Sjögren's Syndrome with Hydronephrosis Caused by Pseudolymphoma

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A 67-year-old woman with Sjögren's syndrome was found to have left hydronephrosis and stenosis of the left ureter. Exploratory laparotomy disclosed a nodule at the ureteropelvic junction of the left ureter. Histopathological examinations of the biopsied specimen of this nodule showed lymphoid hyperplasia within the ureteral wall. Low dose prednisolone improved hydronephrosis and pseudolymphoma within several months. Hydronephrosis secondary to pseudo-lymphomatous infiltration of the ureter is a rare but, if properly treated, reversible complication of Sjögren's syndrome.

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Introduction

Lymphocytic infiltration may occur in many nonexocrine as well as exocrine organs in Sjögren's syndrome (SS). It may simulate clinical and histological findings of lymphoma, hence the term pseudolymphoma (1). We report a case of SS with reversible hydronephrosis secondary to pseudolymphomatous infiltration within the ureteral wall, which was responsive to a small dose of prednisolone.

Case Report

A 67-year-old woman was admitted in July 1985 because of bilateral parotid gland enlargement and xerostomia. In 1975, she noticed painful eyes and dryness of her mouth. In 1976, bilateral submandibular glands became enlarged and they were excised in a local hospital. In 1977, she noticed bilateral enlargement of preauricular lymph nodes, which were also excised. In April 1985, she developed enlargement of bilateral parotid glands and supraclavicular lymph nodes. Her mother died of renal failure at the age of 42 years and her sister died of cancer of the gastrointestinal tract at the age of 15 years.

Physical examination revealed marked enlargement of the parotid glands. The supraclavicular lymph nodes were enlarged bilaterally. Physical examination was

otherwise unremarkable.

Laboratory examinations revealed a sedimentation rate of 125 mm/h and an unremarkable urinalysis. Complete blood counts were essentially normal except for a slight normocytic, normochromic anemia. Serum immunoglobulin concentrations were IgG 5,438 mg/dl, IgA 152 mg/dl, and IgM 32 mg/dl. Serum immunoelectrophoresis did not show M components. Serum creatinine was 1.0 mg/dl, BUN 25.3 mg/dl, GPT 31 U, GOT 16 U, LDH 322 U, and ALP 7.9 U. Serum amylase was increased to 358 IU/l (normal: 60–200), of which 89% was of the pancreatic isoenzyme type. Creatinine clearance was 60.0 ml/min. C-reactive protein was absent. RA test was +-. Antinuclear antibody titer was 1:80 with a diffuse pattern. Anti-DNA, anti-SS-A, anti-SS-B, anti-RNP antibodies were negative. The levels of C3, C4, CH50 were 33 mg/dl (60–116), <5 mg/dl (15–44), <10 U (30–40), respectively.

Chest X ray films showed enlargement of the right hilar lymph nodes. Abdominal ultrasonography and computed tomography revealed left hydronephrosis, a mass at the right renal hilum, and a pancreatic cyst (Fig. 1A). Antegrade pyelography disclosed stenosis of the left ureter adjacent to the ureteropelvic junction. Lymphography revealed enlargement of paraaortic lymph nodes (Fig. 2A).

The results of Shirmer's test and rose bengal dye

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Fig. 1. Abdominal computed tomography showing a mass at the right renal hilum; left hydronephrosis in September 1985 (A). Follow-up tomography showing an improvement of these findings in September 1986 (B).

staining were compatible with the diagnosis of SS. Biopsy specimens of the minor salivary glands and the parotid glands showed marked infiltration with lymphocytes and plasma cells, and fibrosis with acinar atrophy and destruction. A supraclavicular lymph node biopsy revealed proliferation of plasma cells and immunoblasts. To definitely rule out malignant lymphoma, exploratory laparotomy was performed. An induration was palpable in the left upper ureter, which was thought to be the cause of hydronephrosis. A biopsy specimen of this induration revealed marked infiltration with lymphocytes

Fig. 3. Microscopic findings of the left ureteral wall. Marked infiltration with lymphocytes and plasma cells is shown. hematoxylin and eosin, ($\times 400$).

Fig. 2. Antegrade pyelography demonstrates stenosis of the left ureter adjacent to ureteropelvic junction in September 1985 (contrast medium for lymphography remains) (A). Intravenous pyelography shows no significant abnormality in March 1986 (B).

Fig. 4. Clinical course of the patient.

and plasma cells (Fig. 3). There was a mass encircling lymph nodes and vessels at the right renal hilum, which could not be resected. The lymph nodes adjacent to the stomach, pancreas and aorta were enlarged, soft and smooth on palpation. One of these nodes was biopsied and showed no tumor cells on examination. The pancreas was diffusely firm with a solitary cyst, a finding consistent with the diagnosis of chronic pancreatitis.

Prednisolone, 10 mg a day, was started in January 1986 and there was an appreciable decrease in the size of bilateral parotid glands within a few weeks. When she was admitted for reexamination in September 1986, parotid gland enlargement was not seen (Fig. 4). Radiographic examinations demonstrated no mass at the right renal hilum and disappearance of hydronephrosis (Figs. 1B, 2B).

Discussion

SS in this patient was considered to be complicated by pseudolymphoma because of generalized lymphadenopathy with no evidence of lymphoma on histopathological examination. Marked lymphocytic infiltration within the ureteral wall and disappearance of hydronephrosis after treatment with prednisolone suggested that hydronephrosis in this patient was secondary to the ureteral involvement by pseudolymphoma. Pseudolymphoma in SS has been reported to respond to steroid therapy (2, 3). This patient had hypocomplementemia which

improved after treatment with prednisolone (Fig. 4), although there was no evidence to prove a diagnosis of systemic lupus erythematosus except for antinuclear antibody. It has been demonstrated that the levels of circulating immune complexes (IC) and C3d are elevated in primary SS patients, and possibly reflect clinical activity (4). Hence there is a possibility that hypocomplementemia may be found in primary SS patients. IC and C3d were not measured in the present patient. Furthermore, hypocomplementemia is sometimes seen in patients with SS associated with partial lipodystrophy, membranoproliferative glomerulonephritis, lymphoreticular neoplasms, or vasculitis (5). These complications, however, were absent in the present case. The clinical significance of hypocomplementemia in SS remains to be clarified.

Renal tubular defects and glomerulonephritis are known as renal involvement in SS (5). But a search of the literature failed to reveal a single case resembling the present case (2, 6, 7). On the other hand, ureteral obstruction secondary to retroperitoneal nodal enlargement is not an uncommon complication of malignant lymphoma (8, 9). Nevertheless the uretral intreamural lesion similar to that seen in the present case is unusual even in malignant lymphoma. There is a single report of Hodgkin's disease of the ureter with hydronephrosis (10). The patient in that report had an isolated lesion which developed within the ureteral wall near the ureteropelvic junction like the present case, and the cross-section of the ureter showed tumor cells primarily involving the muscular and submucosal layers. In general, lymphoid tissue is seen in the lamina propria of the ureter (11), so that there is a possibility that ureteral involvement may develop in lymphoproliferative disorders.

This is the first report of a case of SS with hydronephrosis caused by pseudolymphoma. As progression from pseudolymphoma to malignant lymphoma has been reported in SS (3), this patient requires close follow-up.

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