. Histology disproved malignancy and cholestatic symptoms recurred postoperatively. Revision of the surgical specimen ultimately disclosed an IgG4-positive infiltrate (Fig. 1) allowing the diagnosis of IgG4-related pancreaticobiliary disease.^{1,2} IgG4-related retroperitoneal fibrosis causing left kidney hydronephrosis required double-J catheter placement. Short-term corticosteroids brought some clinical amelioration.

Seven years after pancreaticoduodenectomy, the patient developed scrotal pain, which was treated initially with oral antibiotics under the suspicion of epididymitis. Days later, a normal right testis and inhomogeneous alterations of the left testis were found on ultrasound. Orchidectomy was performed under the suspicion of testicular malignancy, but histology showed benign alterations. Six months later, the patient developed a swollen and tender right testicle. Ultrasonography showed inhomogeneous testicular alterations, suggestive of an uncomplicated orchitis or epididymitis. One month later, acute scrotal pain because of an abscess-forming inflammation necessitated surgical removal of the right testis.

Given the variety of organ manifestations in IgG4-related disease,³⁻⁵ we wondered if the testicular inflammations could have been related to IgG4-related disease. Histology showed IgG4-positive plasmacellular infiltrates

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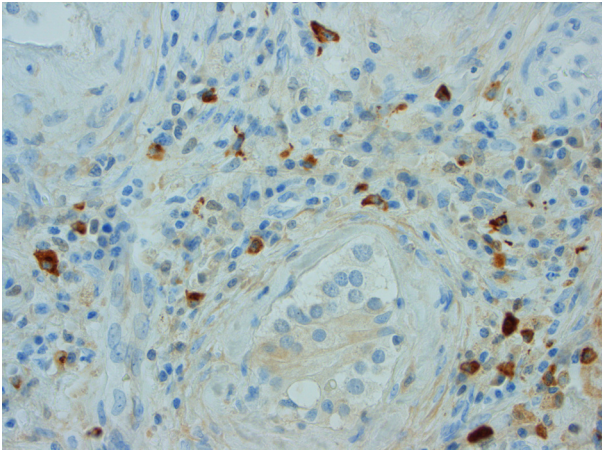


Figure 3. IgG4-staining of the tissue of the right testis showing abnormally high numbers (focally maximum 50 per high powered field) of infiltrating IgG4-positive cells around a seminiferous tubule. The earlier resected left testis showed similar changes including a strongly elevated number of IgG4-positive cells (maximum 80 per high powered field). Meanwhile, a contemporary colonic biopsy showed a normal amount of non-IgG4-positive plasma cells in the mucosa (not shown), supportive of the specificity of IgG4-positive testicular infiltrates.

(Figs. 2 and 3), which are thought to be pivotal in IgG4-related disease,^{6,7} in both testes. IgG4-related disease may thus mimic acute inflammation or malignancy of the testis.

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