Acquired idiopathic generalized anhidrosis in a young Austrian patient

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INTRODUCTION

Acquired idiopathic generalized anhidrosis (AIGA) is a rare disease with approximately 100 reported cases worldwide, many of which occurred in Asia, especially in Japan.[1]

To diagnose AIGA, other causes of anhidrosis (medications; physical agents; and dermatologic, neurologic, metabolic, and endocrine disorders), must be ruled out.

Three subtypes of AIGA with different pathologic conditions can be distinguished: sweat gland failure, sudomotor neuropathy, and idiopathic pure sudomotoric failure.[1,2] We report a case of AIGA in an Austrian patient. At present, only a few cases have been reported in Europe[3-5] and the United States.[6-8]

Abbreviations used:

AIGA:	acquired idiopathic generalized anhidrosis
IPSF:	idiopathic pure sudomotoric failure
QSART:	quantitative sudomotor axon reflex test

CASE REPORT

A 30-year old, white, athletic man presented with progressive lack of body sweat production, pruritus, and urticaria. During exercise or rest, when surrounding temperatures were >24°C, he felt dizzy and weak, overheated, and required more time to recover after exertion. The frequency of his physical training sessions was reduced because of his symptoms.

The patient's medical and family history were inconspicuous, and previous treatment with any medication was denied. Dermatologic, neurologic, and internal clinical statuses were normal. Cranial and spinal magnetic resonance imaging did not indicate any neurologic disorders. Cardiovascular autonomic reflex screening revealed postural orthostatic tachycardia syndrome and cardioinhibitory reflex syncope.

Laboratory examinations (total and differential blood count, electrolytes, renal and liver function tests, blood glucose level, thyroid stimulating hormone, serum immunoglobulin E level, and carcinoembryonic antigen) were all within the normal ranges. No autoantibodies to SS-A/Ro, SS-B/La, and ganglionic $\alpha 3$ acetylcholine receptors were detected. Biochemical analysis revealed normal enzyme activity of α-galactosidase, ruling out Morbus Fabry. Biopsies from affected areas on the dorsum of the hands and the abdomen showed intact sweat glands with slight $CD3^+$ lymphocytic infiltration.

Anhidrosis was confirmed with a qualitative thermoregulatory sweat test (Minor test) combined with thermography and the quantitative sudomotor axon reflex test (QSART). The Minor test confirmed generalized anhidrosis with sweat production only on palms and armpits (Fig 1, *A*, *C*, *E*, and *G*). QSART showed a severe sudomotoric function deficiency without sweat production (Fig 1, *K*).

The diagnostic procedures we used ruled out other causes of anhidrosis and confirmed AIGA, subtype idiopathic pure sudomotor failure (IPSF).

After receiving three cycles of high dose methylprednisolone (1 g on each of 3 consecutive days

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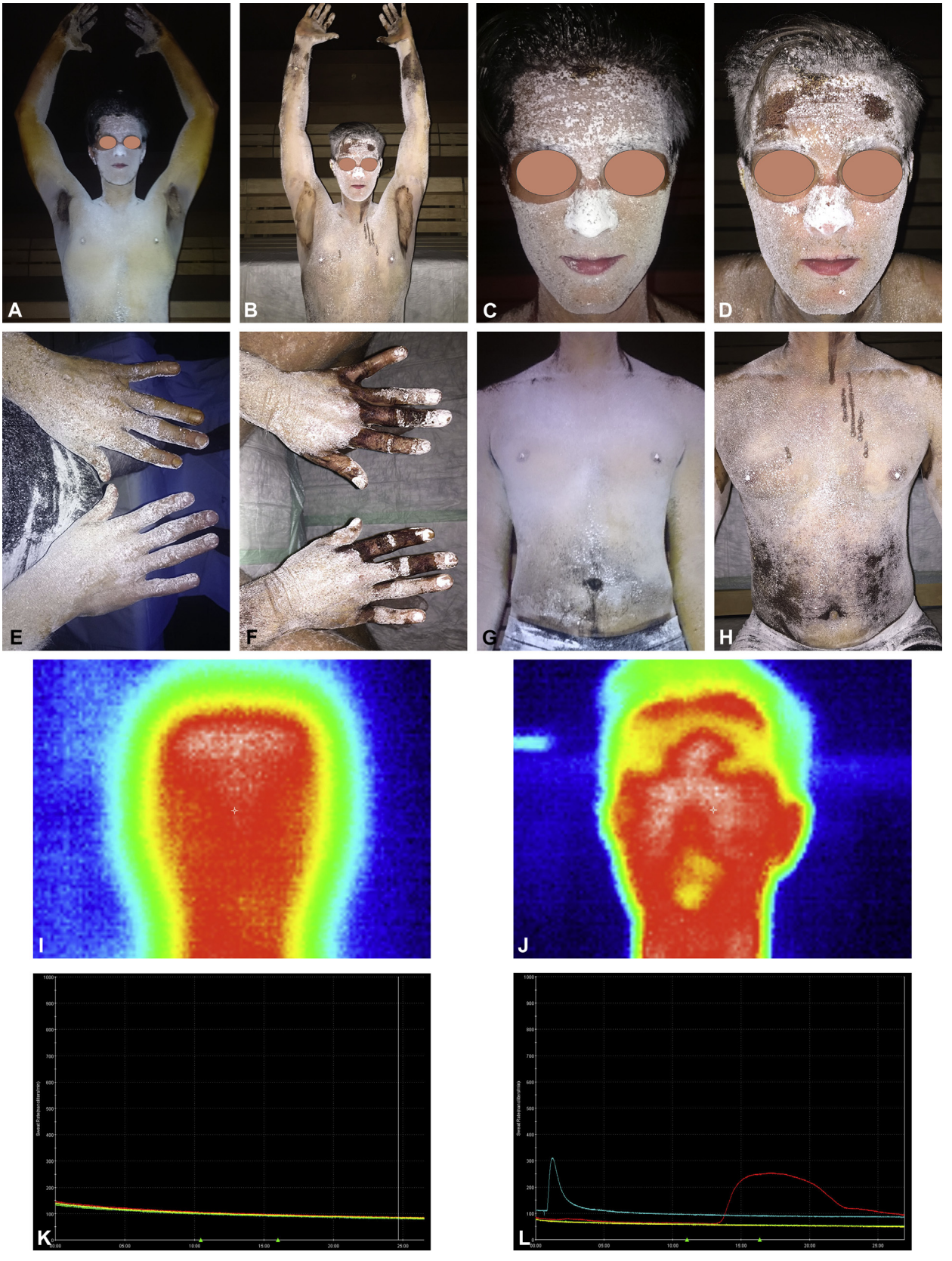
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