


Images in Rheumatology

IgG4-related Disease Mimicking a Paratesticular Tumor and Pelvic Lymph Node Metastasis

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IgG4-related disease (IgG4-RD) is a systemic fibro-inflammatory disease characterized by tumor-like mass with infiltration of IgG4-positive plasma cells.¹

A 58-year-old man visited the Department of Urology at Tonan Hospital with a 3-month history of right scrotal swelling. On computed tomography (CT), a right paratesticular tumor (Figure 1) and pelvic mass, which was suspected to be a metastatic obturator lymph node, were observed (Supplementary Figure S1A, available with the online version of this article). He underwent right high orchiectomy with suspicion of malignancy. Histopathological examination showed fibrotic thickening of serosal membrane accompanied with infiltration of lymphoplasmacytic cells, storiform fibrosis, obliterative phlebitis, and abundant IgG4-positive cells (60 per high-power field, and the IgG4/IgG plasma cell ratio was 50%; Figure 2; Supplementary Figures S2A,B). There was no evidence of malignancy. The serum IgG4 level was 527 mg/dL. The patient was diagnosed to have “atypical IgG4-RD” based on a case control study² that

used the 2019 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for IgG4-RD,³ since he had no typical organ involvement. The pelvic mass was considered to be IgG4-related fibrotic tissue. He was treated with 30 mg/day of oral prednisolone, followed by gradual tapering. Follow-up CT 4 months later showed significant regression of the pelvic mass (Supplementary Figure S1B).

Our patient had an IgG4-related paratesticular pseudotumor and lymph node metastasis-like pelvic fibrosis simultaneously. In previous reports, there has been IgG4-related retroperitoneal fibrosis several years prior to onset of a paratesticular pseudotumor.^{4,5} It is important to recognize that a paratesticular pseudotumor might be one of the manifestations of systemic IgG4-RD.

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Figure 1. Coronal contrast-enhanced computed tomography scan shows a slightly enhanced soft tissue mass in the left hemiscrotum.

ONLINE SUPPLEMENT

Supplementary material accompanies the online version of this article.

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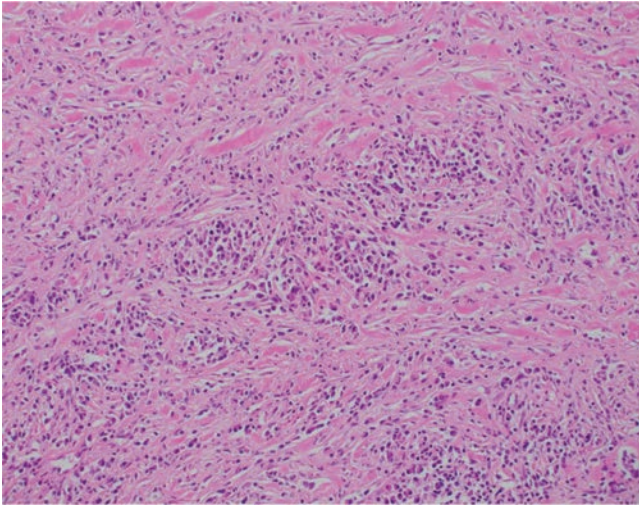


Figure 2. Histopathological sections of the right paratesticular mass showing storiform fibrosis and lymphoplasmacytic infiltration ($\times 200$; H&E staining).