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Case Report

Prostatitis and Acute Urinary Retention as First Manifestations of Wegener's Granulomatosis☆

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ABSTRACT

Objectives: We present a case of prostatitis with acute urinary retention as a rare initial manifestation of Wegener's granulomatosis.
Methods: The case was a 48-year-old male with symptoms of prostatitis over 10 days. The patient presented urinary retention, with partial response to antibiotic treatment. High levels of cytoplasmic antineutrophil cytoplasmic antibody and a prostatic biopsy were compatible with Wegener's granulomatosis.
Results: After starting treatment with glucocorticoids and cyclophosphamide, a significant improvement to the point of disappearance of symptoms was observed. At 3 months pulmonary and upper airway symptoms began, requiring higher doses of cyclophosphamide to control symptoms.
Conclusions: Wegener's granulomatosis is a multisystem entity whose presentation as prostatitis with urinary retention is rare.

Prostatitis y retención aguda de orina como comienzo de granulomatosis de Wegener

Palabras clave:
Granulomatosis de Wegener
Prostatitis
Anticuerpos citoplasmáticos contra los neutrófilos con patrón citoplasmático

RESUMEN

Objetivos: Presentar un caso de prostatitis con retención aguda de orina como manifestación inicial poco frecuente de granulomatosis de Wegener.
Métodos: Se presenta el caso de un varón de 48 años con un cuadro de prostatitis de 10 días de evolución, que presentó retención de orina, con respuesta parcial al tratamiento antibiótico, y con niveles elevados de anticuerpos citoplasmáticos contra los neutrófilos con patrón citoplasmático y estudio anátomo-patológico de la biopsia prostática compatible con granulomatosis de Wegener.
Resultados: Tras el inicio de tratamiento mediante glucocorticoides y ciclofosfamida se observa mejoría notable de los síntomas hasta su desaparición. A los 3 meses inicia clínica pulmonar y de vías aéreas superiores, precisando para el control de sus síntomas dosis mayores de ciclofosfamida.
Conclusiones: La granulomatosis de Wegener es una entidad multisistémica cuya forma de presentación como prostatitis con retención de orina es poco frecuente.

Introduction

Granulomatosis with polyangiitis (GPA)[1] previously known as Wegener's granulomatosis, is a clinicopathologic entity described by F. Wegener in 1936, characterized by granulomatous lesions in multiple organs and varying degrees of disseminated vasculitis of arteries and veins of medium and small caliber.[2] The pathogenesis of the disease remains unknown, although it is believed that an abnormal hypersensitivity response develops against exogenous or endogenous antigens, probably located in the upper respiratory

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tract, synthesizing antibodies to proteinase 3, an enzyme present in polymorphonuclear cells. These antibodies are associated to a granular cytoplasmic staining pattern in these cells using indirect immunofluorescence, or anti-neutrophil cytoplasmic antibodies (c-ANCA) and may contribute to the pathogenesis of the disease.[3] It affects men and women, with a 1:1 ratio, and the highest incidence occurs in the fifth decade of life, with an estimated 20 cases per million per year prevalence.

It affects the upper airway (92%) and lungs (85%), with granulomatous lesions, necrotizing vasculitis, as well as the kidneys (77%) in the form of glomerulonephritis.[4] Urological extrarenal manifestations are very low in frequency and almost always develop in an advanced multisystem disease. We present a case of prostatitis as the first manifestation of Wegener's granulomatosis.

Clinical Case

The patient was a 48-year-old male who was admitted with fever, urinary symptoms, suprapubic pain and acute urinary retention. The onset of symptoms had been 10 days before without other accompanying phenomena present. His medical history was unremarkable, except for being an ex-smoker of 15 cigarettes/day. On physical examination, temperature was 39 °C, he had a distended bladder (evacuating, after a suprapubic cystostomy, 750 ml), with painless renal fist percussion, and normal external genitalia .Rectal examination was very painful, with a grade 2/4 prostate, congestive without suspicious fluctuating abscessed areas.

Other initial tests highlighted leukocytosis ($14.4 \times 10^3/\mu l$, neutrophils 70.2%), with the rest of the parameters being a normal blood count, a urine sediment with pyuria (410–420 cells/field), hematuria (20–25 red blood cells/field) and intense bacteriuria. All other laboratory tests were normal.

With the presumptive diagnosis of acute bacterial prostatitis, treatment was started with ceftriaxone 2 g/24 h intravenously (iv), IV anti-inflammatory drugs and alpha blockers orally (po). The evolution was torpid, with fever and suprapubic pain yielding partially, but then developing asthenia, anorexia, joint/muscle pain and weight loss 10 kg in 1 week. Given the negative results of blood cultures and urine, we started meropenem 1 g/8 h iv and new laboratory tests were performed: complete blood count with WBC 17,000 (*N* 93%), hemoglobin 12 g/dl, hematocrit 36%, mean corpuscular volume 87 fL, ESR: 36 mm/h. Biochemistry: alanine aminotransferase 81 U/l, gammaglutamil transpeptidase: 159 U/l. Rheumatoid factor negative. C-reactive protein: 69. Salmonella and Brucella agglutinations were negative. C3, C4, AAT, copper and ceruloplasmin: normal. Antinuclear antibodies, anti-mitochondrial antibodies, antibodies to extractable nuclear antigens and anti-LKM1: negative. Anti-neutrophil cytoplasmic antibodies (ANCA) were positive at a titer of 1/160 (16 U/ml) by indirect immunofluorescence, with a cytoplasmic pattern. Proteinogram: normal. Human chorionic gonadotropin beta and α-fetoprotein: normal. PSA 1.7 ng/ml. Syphilis, human immunodeficiency virus, B and C hepatitis virus serology were negative. The Löwenstein–Jensen urine culture was negative.

Chest X-ray and CT were normal. Abdominal ultrasound and CT showed a prostate with a volume of 56 cm^3, without evidence of abscess. Flexible cystoscopy confirmed the enlargement of the prostate.

An evaluation by the Rheumatology Department of our center was requested and, suspecting GPA, a prostate biopsy was performed, the pathologic diagnosis being necrotizing epithelioid granuloma and vasculitis of a Wegener type (Fig. 1). Treatment with methylprednisolone was established 40 mg/8 h iv and cyclophosphamide 100 mg/24 h vo, increasing to 150 mg/24 h 5 days later, with disappearing symptoms and a reduction of the prostatic volume to 34 cm^3. The suprapubic cystostomy was removed to recover spontaneous voiding, with postvoid residue less than 50 cm^3. Analytically, the c-ANCA became negative.

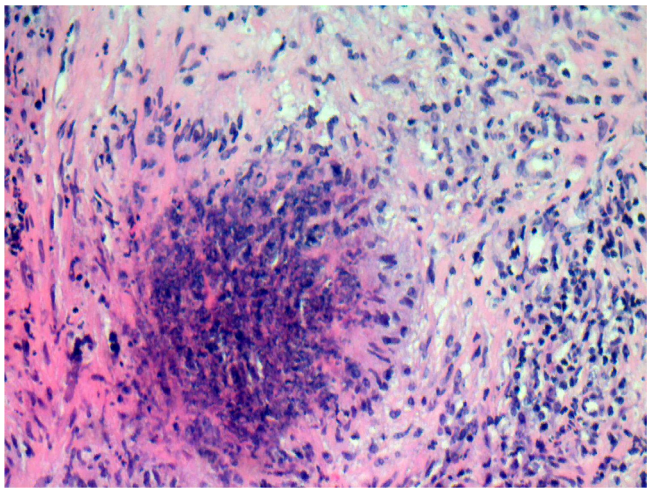

Fig. 1. Detail of necrotizing granulomatous epithelioid cells surrounded organized as a palisade.

The patient was discharged with prednisone 60 mg/day and cyclophosphamide 150 vo mg/vo day. He presented systemic recurrence 4 months later, coinciding with the decrease in corticosteroid treatment.

Twelve months into the process, while in remission, we replaced cyclophosphamide for azathioprine due to its better toxicity profile and, 6 months later, for methotrexate, reaching the target of a 20 mg/week dose, gradually reducing prednisone to 12.5 mg/day, while the patient remained asymptomatic. We made the changed from azathioprine to methotrexate because of the latter's better dosaging profile upon request by the patient, to ensure compliance.

Comments

The urological manifestations of extrarenal GPA are uncommon, especially in isolation, and in most cases are associated with concomitant renal injury, or may precede the clinical manifestation of the disease, making diagnosis difficult. The prostate is the most frequently genitourinary organ involved (2%–7.4%) after the kidney, but onset with urinary symptoms is rare.[5] There is also sporadic involvement of the testicles, bladder, seminal vesicles, urethra, penis, ureter, and adrenal glands[6] (Table 1).

The diagnosis of GPA is performed by a combination of clinical and histological data. The initial and characteristic involvement of the upper and lower airways and the subsequent development of glomerulonephritis of varying severity leads the clinician toward the diagnosis. However, a biopsy of the affected organ, and visualization of lesions of necrotizing granulomatous vasculitis, is essential to confirm the diagnosis.

High titers of c-ANCA were identified in 88% of patients with active disease. The finding of high levels of c-ANCA shows a sensitivity of 41%–96% depending on the degree of activity, and a specificity of 99%, in the diagnosis of GPA.[2,7]

ANCA are a group of autoantibodies, mainly IgG, directed against antigens which are present in the cytoplasm of the neutrophil granulocytes and in the cytoplasm of monocytes, and which are particularly associated with systemic vasculitis. pANCA or perinuclear fluorescence pattern are cytoplasmic autoantibodies against myeloperoxidase (anti-MPO). c-ANCA fluorescence or cytoplasmic pattern autoantibodies are specific against serine proteinase-3 (anti-PR3).

Plasma levels of c-ANCA can be used as immunological (not biochemical) markers of the disease, although they not always have