Acquired generalized anhidrosis associated with lymphocytic infundibuloneurohypophyseitis

M. Asahina, MD; A. Suzuki, MD; N. Tamura, MD; T. Yoshida, MD; and T. Hattori, MD

Acquired idiopathic generalized anhidrosis (AIGA) has an acute or insidious onset without additional symptoms. There may be eccrine gland dysfunction, and the beneficial effects of corticosteroids lend support to an autoimmune etiology. Lymphocytic infundibuloneurohypophyseitis (LIN) is an inflammatory disorder that can result in central diabetes insipidus (DI); histologic findings showing infiltration of lymphocytes suggest an autoimmune etiology. We describe a case of AIGA associated with LIN.

Case report. A 25-year-old man was in good health until December 1999, when he noticed a cutaneous eruption with itching in the trunk and arms after physical exercise or exposure to a hot environment, accompanied by generalized anhidrosis. He also experienced frequent thirst and increased fluid consumption. There was no history of syncope, impotence, or urinary symptoms. On his first admission in July 2001, general physical examination revealed normal findings except for dry skin. On neurologic examination, pupils were equal and reacted to light well. Muscle strength, tone, and muscle stretch reflexes were normal in all extremities. Sensory system was intact. The patient’s blood pressure was 135/80 mm Hg in the supine position and 138/84 mm Hg at 70° of head-up tilt. The temperature was intact. The patient’s blood pressure was 135/80 mm Hg in the supine position and 138/84 mm Hg at 70° of head-up tilt. The coefficient of variation of R-R intervals was within normal limits. A 2-hour urine output was 7 to 10 L/d. Blood tests, including electrolytes and anterior pituitary hormone levels (growth hormone, adrenocorticotrophic hormone, prolactin, luteinizing hormone, follicle-stimulating hormone, and thyroid-stimulating hormone), were normal. Urinary and plasma osmolarity were 65 and 278 mOsm/kg with oral free intake. The baseline level of plasma antidiuretic hormone (ADH) was 0.7 pg/mL (normal range, 0.3 to 3.5 pg/mL). During the water deprivation test, urinary osmolarity was persistently low (urine, 145 mOsm/kg; plasma, 268 mOsm/kg), and the plasma ADH level did not increase (0.9 pg/mL). Subcutaneous injection of pitressin led to increased urinary osmolarity (315 mOsm/kg). MRI showed thickening of the pituitary stalk, lack of hyperintense signal of the posterior pituitary gland, and destruction, whereas others have normal findings. The clinical findings in our patient were characteristic of AIGA. Our patient also had central DI with thickening of the pituitary stalk, suggesting LIN. Two cases of AIGA and central DI have been reported, but they did not show thickening of the pituitary stalks. However, MRI was examined 10 years after onset of DI in one case, and onset of DI was unidentified in the other. Although thickening of the pituitary stalk is an important finding in diagnosing LIN, it may spontaneously disappear within several years of onset. AIGA is a rare disease. Although three reported AIGA cases with DI, including our case, indicate a relatively high incidence of DI in AIGA patients, our case associates AIGA with LIN, both of which are likely autoimmune diseases.

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From the Department of Neurology (Drs. Asahina, Suzuki, Tamura, and Hattori) and Second Department of Internal Medicine (Dr. Yoshida), Chiba University Graduate School of Medicine, Japan.

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Address correspondence and reprint requests to Dr. Masato Asahina, Department of Neurology, Chiba University Graduate School of Medicine, 1-8-1 Inohana, Chuo-ku, Chiba 260-8670, Japan; e-mail: asahinam@hs.chiba-u.ac.jp

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