Case Report

Limited Wegener’s granulomatosis of the epididymis and testis

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Abstract

A case is presented of Wegener’s granulomatosis limited to the testis and epididymis, simultaneously, in a 69-year-old man. Orchiectomy was carried out through an inguinal incision under the presumptive diagnosis of a right testicular tumor. A hard, irregular mass occupied the upper testicle and a portion of the epididymal head was visualized. Histopathologic examination of the specimen showed granulomatous inflammation of the testis and epididymis with prominent angiocentric granulomata in the walls of arteries, veins and foci of fibrinoid necrosis, surrounded by palisading inflammatory cells with a few giant cells. The diagnosis of limited Wegener’s granulomatosis was considered, although antineutrophil cytoplasmic antibody (c-ANCA) test was negative 2 weeks after orchiectomy. The patient showed an excellent response after local complete excision. He remains free of disease 18 months after orchiectomy. (Asian J Androl 2006 Nov; 8: 737–739)

Keywords: Wegener’s granulomatosis; testis; epididymis

1 Introduction

Wegener's granulomatosis is a distinct clinicopathologic entity characterized by necrotizing granulomatous vasculitis affecting various organs in the body, with a propensity to involve the upper and lower respiratory tract and the kidneys. Klinger [1] first reported this disease in 1931, followed by Wegener in 1939, who further described a series of cases about the clinical features and pathological findings of the syndrome that would eventually bear this name [2]. Its clinical presentation includes upper airway symptoms (nasal obstruction, ulcer or discharge, sinusitis and otitis), pulmonary problems (cough, hemoptysis and dyspnea) or renal disease in more advanced cases (necrotizing glomerulonephritis and uremia). The diagnosis of Wegener’s granulomatosis is mainly made by the histological demonstration of vasculitis, granulomatous inflammation and necrosis in involved organs. Immunosuppressive agents are the choice of treatment for the Wegener’s granulomatosis affecting many organs, which carries a poor prognosis if untreated. Even though Wegener’s granulomatosis is considered to be a systemic disease, the central nervous system, heart, parotid gland and urogenital tract are likely to be involved in less than 1% of cases. Urogenital involvement other than the kidneys is rare and, when encountered, is mostly confined to the prostate gland, but nearly all cases have been in the setting of concurrent active renal involvement, multisystem disease or patients already diagnosed as having Wegener’s granulomatosis.
The limited form of Wegener’s granulomatosis involving the urogenital tract (scrotum, testis, bladder, prostate, urethra, epididymis and penis) is very rare, though has been reported before [3]. The present case report presents the first case of Wegener’s granulomatosis affecting the epididymis and testis simultaneously. Besides immunosuppressive agents, high surveillance following complete excision might be the only choice of treatment for this limited Wegener’s granulomatosis.

2 Case report

A 69-year-old man presented with an 8-month history of right testicular mass with tenderness reported at the initial month. The mass was not associated with genitourinary symptoms, fever, night sweating or weight loss. The patient’s past medical history was unremarkable. No symptoms or disorders involving the ear, nose, throat, respiratory tract or eyes were noted. No allergic history was noted. Clinical examination revealed a non-tender irregular mass of 20 × 10 mm in size at the upper pole of the right testicle. Vital signs showed a pulse rate of 80 beats/min, respiratory rate of 16/min, temperature 37°C and blood pressure 136/78 mmHg. Examination of the head and neck was unremarkable. A complete blood count revealed hemoglobin of 13.6 mg/dL with normal white blood cell and platelet counts. Serum biochemistry was unremarkable with blood urea nitrogen (BUN) of 16 mg/dL and creatinine of 1.5 mg/dL. Renal and liver chemical profiles and urinalysis were all normal. Tumor markers, including alpha fetal protein, beta-HCG and low-dose heparin (LDH), were all within normal range. Urine culture was negative. Chest X-ray was also normal.

Orchiectomy was carried out through inguinal incision under the presumptive diagnosis of right testicular tumor. A hard, irregular mass, brown to gray in color occupied the upper testicle and a portion of the epididymal head was visualized (Figure 1). The histopathologic examination of the specimen showed granulomatous inflammation of the testis and epididymis with prominent angiocentric granulomata in the walls of arteries, veins, and foci of fibrinoid necrosis, surrounded by palisading inflammatory cells with a few giant cells (Figure 2). Blood vessels showed intimal fibrosis and luminal occlusion. The tuberculin polymerase chain reaction (PCR) study of epididymis and testis tissue showed negative findings. The diagnosis of limited Wegener’s granulomatosis was considered, although antineutrophil cytoplasmic antibody (c-ANCA) test was negative 2 weeks after orchiectomy. Because the patient had no other constitutional symptoms or evidence of systemic disease associated with Wegener’s granulomatosis, and a negative c-ANCA test, no immunosuppressive drugs were given. The patient was free of Wegener’s granulomatosis for 20 months after surgery. The c-ANCA test has remained negative during the follow-up period.

3 Discussion

Wegener’s granulomatosis is an uncommon condi-
tion characterized by inflammatory processes of small and medium-sized arteries. It is characterized by glomerulonephritis, necrotizing vasculitis and granuloma typically involving the upper and lower respiratory tract. Lower urogenital tract involvement is a rare presentation of this uncommon disease and mainly involves the prostate [4]. Wegener’s granulomatosis localized only to the lower urogenital tract without respiratory and renal system involvement is very rare. Davenport et al. [3] presented eight patients with limited Wegener’s granulomatosis localized to the urogenital tract, and only two cases presented with testicular involvement. Other sites involved in limited Wegener’s granulomatosis in their series were scrotal skin, urinary bladder, urethra and penis. To date, only one limited Wegener’s granulomatosis of the epididymis was reported by Al-Arfaj [5]. In the present report, we present the first case of limited Wegener’s granulomatosis affecting both the testis and epididymis concurrently.

The diagnosis of Wegener’s granulomatosis usually requires an open or thoracoscopic lung biopsy or a kidney biopsy. However, Agraharkar et al. [6] reported one patient with Wegener’s granulomatosis diagnosed by testicular biopsy. Therefore, the importance of a thorough genital examination for testicular involvement in Wegener’s granulomatosis was stressed and its potential as an easily accessible biopsy site for confirmation of diagnosis in Wegener’s granulomatosis was recognized.

The diagnosis of Wegener’s granulomatosis in the present case was difficult to establish preoperatively. The diagnosis was made after complete excision of testis and epididymis. Antineutrophil cytoplasmic antibody is widely used as a diagnostic test for Wegener’s granulomatosis and as a marker of disease activity. The sensitivity of c-ANCA is 70–90% in classic Wegener’s granulomatosis and 65–70% in the limited or inactive disease [7]. Although a positive c-ANCA test might be useful in the diagnosis of Wegener’s granulomatosis, it should not replace tissue biopsy to make the final diagnosis. Negative c-ANCA test in this patient might be explained by the limited nature of the patient’s disease and also by complete excision of the lesion.

Currently, early diagnosis and prompt initiation of immunosuppressive therapy is recommended for treating patients with limited Wegener’s granulomatosis. However, because this patient showed no evidence of systemic disease or a positive c-ANCA test, no immunosuppressive agents were given to the patient. This patient remains free of disease 20 months after complete excision. The present case is the first report of limited Wegener’s granulomatosis affecting both the testis and epididymis, and the excellent outcome of complete surgical excision is highlighted.

References


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