

## Letter

### IgG4-Related Disease With Testicular Involvement: Association or Coincidence?

To the Editor:




We read the recent article by Sugawara and colleagues in *The Journal of Rheumatology* on IgG4-related disease mimicking a paratesticular tumor and pelvic lymph node metastasis with interest.<sup>1</sup> We strongly endorse the point made by the authors regarding the importance of recognizing that paratesticular pseudotumors may be one of the manifestations of systemic IgG4-related disease. We acknowledge the authors' work on this aspect, but we would like to add something new regarding the points made in the article.

First, IgG4-related disease is an immune-mediated chronic, systemic, and autoinflammatory disease that can affect various organs throughout the body. The most commonly affected regions are the biliary system and the pancreas.<sup>2</sup> IgG4-related disease with testicular involvement is a rare entity, with no more than 20 to 30 related cases reported worldwide.<sup>3</sup> Therefore, it is highly likely to be misdiagnosed as testicular malignancy, especially in the elderly population and in patients who are considered to have metastatic manifestations from other sites of the tumor. In the case report by Sugawara et al, a right paratesticular and pelvic mass were observed on computed tomography (CT), making it easy to suspect paratesticular tumor and pelvic lymph node metastasis.

Second, the vast majority of patients with IgG4-related disease with testicular involvement first consult with urologists rather than rheumatologists. However, urologists in conjunction with rheumatologists and pathologists play a crucial role in the diagnosis and treatment of IgG4-related disease, especially when diagnosis is between autoimmune disease and malignancy. Early identification and detection of abnormally elevated IgG4 levels can shift diagnostic thinking to rheumatic diseases. Therefore, testing for Ig levels, such as IgG4, is recommended as a priority in patients with a suspected autoimmune disease with tumor-like manifestations. If abnormal test results are found, prompt consultation with rheumatologists and pathologists is required. The patient can be observed for symptom relief after standard-dose hormone therapy. For those patients who have remission on CT after treatment, removal of the testes may be withheld. To say the least, testicular puncture biopsy is also preferred over orchiectomy because the testes are important reproductive organs in men, especially in younger individuals, and perform an important function in maintaining secondary sexual characteristics. Therefore, orchiectomy at the outset is not a wise move.

Finally, a study revealed that intrascrotal masses, testicular swelling, and pain were usually the first symptoms of the disease, with testicular swelling usually seen in 44.4% of patients and testicular pain in 27.7%.<sup>3</sup> At the same time, the right testicle was significantly more frequently involved than the left, with unilateral involvement accounting for the majority of cases,<sup>3</sup> which was verified in the present patient.<sup>1</sup> Similarly, in the previous report,<sup>3</sup> 22% of patients had a history of IgG4-related retroperitoneal fibrosis several years prior to the onset of paratesticular pseudotumor. We need to emphasize that primary retroperitoneal fibrosis causing hydrocele involving the ureter, inferior vena cava, and abdominal aorta and its branches needs to be differentiated from IgG4-related disease with testicular involvement.<sup>4</sup> Multiple abdominal CT scans and pathologic biopsies after treatment can help in the differentiation. Retroperitoneal fibrosis provides an important clue for the diagnosis of testicular involvement in IgG4-related disease.

In conclusion, we thank Sugawara and colleagues for their work,<sup>1</sup> which adds important real-world data on IgG4-related diseases with testicular involvement as the first manifestation.

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The authors declare that they have no conflicts of interest relevant to this article.

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