A case of Behçet’s disease associated with aortic regurgitation and nephrotic syndrome

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Abstract—A 50-year-old housewife with chief complaint of dyspnea is reported. In 1975, the patient developed aphthous stomatitis, genital ulcers, uveitis and erythema nodosum, and was diagnosed as having Behçet’s disease at the Department of Dermatology of our hospital. Oral prednisolone was started in May 1985. Proteinuria was first detected in April 1987. The patient developed orthopnea in July 1992 and was admitted to our hospital in September of that year. Her past history revealed hyperlipidemia since January 1984 and a diagnosis of aortic regurgitation (AR) was made in 1984. Physical examination revealed a systolic/diastolic murmur in the chest and pretibial edema. Laboratory findings showed proteinuria (3.8 g/day) and hypoproteinemia. Microscopic findings of renal biopsy revealed mesangial proliferative glomerulonephritis and arteriosclerosis. Immunofluorescent studies demonstrated deposits of Apo B in the basement membrane and the mesangium. She was discharged from our hospital as proteinuria was decreased after combination treatment with camostat mesilate 600 mg/day and sairei-to 9.0 g/day. Hypertension and hyperlipidemia appeared to have acted as aggravating factors for renal lesions caused by Behçet’s disease, so that our patient developed the nephrotic syndrome. We report a rare case of Behçet’s disease complicated by aortic regurgitation and the nephrotic syndrome.

Key words: aortic regurgitation; Behçet’s disease; hyperlipidemia; nephrotic syndrome.

INTRODUCTION

Behçet’s disease is a multisystem disorder presenting with recurrent oral and genital ulcerations as well as ocular involvement. Renal and cardiac involvement is relatively rare in Behçet’s disease. We report a case of Behçet’s disease associated with aortic regurgitation and nephrotic syndrome.
CASE REPORT

The patient was a 50-year-old housewife with a chief complaint of orthopnea. In 1975, the patient developed aphthous stomatitis, pudendal ulcers, vitreous body opacities, uveitis and erythema nodosum. Diagnosis of Behçet’s disease was made at the Department of Dermatology in our hospital and she was subsequently followed up by the same department. The patient developed chest pain in June 1984 and her local physician suspected angina pectoris. When she was referred to our department in September 1984, cardiac catheterization was performed and diagnosis of aortic regurgitation (AR; Sellers grade IV) complicated with Behçet’s disease was made. At that time, her systolic blood pressure was elevated and proteinuria was 2+. In May 1985, she suffered from oral ulcers, pudendal ulcers, folliculitis and frequent attack of chest pain, and oral prednisolone (PSL; 20 mg/day) was started. The oral ulcers, pudendal ulcers and folliculitis improved, and the episode of chest pain decreased. From January 1988, the urinary protein level was above 300 mg/dl (urine sediment was normal) and she was admitted to our department from November to December of that year. The proteinuria resolved after rest and she was discharged. However, proteinuria greater than 300 mg/dl recurred in February 1989 and the patient began to suffer from orthopnea in July 1992. She was admitted to our department on September 14 of that same year. Her past history revealed hyperlipidemia since January 1984 and a diagnosis of AR in 1984.

Physical examination revealed the following: height 156 cm, weight 73 kg, BP 166/0 mmHg, PR 80/min. The palpebral conjunctiva revealed anemia. No aphthous ulcers were detected in the mouth.

A systolic/diastolic murmur (Levine V/VI) which radiated to the neck was heard in the chest. Breath sounds were normal and there were no rales. The abdomen was flat, soft, non-tender and no hepatosplenomegaly was detected on palpation. Edema was present in the lower extremities. No pudendal ulcers or erythema nodosum were observed. The pinprick reaction was negative and neurological examination was unremarkable.

On admission, urinalysis showed the following: protein 3.8 g/day; occult blood 3+; sediment 5–10 RBC/high-power field and 10–20 granular casts/whole visual field. The urinary NAG level was 107.6 IU/l. A complete blood count was as follows: WBC 8730/mm³; RBC 389 × 10⁶/mm³; Hb 10.1 g/dl; Ht 32.1%; Plts 36.1 × 10⁴/mm³. Biochemical studies showed an elevated LDH level (432 IU/l) and hypoproteinemia (serum total protein level was 5.1 g/dl and serum albumin level was 3.1 g/dl). Serum creatinine was 0.9 mg/dl and urea nitrogen was 10 mg/dl, while the uric acid level was slightly elevated at 6.5 mg/dl. The erythrocyte sedimentation rate was 21 mm/h and CRP was 0.42 mg/dl. Immunoglobulin and complement levels were as follows: IgG 726 mg/dl; IgA 196 mg/dl; IgM 49 mg/dl; C₃ 78.6 mg/dl; C₄ 36.6 mg/dl; CH₅₀ 37.4 U/ml. She was negative for antinuclear antibody, anti-DNA antibody and anti-RNP antibody, while immune complexes were below 1.5 µg/ml and haptoglobin was below 20.6 mg/dl. Both the direct and indirect Coombs’ tests were negative.