Urology Case Reports 24 (2019) 100844

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Functional medicine

Female urinary retention from a huge periurethral mass caused by immunoglobulin G4-related disease (IgG4-RD)

Phadungsak Sangsoad[a], Patkawat Ramart[a,*], Pornpim Korpraphong[b], Vilasinee Rerkpichaisuth[c], Kanapon Pradniwat[c], Jitsupa Treetipsatit[c]

[a] Division of Urology, Department of Surgery, Faculty of Medicine, Siriraj Hospital, Mahidol University, 12th Fl. Syamindra Bldg., Siriraj Hospital, Prannok Rd., Bangkok-Noi, Bangkok, 10700, Thailand
[b] Division of Diagnostic Radiology, Department of Radiology, Faculty of Medicine, Siriraj Hospital, Mahidol University, Bangkok, Thailand
[c] Department of Pathology, Faculty of Medicine, Siriraj Hospital, Mahidol University, Bangkok, Thailand

ARTICLE INFO

Keywords:
IgG4-related disease
Urinary retention
Periurethral mass

ABSTRACT

Immunoglobulin G4-related disease is a systemic disease, recognized as extensive T-lymphocyte and IgG4-positive plasma cells. It can present as inflammatory pseudotumor in various organs. A female 75 years old, diagnosed IgG4-related autoimmune pancreatitis, presented with urinary retention. Pelvic examination showed well-defined, soft tissue mass, bulging from anterior vaginal wall. MRI pelvis demonstrated a huge periurethral mass, size 6.2 × 4.4 × 4.2 cm, encasing the urethra, extending from bladder neck to distal urethra, and mimicking the prostate gland. Tissue biopsy showed compatible with IgG4-related disease. Immunosuppresive drugs were given for few months and the patient could void normally.

Introduction

IgG4-related disease (IgG4-RD) is a systemic fibroinflammatory condition characterized by a mass-like inflammatory lesion comprising dense lymphoplasmacytic infiltrate rich in IgG4+ plasma cells and storiform fibrosis, and elevated serum IgG4 concentrations.[1,2] Organs commonly involved include pancreas, salivary gland, and lacrimal gland. Urethral involvement is much less common.

Case presentation

A 75 years old woman presented with urinary retention. The patient was diagnosed IgG4-related autoimmune pancreatitis 2 years ago and treated with corticosteroid. She had never had lower urinary tract symptoms. Physical examination, in conjunction with ultrasonography (USG), showed a hypoechoic mass in the periurethral area resulting in a bulge in the anterior vaginal wall. Magnetic resonance imaging (MRI) revealed extent of the mass from the distal urethra to the bladder neck, which measured 6.2×4.4 × 4.2 cm in size and the mass encased the urethra. Transvaginal biopsy was performed. Histopathology of the biopsy showed dense lymphoplasmacytic and eosinophilic infiltration in the subepithelial connective tissue with patchy storiform fibrosis. $CD3^+$ T-lymphocytes comprised the majority of the lymphoid infiltrates with a minor component of $CD20^+$ B-lymphocytes. Infiltrating plasma cells were polytypic. IgG4 and IgG immunostains demonstrated that there were > 100 IgG + plasma cells per high power field with IgG4+/IgG + plasma cell ratio up to 50% (Fig. 1). IgG4-RD with the periurethral mass-forming lesion was diagnosed based on the clinical, radiologic, and histopathologic findings. The patient was treated with high dose corticosteroids and Azathioprine. After few months, the patient was able to void without catheter and did not have lower urinary tract symptoms. Follow-up USG and pelvic MRI showed that the lesion markedly decreased in size (Fig. 2). Uroflowmetry showed normal. (Fig. 3).

Discussion

IgG4-RD is an immune-mediated, fibroinflammatory condition that affect nearly organ system.[1] The disease has been described in virtually every organ system: the pancreas, biliary tree, salivary glands kidneys, lungs, lymph nodes, meninges, aorta, breast, prostate, thyroid, pericardium and skin1. Only few cases of urethral involvement were reported.[3] In addition, IgG4-RD could also manifest as inflammatory pseudotumor in many organs.[4] Pathophysiology of IgG4-positive cell remained unclear in pseudotumor. Importantly, the mass must be histologically proved that is related with IgG-4 RD because the

* Corresponding author.
E-mail address: patkawat.ram@mahidol.ac.th (P. Ramart).

https://doi.org/10.1016/j.eucr.2019.100844
Received 1 January 2019; Received in revised form 19 January 2019; Accepted 30 January 2019
Available online 31 January 2019

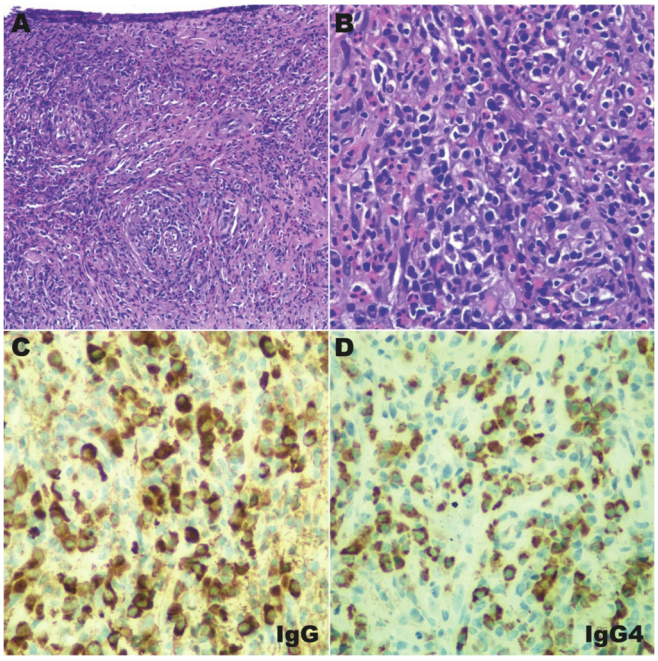

Fig. 1. Histopathology of the transvaginal biopsy. (A and B) Dense lymphoplasmacytic and eosinophilic infiltration with storiform fibrosis in the subepithelial connective tissue. (C and D) More than 100 IgG + plasma cells/HPF noted with IgG4+/IgG + plasma cell ratio up to 50%. [Hematoxylin & Eosin, original magnification x100 (A), x400 (B); immunohistochemistry for IgG, original magnification x400 (C); immunohistochemistry for IgG4, original magnification x400 (D)].

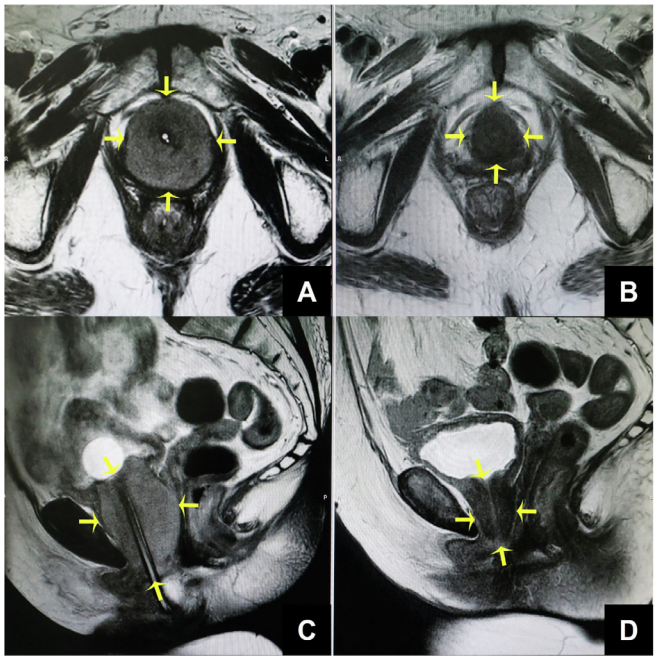

Fig. 2. T2-weighted MRI pelvis compared between before and after treatment, (A and C) were cross-section and sagittal view at the time of urinary retention. The mass was 6.2 × 4.4 × .4.2 cm. in size. (B and D) were cross-section and sagittal view after treatment. The mass was significantly decreased and measured 3.0 × 2.2 × 2.4 cm. in size.

pseudotumor can mimic to the malignant tumors.[5] There is still

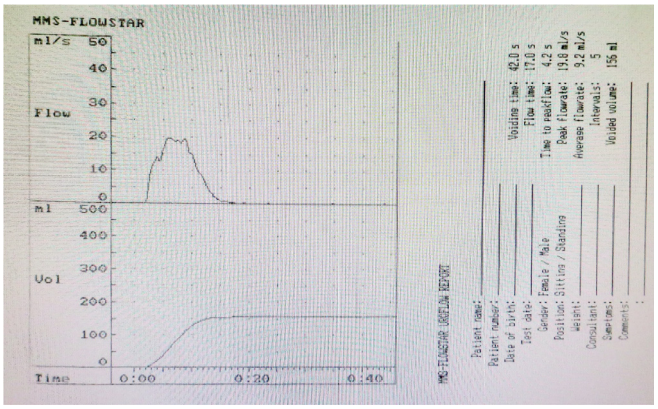

Fig. 3. Uroflowmetry showed the voiding pattern that nearly normal after treatment with immunosuppressive drugs after few months.

unexplored field form a radiological view to identify. Because of history, physical examination and findings on imaging studies in this case, the core needle biopsy was considered and performed in order to get the tissue for definite diagnosis. An ultrasound guided core needle biopsy was very helpful and safe to identify as well as give us about detail of the mass. This method should be considered as an important tool. There are few reports about IgG4-RD involving the urethra or how to manage appropriately, therefore, the patient with history of IgG4-RD who presents with periurethral mass should be considered as clinical manifestation of the IgG4-RD and avoid the unnecessary treatment.

Conclusion

The patient with history of IgG4-RD who present with periurethral mass should think about the inflammatory pseudotumor in differential diagnosis. The lesion can mimic the malignant tumor. The biopsy is investigation for helping in diagnosis, giving the proper management, and avoiding unnecessary treatment.

Conflicts of interest

The authors declare that they have no competing interests.

Acknowledgement

Supot Pongprasobchai M.D., Division of Gastroenterology, Department of Medicine, Faculty of Medicine, Siriraj Hospital, Mahidol University.

References

1. Stone JH, Zen Y, Deshpande V. IgG4-related disease. *N Engl J Med.* 2012;366(6):539–551.
2. Deshpande VZY, Chan JKC, Yi EE, Sato Y, Yoshino T, et al. Consensus statement on the pathology of IgG4-related disease. *Mod Pathol.* 2012;25(9):1181–1192.
3. Choi JW, Kim SY, Moon KC, Cho JY, Kim SH. Immunoglobulin G4-related sclerosing disease involving the urethra: case report. *Korean J Radiol.* 2012;13(6):803–807.
4. Park SB, Cho KS, Kim JK, et al. Inflammatory pseudotumor (myoblastic tumor) of the genitourinary tract. *AJR Am J Roentgenol.* 2008;191(4):1255–1262.
5. Yamamoto H, Yamaguchi H, Aishima S, et al. Inflammatory myofibroblastic tumor versus IgG4-related sclerosing disease and inflammatory pseudotumor: a comparative clinicopathologic study. *Am J Surg Pathol.* 2009;33(9):1330–1340.