Clinical Image – General Urology

Scrotal Hydrocele in Immunoglobulin G4-related Disease

İmmünoglobulin G4 ile İlişkili Hastalıkta Skrotal Hidrosel

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A 78-year-old male patient presented to our hospital with a slowly enlarging, painless right inguinal mass that had been present for 15 years. On physical examination, a non-tender right scrotal mass was palpable. Computed tomography revealed a 12-cm encapsulated cystic lesion containing a calculus (**Figure 1**). Blood tests showed a mildly elevated C-reactive protein (CRP) level of 0.74 mg/dL (reference range: < 0.14 mg/dL). Based on these findings, a diagnosis of right scrotal hydrocele was made, and the patient underwent right orchiectomy with hydrocelectomy. Intraoperative findings indicated that the mass was located in the scrotum and extended toward the external inguinal ring. Orchiectomy was selected because the hydrocele was firmly adherent to the testis, with chronic inflammation and fibrosis, making safe separation technically unfeasible. The resected specimen contained turbid fluid, a calculus, and a markedly thickened, inflamed, and partially necrotic cyst wall (**Figure 2**). Histopathological examination revealed diffuse thickening of the tunica vaginalis with dense lymphoplasmacytic infiltration and characteristic storiform fibrosis (**Figure 3A**). The testis itself was markedly atrophic, and the seminiferous tubules showed diffuse atrophy with thickened basement membranes and hyalinization. The hydrocele was not located within the testicular parenchyma. Immunohistochemical staining demonstrated 27-48 Immunoglobulin G4 (IgG4)-positive plasma cells per high-power field (HPF) and an IgG4-positive/IgG-positive plasma cell ratio of 25-55% (**Figure 3B**). Given the patient's long-standing, slowly enlarging inguinal mass without prior hospital visits, and the marked chronic inflammatory changes observed intraoperatively and pathologically, the exact origin and progression from the inguinal area cannot be determined.

Serum IgG4 was within normal limits, and no involvement of other organs was observed. According to the 2019 American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) classification criteria for IgG4-related disease [1], a diagnosis of IgG4-related disease (IgG4-RD) was established. The postoperative course was uneventful, and the patient remains well on follow-up.

IgG4-RD is a recently recognized, immune-mediated fibroinflammatory disorder characterized by tissue infiltration with IgG4-positive plasma cells, lymphoplasmacytic inflammation, storiform fibrosis, and frequently obliterative phlebitis [2]. It can involve multiple organs, most commonly the pancreas, salivary glands, and lacrimal glands, resulting in clinical entities such as autoimmune pancreatitis, sialadenitis, and dacryoadenitis. Corticosteroid therapy is considered the first-line treatment, although no randomized controlled trials have been conducted

to date. In the present case, systemic therapy was not initiated because serum IgG4 was normal and there was no evidence of extra-scrotal disease.

Reports of IgG4-RD involving the male reproductive system are rare, with only a few testicular cases described in the literature. To our knowledge, presentation as chronic scrotal hydrocele represents an exceptionally uncommon manifestation. This case underscores the importance of considering IgG4-RD in the differential diagnosis of long-standing scrotal masses, particularly when histological features are suggestive. Increased awareness of this entity may facilitate accurate diagnosis, appropriate management, and a deeper understanding of its diverse clinical spectrum.

Keywords: Immunoglobulin G4-related disease, scrotal hydrocele, storiform fibrosis

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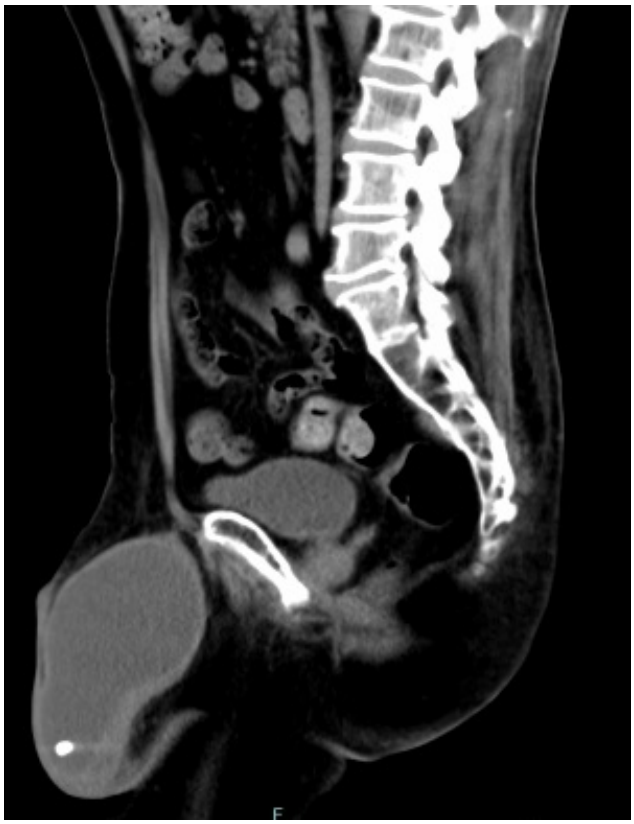

Figure 1. Computed tomography image. Computed tomography scan showing a 12-cm encapsulated cystic lesion with a calcified nodule in the right inguinal region

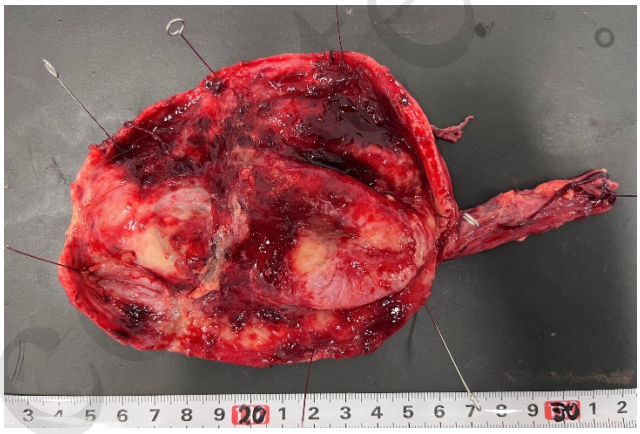

Figure 2. Gross appearance of the resected specimen showing a thickened, inflamed, and focally necrotic cyst wall

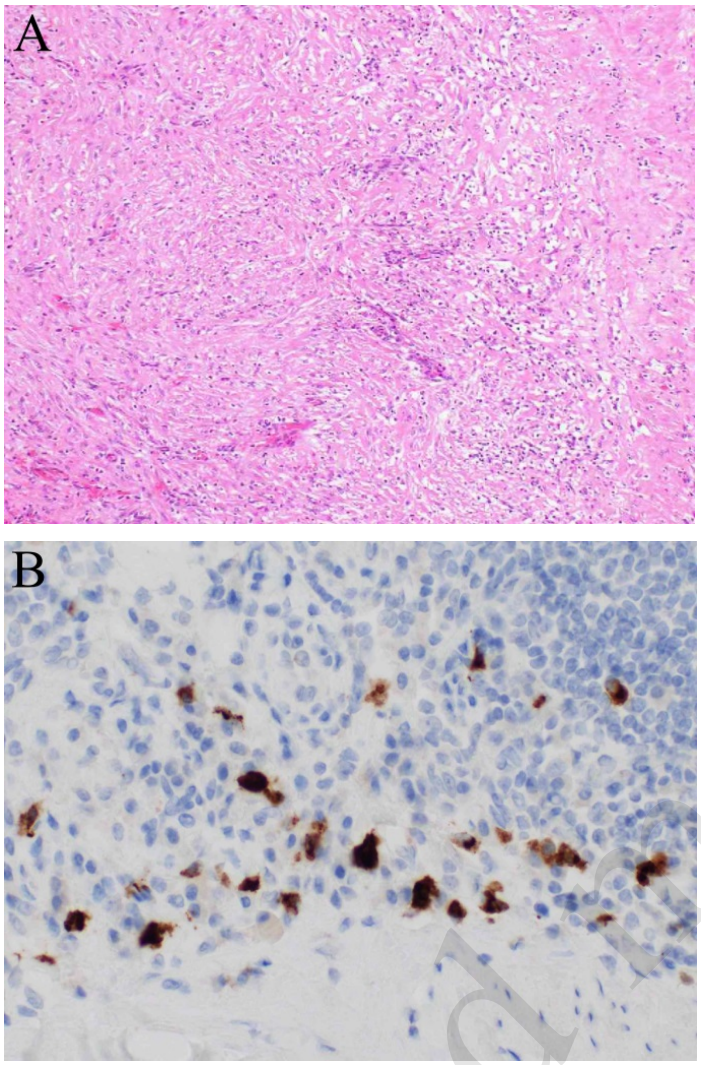

Figure 3. A- Histopathological examination revealing lymphoplasmacytic infiltration and storiform fibrosis (H&E ×100), B- Immunohistochemical stain highlighting numerous IgG4 positive plasma cells (×400)