Acquired Idiopathic Generalized Anhidrosis: Case Report

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We report a case of acquired idiopathic generalized anhidrosis (AIGA) in a 56-year-old white woman. Acquired idiopathic generalized anhidrosis is an exceedingly rare group of heterogeneous disorders that has been almost exclusively reported in young Japanese males. Our case is unique in that AIGA may be underrecognized in this patient population.

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Acquired idiopathic generalized anhidrosis (AIGA) was first described in 1917 by Lutembacher.[1] It is an exceedingly rare heterogeneous group of disorders characterized by an episodic burning sensation and erythema associated with decreased or absent sweating. Most cases of AIGA have been described in young Japanese males.[2-6] We report a case of AIGA in a 56-year-old white woman.

Case Report

A 56-year-old white woman presented with an acute cutaneous burning sensation upon heat exposure of 10 years' duration that was associated with decreased sweating, elevated body temperature, and diffuse erythema. The phenomenon initially involved her extremities and then progressed to involve her trunk and face. Symptoms markedly improved with cool showers and decreased body temperatures. Gabapentin 300 mg every 8 hours and a 4-week course of prednisone 40 mg daily provided no relief, while oral triamcinolone caused an acute increase in symptoms. A complete review of systems revealed no abnormalities. Her medical history included hyperlipidemia and hypertension. At the time of onset, she was not taking any medications or supplements. Her current medications included lisinopril, conjugated estrogens, and atorvastatin. The result of a full skin examination revealed no abnormalities. A starch-iodine test revealed minimal axillary hidrosis and complete absence of sweating elsewhere. Her workup included a complete blood cell count as well as a renal, liver, and metabolic panel; all results were normal. An electromyogram was normal. Magnetic resonance imaging of the spine revealed multi-level cervical spondylosis with mild midcervical compression. The result of a complete neurologic evaluation, however, was negative, and it was determined that the compression was not related to her symptoms. Skin biopsy revealed a slight decrease in the number of eccrine coils with a few adjacent lymphocytes.

Comment

Acquired idiopathic generalized anhidrosis is an exceedingly rare group of heterogeneous disorders that clinically manifest as diffuse progressive loss of sweating. The typical patient is a young and otherwise healthy male, occurring almost exclusively in Japanese males.[2-5,7] In general, heat exposure produces an acute elevation of body temperature as well as a burning stinging sensation of the skin associated with bright erythema and a lack of or marked decrease in sweating. Many patients also have associated cholinergic urticaria and increased IgE levels.[2,3,5,6] Few cases have occurred immediately after heat stroke.[8,9] A minority of the patients may have a progressive insidious onset of symptoms.[6]

Several histopathologic findings have been observed in AIGA, further contributing to the heterogeneous nature of the disorder. The majority of cases demonstrate with normal-appearing eccrine glands; some cases have eccrine gland atrophy, periecrine lymphocytic or mast cell infiltrates, irregular eccrine arrangements within the dermis and subcutaneous tissues, or rarely poral occlusion.[3-7,10,11]

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In cases reported following heat stroke, electron microscopy often revealed vacuolar changes in the cytoplasm of eccrine secretory cells, which was sometimes appreciated on light microscopy.[6,8,9] In some cases, immunohistochemistry studies have shown changes in membrane glycoproteins, such as decreased lectin binding.[12] The significance of this observation is not known.

The role of neurotransmitters has been evaluated. Eccrine glands primarily are innervated by postganglionic cholinergic sympathetic nerves. Although few patients with AIGA have been found to have a decrease in terminal synapse densities, most have normal neural density and activity.[3,13] However, several case studies have demonstrated a lack of response to cholinesterase agonists such as pilocarpine, suggesting that the postsynaptic muscarinic receptors may be at fault.[2,3,14]

Given the heterogeneous pathobiologic, clinical, and histopathologic findings in AIGA, attempts have been made to further classify this disorder into 3 subgroups: idiopathic pure sudomotor failure, sudomotor neuropathy, and sweat gland failure. Idiopathic pure sudomotor failure represents the vast majority of cases and includes patients thought to have cholinergic transmission and reception deficits. Sudomotor neuropathy reflects postganglionic cholinergic dysfunction, and sweat gland failure refers to the small population of patients with inflammation and degeneration of sweat glands on histopathology.[15,16]

Acquired idiopathic generalized anhidrosis often is resistant to treatment. Some patients respond to short courses of prednisone, while some cases spontaneously resolve; however, most cases are chronic.[10,17] Treatment is aimed at symptomatic relief and decreasing the core body temperature with the application of cool water and avoidance of overheating.[6] Our patient adapted to her environment by avoiding high ambient temperatures and spraying her skin with cold water to maintain a normal core body temperature.

Acquired anhidrosis has occurred in association or conjunction with several conditions including hereditary metabolic disorders such as Fabry disease, diabetes mellitus, and Hodgkin lymphoma; as part of paraneoplastic syndromes, Sjögren disease, and other connective tissue diseases; lymphocytic infundibuloneurohypophysitis, an autoimmune disorder resulting in neurohypophyseal dysfunction; drugs; and other causes of neuropathy.[18-25] Therefore, a complete history and workup should be done to rule out other conditions before diagnosing AIGA.

Conclusion

Acquired idiopathic generalized anhidrosis is a rare group of heterogeneous disorders reported almost exclusively in young Japanese males. We report a white woman with AIGA.

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