A case report of bilateral hydroureteronephrosis associated with systemic sclerosis

Received: November 6, 1999 / Accepted: April 24, 2000

Abstract This report describes a case of atrophic bladder and bilateral hydroureteronephrosis that occurred in a patient with systemic sclerosis (SSc). A 49-year-old woman who had a 12-year history of SSc was admitted to our hospital because of bilateral hydroureteronephrosis indicated by uroflowmetric and radiological studies. Histological examination of the patient's bladder after biopsy revealed fibrotic replacement of submucosa and infiltration of mononuclear cells, but no deposition of immunoglobulins and complement components were observed. Nephrostomy to relieve the urinary retention was required. There have been few reports regarding SSc complications in hydronephrosis. The association between hydronephrosis and the pathological disorder of SSc is discussed.

Key words Systemic sclerosis · Hydronephrosis · Atrophic bladder · Nephrostomy

Introduction

Systemic sclerosis (SSc) is a connective tissue disease characterized by fibrosis of the skin and visceral organs, including lungs, gastrointestinal tract, heart, and glomeruli in the kidneys. Renal parenchymal involvement, which causes rapid reduction of creatinine clearance, azotemia, and sudden onset of severe hypertension, is a documented life-threatening symptom of SSc. In contrast, the involvement of SSc in the lower urinary tract is an uncommon manifestation. Although atrophic bladder has been reported as a chronic lower urinary tract disorder in patients with systemic lupus erythematosus (SLE), there have been few reports on the involvement of the lower urinary tract in SSc. Bilateral hydronephrosis has rarely been mentioned, and only one investigator has reported hydronephrosis associated with SSc.

Here we present a case of bilateral hydronephrosis with contracted bladder in a patient with SSc. We evaluated the lower urinary tracts disorder using radiological examinations, including intravenous pyelography (IVP), computed tomography (CT), and magnetic resonance imaging (MRI). We also performed immunohistological staining of the urinary bladder. Although immunohistological studies have frequently been performed in lupus cystitis cases, atrophic bladder in patients with SSc has not been examined in detail. In this communication, we report the results of uroflowmetric, radiological, and immunohistological examinations of atrophic bladder observed in a patient with SSc and discuss the pathogenesis of hydronephrosis.

Case report

The patient was a 49-year-old Japanese woman who was diagnosed with SSc based on Raynaud’s phenomenon, bilateral sclerodactyly, sausage-like swelling of the fingers, and the histological findings of a skin biopsy in 1987 at another hospital. She had been treated with 5mg/day prednisolone since then. She had acute myocardial infarction caused by stenosis of the 7th segment of the coronary artery, and percutaneous transarterial coronary angioplasty was performed in March 1997. From 1998, she had felt polakisuria and decreased volume of urine, which were
getting worse, and bilateral sclerodactylyia had progressed gradually. In March 1999, she was admitted to the hospital with complaints of lower abdominal pain and complete urinary retention. Although an improvement of the bilateral hydroureteronephrosis was observed within 24 h after insertion of a urinary catheter, she was referred to our university hospital on April 12, 1999, so that the disease activity of the SSc could be evaluated and the cause of the hydronephrosis examined.

On admission to our hospital, physical examination revealed bilateral sclerodactylyia, and scleroderma on the face and forearm. Neither pitting scars on the fingers nor shortening of the sublingual ligament were present. Blood pressure was 128/70 mmHg, and funduscopic findings indicated no hypertensive retinopathy. A slight fine crackle was audible at the bilateral lower back. Bowel sound was normal, and no abnormalities were found in neurological examination. Laboratory findings revealed a normal blood cell count. The serum albumin value was slightly low at 3.8 mg/dl, and the triglyceride level was mildly elevated at 158 mg/dl. The serum creatinine level was 0.6 mg/dl, and renin activity was within normal limits. Urinalysis demonstrated 1+ proteinuria and 20 red blood cells and 10 white blood cells per high-power field. Although antinuclear antibodies were positive at a titer of 1:640, anti-Scl 70, anticentromere, anti-RNP, anti-DNA, anti-SS-A, and anti-SS-B antibodies were negative.

Ultrasonography and intravenous pyelography (IVP) showed bilateral hydroureteronephrosis and engorgement of the bilateral urinary tracts, as shown in Fig. 1. An improvement of bilateral hydroureteronephrosis due to catheterization was indicated by IVP. Cystometric analysis revealed a reduced compliance of bladder with no residual urine. The maximal volume of the bladder was estimated at less than 40 ml. Right vesicoureteral reflux was observed on voiding cystography. Cystoscopy showed edematous mucosa and scattered erosion of the bladder wall. A biopsy of the bladder was performed by transurethral resection, and its histological examination revealed fibrotic replacement of submucosal tissue and infiltration of mononuclear cells, as shown in Fig. 2. However, when immunohistological staining was performed using anti-IgG, anti-IgM, anti-IgA, and anti-C3 antibody, no deposition was detected in the specimen. Computed tomography (CT) did not detect an obstruction of the ureters by extrinsic involvement such as a classic retroperitoneal fibrosis. Magnetic resonance imaging (MRI) reconfirmed no organic region around the ureters by both horizontal and sagittal views. In addition, MRI also indicated thickening and inhomogeneous intensity of the bladder wall, as shown in Fig. 3. Based on these findings, a diagnosis of obstructive uropathy was made, which was...