Triple Wegener's Granulomatosis in the Urogenital Tract

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thrae. TUR of the prostate was done, and a histological diagnosis of necrotizing granulomatous prostatitis was obtained (fig. 1). Upper airway symptoms reappeared at that time and tests for ANCA were repeatedly positive in that period with an antibody titer of 1:32 or 1:64. Therefore, a clinical relapse of WG was obvious and he was treated again successfully with immunosuppressive therapy.

This case is unique because of the development of 3 different manifestations of WG in the urogenital tract: retroperitoneal fibrosis, extracapillary glomerulonephritis and granulomatous prostatitis. Extrarenal urogenital manifestations of WG have been reported infrequently. Prostatic involvement in WG has been estimated to amount to 2-7% [1], and involvement of seminal vesicles, testes, urethra and bladder wall has been reported sporadically [2-8]. Four cases have been described with ureteric obstruction due to an

Dear Sir,

Involvement of the urogenital tract in Wegener's granulomatosis (WG) is usually restricted to the kidneys. We describe a case with 3 different disease localizations in the urogenital tract. A diagnosis of retroperitoneal fibrosis was made in a 49-year-old man complaining of a persistent ache in the groins in 1981. Ureterolysis was performed because of obstruction of the left renal system. Histology showed necrotizing granulomatous vasculitis. He developed a rapidly progressive nonoliguric renal insufficiency 1 month later. A renal biopsy disclosed local and focal extra-capillary glomerulonephritis with necrotizing vasculitis. Apart from this, he had developed conjunctivitis and rhinitis; ENT evaluation showed erosions on the mucous membrane of the nose septum with signs of chronic inflammation. Pulmonary examination revealed a cavitary coin lesion of the right lung. The diagnosis of WG was unmistakable with involvement of all 3 classic target organs. The patient was treated with cyclophosphamid, corticosteroids and temporary hemodialysis. He made an uneventful recovery albeit with an impaired renal function (serum creatinine 280 $\mu\text{mol/l}$). Renal function gradually deteriorated in the course of the next 7 years and chronic intermittent hemodialysis had to be started. A test for the presence of anticytoplasmatic antibodies (ANCA) had become available at that time, and antibodies against proteinase 3 were detected in a titer of 1:64.

The patient was readmitted twice within 2 months to our hospital in 1991 because of an acute urinary retention 9 years after the first event. The initial cystoscopy showed bleeding rags of prostate tissue, the one performed during the second admission showed granulomatous tissue in the pars prostatica ure-

Fig. 1. Prostatic gland tissue (top left) and necrotizing granulomatous prostatitis (bottom). HE. × 100.

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inflammatory infiltration associated with a generalized WG [9-13].

Apart from the above, this case is the more noteworthy because both active disease periods were heralded by atypical manifestations of urogenital disease. Patients with symptoms of prostatic involvement as the first clinical evidence of WG have been reported occasionally [1,2,14-17]. However, retroperitoneal fibrosis as the sole preliminary symptom of WG has not been described before.

Early diagnosis of WG may be hampered by early atypical disease manifestations and lack of pathognomonic histologic evidence. Nevertheless, the possibility of WG should be remembered in the case of an uncommon-prehended urogenital chronic aspecific inflammation, especially with the presence of granuloma formation or necrotizing vasculitis. The detection of anticytoplasmatic antibodies is a useful aid in making this diagnosis.

References

- Stillwell TJ, DeRemee RA, McDonald TJ, Wei-land LH, Engen DE: Prostatic involvement in Wegener's granulomatosis. *J Urol* 1987; 138: 1251-1253.
- Friend DS: Wegener's granulomatosis: A case report. *Arch Intern Med* 1963;111:703-709.
- Fienberg R: Necrotizing granulomatosis and angitis of the lungs with massive splenic necrosis and focal thrombotic granulomatous glomerulonephritis. *Am J Clin Pathol* 1953; 23: 413-428.
- Fahey JL, Leonard E, Churg G, Godman GC: Wegener's granulomatosis. *Am J Med* 1954; 17: 168-179.
- Berman DA, Rydall RE, Eichenholz A: Wegener's granulomatosis: A clinicopathological study of four cases. *Ann Intern Med* 1963; 59: 521-530.
- Tuhy JE, Maurice GL, Niles NR: Wegener's granulomatosis. *Am J Med* 1958;25:638-646.
- Fowler M, Martin SA, Bowles W, Packman R, Katzenstein A-L: Wegener's granulomatosis: Unusual cause of necrotizing urethritis. *Urology* 1979;14:66-69.
- Jensen KM-E, Nielsen KK, Kock K: Necrotizing urethritis in Wegener's granulomatosis. *Br J Urol* 1982; 54:434.
- Adelizzi RA, Shockley K, Pietras JR: Wegener's granulomatosis with ureteric obstruction. *J Rheumatol* 1986;13:448-451.
- Case records of Massachusetts General Hospital. *N Engl J Med* 1977;297:1164-1172.
- Leche J, Feuilhade C, Ferroir JP, Moulonget Doleris L, Guillard A: Maladie de Wegener à début urologique et neurologique. *Ann Méd Interne* 1985 ;136:353.

Ronco P, Mignon P, Lanoe Y, Roland J, Morel-Maroger L, Gattegno B: Ureteral stenosis in Wegener's granulomatosis. *Nephron* 1982;30: 201-204.

Baker SB, Robinson DR: Unusual renal manifestations of Wegener's granulomatosis: Report of two cases. *Am J Med* 1978;64:883-889.

Yalowitz PA, Greene LF, Sheps SG, Carlin MR: Wegener's granulomatosis involving the prostate gland: Report of a case. *J Urol* 1966;96: 801-804.

Sözer IT: Wegener's granulomatosis with infiltration of the prostate. *Br J Urol* 1986; 58:335.

Fanger H, Hoffman M: Wegener's granulomatosis: A case report. *J Urol* 1957;77:78-81.

Murty GE, Powell PH: Wegener's granulomatosis presenting as prostatitis. *Br J Urol* 1991;63: 107-108.