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Conn's Syndrome Associated with Behçet's Disease

A 39-yr-old woman, who had been treated for Behçet's disease for 4 yr, was admitted for further investigation of recently identified hypokalemia and hypertension. Suppressed plasma renin activity with elevated plasma aldosterone concentration and an anomalous postural decrease in plasma aldosterone were observed. An abdominal CT scan revealed a right adrenal mass. The patient was diagnosed with Conn's syndrome. The association of Conn's syndrome with Behçet's disease was thought to be coincidental. To our knowledge, this is the first case of Conn's syndrome associated with Behçet's disease.

Key Words: *Adenoma, Adrenal Cortical; Hyperaldosteronism; Behçet Syndrome*

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INTRODUCTION

Behçet's disease (BD) is a multisystem disorder that presents with recurrent oral and genital ulcerations as well as ocular lesions (1). Primary hyperaldosteronism, an uncommon but important cause of secondary hypertension, is confirmed by demonstrating elevated aldosterone levels in the presence of suppressed plasma renin activity (PRA) (2). Conn's syndrome, which is caused by an aldosterone-producing adenoma, is the most frequent cause of primary hyperaldosteronism and accounts for approximately 70% of such cases. The occurrence of BD in association with neoplasms is very rare but has been reported (3-12). We report here an unusual association of BD and Conn's syndrome in a middle-aged Korean woman. We believe this to be the first report of such a case.

CASE REPORT

The patient was a 39-yr-old woman, who had been troubled by recurrent oral and genital ulcers and erythema nodosum-like skin lesions since 1994. She had also occasionally experienced localized pain in the right lower quadrant of the abdomen, and was diagnosed with intestinal BD in 1997. At that time, she had a positive pathergy test and posterior uveitis. She had been treated with prednisolone, sulfasalazine, and colchicine. During follow-up in our outpatient clinic, she suddenly developed hypertension and hypokalemia, and on December 11, 2000, was admitted to Chonnam National University Hospital for further investigation.

On admission, her blood pressure was 160/120 mmHg, pulse rate was 70/min, body temperature was 36.5℃, and respiration rate was 20/min. On physical examination, the patient appeared chronically ill. The patient's laboratory findings were as follows: hemoglobin 10.5 g/dL, white blood cell count 5,400/μL, platelet count 243,000/μL, C-reactive protein 0.336 mg/L, erythrocyte sedimentation rate 4 mm/hr, rheumatoid factor 10.5 IU/mL, C3 92.8 mg/dL, and C4 11.8 mg/dL. Liver function tests were normal, and a test for antinuclear antibody was negative. Serum sodium was 139 mEq/L, potassium was 3.0 mEq/L, chloride was 105 mEq/L, blood urea nitrogen was 8.4 mg/dL, and creatinine was 0.9 mg/dL. A 24-hr urine specimen showed that her urinary potassium was 52.8 mEq/day and her urinary sodium was 123 mEq/day. Postural studies revealed suppressed peripheral PRA of 0.53 ng/mL/hr and an elevated aldosterone level of 39.3 ng/dL with an aldosterone/PRA ratio of 68 in the supine position. After standing for four hours, her PRA decreased to 0.27 ng/mL/hr, and her aldosterone level decreased to 31.2 ng/dL. Serum cortisol levels that were measured at the same time were 4.5 μg/dL supine and 8.6 μg/dL standing. The plasma ACTH concentrations were 12.8 pg/mL and 6.8 pg/mL in the supine and standing positions, respectively. The levels of urinary catecholamines and their metabolites were normal. An abdominal ultrasonogram revealed a right adrenal mass measuring 1.5 × 2.5 cm. The presence of the mass was confirmed by computed tomography (Fig. 1).

On February 5, 2001, the patient underwent an abdominal

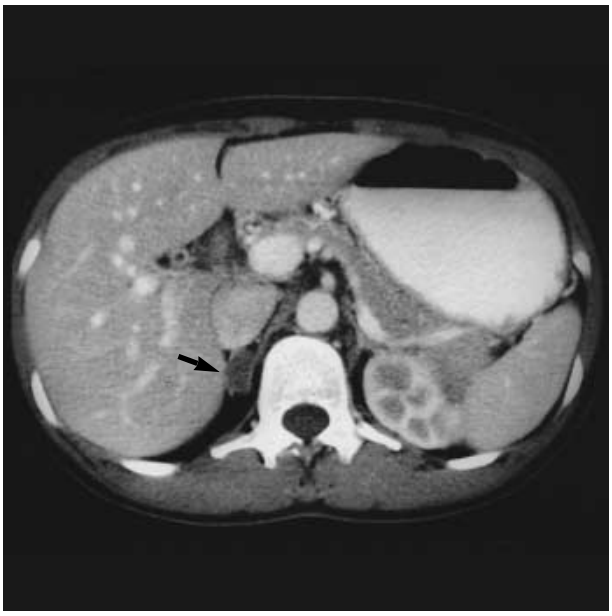

Fig. 1. Computed tomographic scan of the abdomen showing a right adrenal mass (arrow).

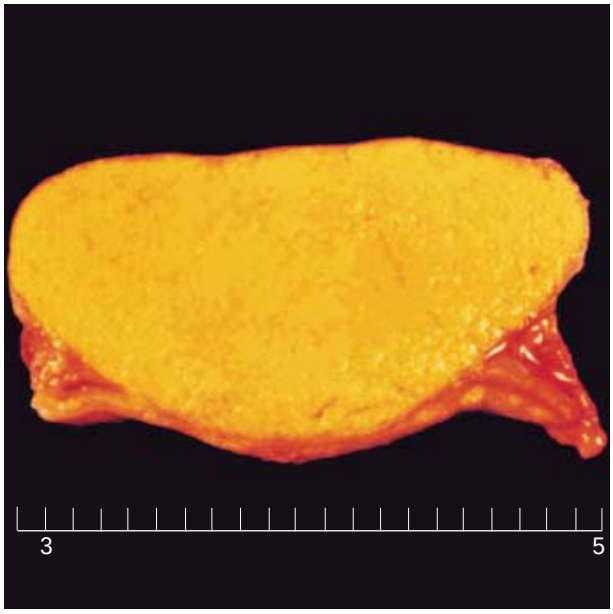

Fig. 2. The cut surface of the right adrenal mass shows a golden-yellow color.

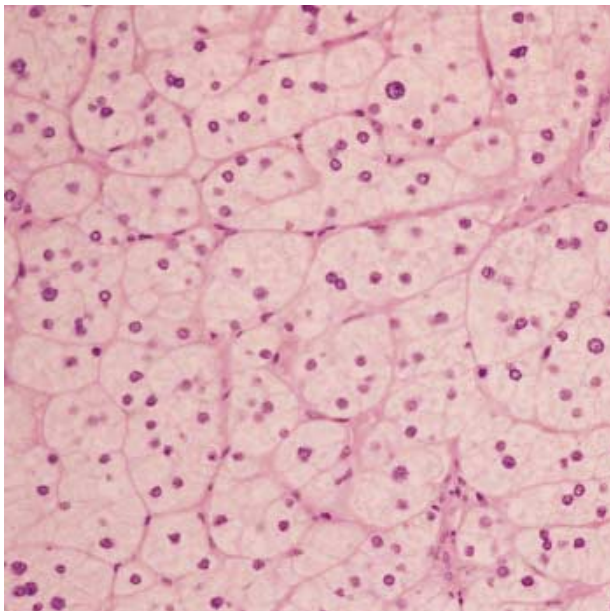

Fig. 3. Histologic appearance of the mass. The tumor is composed of large, clear, and finely granular cells in lobular arrangements (H &E stain; × 400).

exploration. A right total adrenalectomy was performed, with removal of a 1.5 × 1 × 2 cm right adrenal mass that weighed 10 g. On gross examination, the mass was golden-yellow in color and showed no evidence of necrosis or hemorrhage (Fig. 2). Histologically, the tumor was composed of large, clear, and finely granular cells in lobular arrangements that were divided by vascular septae (Fig. 3). The pathological diagnosis was benign adrenocortical adenoma. The combined biochemical and pathological profile of the patient was consistent with that of Conn's syndrome.

Postoperatively, the patient was normotensive, and the serum potassium was 3.7 mEq/L. The patient was discharged on 12 February 2001 and was followed at the outpatient clinic. Follow-up at 12 months showed that the patient remained normotensive and normokalemic.

DISCUSSION

BD is endemic in the Middle East and the Mediterranean region (1). It also occurs in Central and East Asian countries, such as Korea, Japan, and China. The incidence of BD in Korea has been increasing (13). The etiology of BD remains unclear, but genetic and environmental factors probably play a role in its pathogenesis.

There have been several reports on concurrent neoplasms in BD (3-12). However, as far as we know, no association between BD and Conn's syndrome has ever been reported. Previous case reports provide no documented pathophysiological mechanisms for the development of tumors in BD patients, other than possible drug-related etiologies or the autoimmune nature of BD itself. Harada et al. (8) suggested that long-term administration of colchicine in BD patients may induce non-Hogkin's lymphoma, because of colchicine's immunosuppressive properties. Abe et al. (9) attributed the transformation of lymphoma to the immune disturbances of T-helper cells, neutrophils, and macrophages in BD. However, the authors believed that most cases of solid tumors were incidental.

Aldosterone-producing adenoma has been recognized as being second only to renovascular disease as a cause of surgically correctable hypertension (2). In our patient, the presence of hypertension, hypokalemia resulting from inappropriate kaliuresis, suppressed peripheral PRA, increased aldosterone excretion, and a plasma aldosterone/PRA ratio greater than 30 was consistent with primary aldosteronism (2). Postoperatively, our patient has remained normotensive and normokalemic.

There have been some reports of high surgical morbidity in BD. Sayek et al. (14) reported increased surgical morbidity in seven cases of intestinal BD. Paccagnella et al. (15) reported a worsening of BD symptoms after surgery, and fatal infection due to impaired immunity. However, our patient did not experience any complications.

In conclusion, there is no clear evidence that autoimmune disorders or immunosuppressive drugs cause epithelial cancer or adenoma, and therefore, we consider our case to be a sporadic occurrence of Conn's syndrome concurrent with BD.

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