CASE REPORT

Acquired idiopathic generalized anhidrosis.

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ABSTRACT

Acquired idiopathic generalized anhidrosis (AIGA) is a rare disorder characterized by the inability to sweat in the absence of identifiable dermatological, neurological, or metabolic causes. We present the case of a 23-year-old woman whose symptoms began at the age of 8 years, with the progressive development of generalized anhidrosis associated with heat intolerance, pruritus, and erythematous cutaneous lesions exacerbated by elevated temperatures. Physical examination revealed erythematous to violaceous macules, papules, and plaques involving the trunk and axillary regions. The Minor starch-iodine test was negative, confirming the absence of sweating. Histopathological findings demonstrated preserved cutaneous architecture with structurally normal eccrine glands, although ductal degenerative changes and a mild lymphocytic infiltrate were observed.

AIGA represents a diagnostic challenge because of its low prevalence and clinical overlap with other disorders. Its pathophysiology is heterogeneous and includes neuropathic, glandular, and immunological mechanisms. Early recognition is essential to prevent complications such as hyperthermia and heat stroke. This case highlights the importance of clinicopathological correlation in the diagnosis of AIGA and contributes to expanding the available evidence regarding this uncommon condition.

INTRODUCTION

Acquired idiopathic generalized anhidrosis (AIGA) is a rare disorder characterized by systemic anhidrosis or hypohidrosis in the absence of dermatological, metabolic, or neurological causes.[1,2] In simple terms, anhidrosis refers to the failure of sweat production in response to physiological, thermal, or chemical stimuli.[3]

As of 2017, according to the Japanese guidelines for the diagnosis and treatment of AIGA, there was no specific definition or published epidemiological information regarding this condition. Consequently, prevalence and morbidity data remain unavailable, and the only published information consists of approximately 100 case reports, the majority originating from Japan.

AIGA is more common in men, accounting for more than 80% of reported cases, and typically presents during the second to fourth decades of life.[2]

From a pathophysiological perspective, three mechanisms have been proposed to explain the development of this condition: (a) sudomotor neuropathy, (b) idiopathic pure sudomotor failure, and (c) sweat gland failure. Recent studies suggest the existence of peripheral resistance to cholinergic stimulation associated with decreased expression of muscarinic M3 receptors in eccrine glands. Furthermore, the association with elevated immunoglobulin E levels, cholinergic urticaria, and favorable responses to immunosuppressive therapy support an immunological mechanism.[4,5]

AIGA continues to be an underreported condition due to underdiagnosis, its clinical variability, and its association with other dermatological disorders. This highlights its relevance for this journal and the scientific community. In this context, we present the following clinical case illustrating the clinical and histopathological characteristics of this uncommon disorder, along with a brief review of the literature.

CASE REPORT

A 23-year-old woman with no relevant past medical history or known allergies presented with symptoms that began at the age of 8 years. The patient reported normal sweating until that age, after which she noticed a progressive decrease in perspiration, eventually developing generalized anhidrosis.

Physical examination revealed diffuse erythema involving the abdomen, accompanied by erythematous papules on the chest and the inner aspects of the arms and forearms (Figure 1A). Additionally, frontal erythema associated with erythematous papules on the face was observed (Figure 1B), with lesions worsening during exposure to elevated temperatures. The back demonstrated ill-defined erythematous papular lesions (Figure 1C), while erythematous to violaceous plaques were present in the axillary regions (Figure 1D). Associated symptoms included pruritus

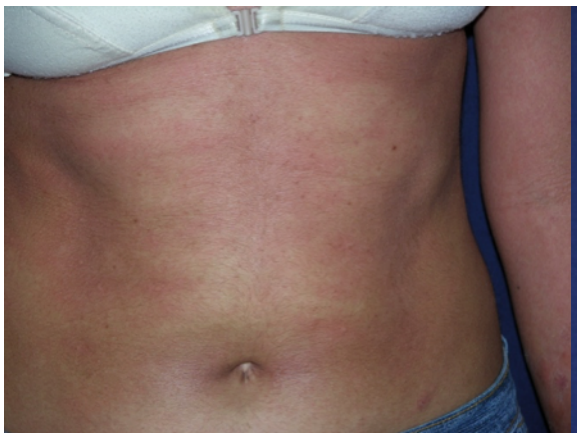


Figure 1A.Clinical presentation of the abdomen.



Figure 1B.Clinical presentation of the face.



Figure 1C. Clinical presentation of the back.

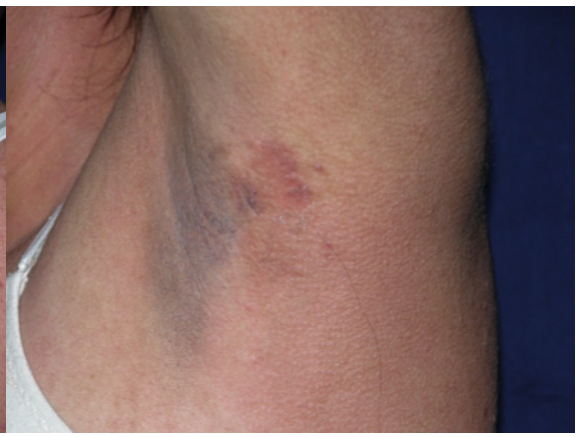


Figure 1D.Clinical presentation of the axillary region.

involving the axillary and dorsal back regions, as well as a burning sensation affecting the thorax and abdomen. The patient also reported exercise intolerance and avoided physical activity due to a sensation of overheating.

The Minor starch-iodine test performed in the axillary region was negative, demonstrating the absence of sweating in this area (Figure 2).



Figure 2. Negative Minor starch-iodine test.

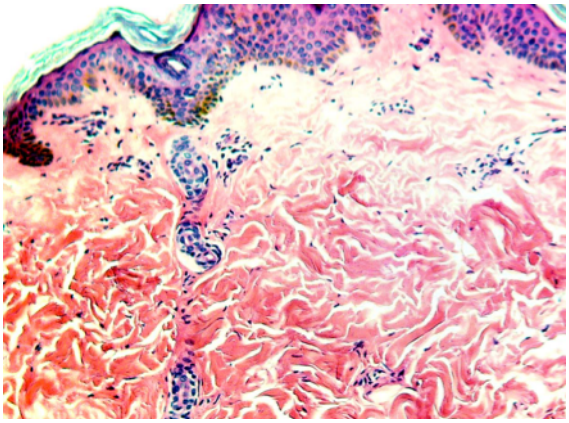


Figure 3A. Left arm biopsy showing eccrine glandular structures with preserved architecture.

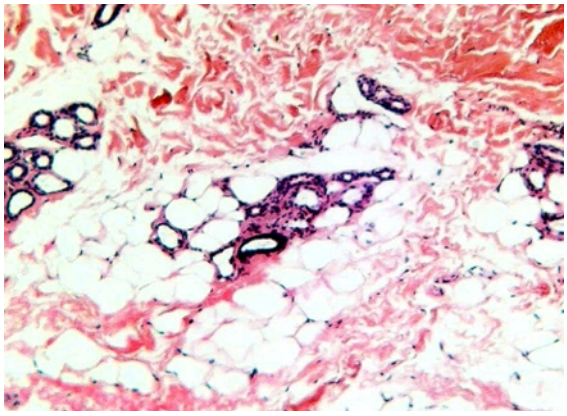


Figure 3B. Eccrine glandular structures showing degenerative changes and associated inflammatory infiltrate.

Skin biopsies were performed from the left arm and the left axillary region. The arm specimen demonstrated preserved epidermal and dermal architecture, a mild superficial perivascular lymphocytic infiltrate, and eccrine ducts with normal morphology (Figure 3A). The axillary biopsy revealed preserved cutaneous architecture, with a mild superficial perivascular lymphocytic infiltrate and structurally preserved eccrine glands. At the ductal level, a single eccrine duct showed degenerative changes characterized by cellular swelling, cytoplasmic eosinophilia, apoptosis, and karyorrhexis, associated with a mild lymphocytic infiltrate (Figure 3B).

DISCUSSION

Anhidrosis is defined as the failure to produce sweat in response to physiological, thermal, or chemical stimuli.[3] Acquired idiopathic generalized anhidrosis (AIGA) is a rare disorder characterized by the inability to sweat in the absence of an identifiable cutaneous, metabolic, or neurological cause. This condition typically affects young men and significantly impairs quality of life. When exposed to elevated temperatures, patients with AIGA are particularly susceptible to heat stroke and may experience symptoms such as malaise, hyperthermia, dizziness, palpitations, syncope, and ultimately loss of consciousness due to impaired thermoregulation. The true incidence, prevalence, and morbidity of the disease remain unknown.[3,6] AIGA is considered an uncommon disorder, with only approximately 100 reported cases to date. Most reported patients have been of Asian ethnicity, although the condition is not exclusive to this population, and therefore the incidence and clinical characteristics in other countries remain uncertain. Alternatively, patients may be misdiagnosed with other conditions, such as cholinergic urticaria associated with anhidrosis or atopic dermatitis associated with anhidrosis. Consequently, only a small proportion of actual cases are ultimately recognized and diagnosed.[7]

Regarding its pathophysiology, three principal mechanisms have been identified: sudomotor neuropathy, idiopathic pure sudomotor failure (IPSF), and primary sweat gland failure.[5,7] In IPSF and in the early stages of glandular failure, cutaneous sympathetic activity is

generally preserved or increased, suggesting peripheral resistance to cholinergic stimulation rather than a central defect.[7] This phenomenon has been associated with decreased expression of muscarinic M3 receptors in eccrine glands. The frequent elevation of IgE levels, coexistence with cholinergic urticaria, and favorable response to corticosteroid therapy support an autoimmune mechanism, possibly involving anti-M3 autoantibodies. In advanced glandular failure, irreversible structural damage predominates, resulting in a more prolonged clinical course and poorer therapeutic response, reinforcing the concept of AIGA as a disorder with a heterogeneous pathophysiological spectrum.[1,7]

Patients with AIGA fail to produce adequate sweat in response to heat-related stimuli such as elevated temperatures, humidity, and physical exercise. Anhidrosis or hypohidrosis usually affects the trunk with a symmetrical distribution.[8,9] The extremities and occasionally the face may also be involved, whereas the palms, soles, and axillae are less frequently affected. Approximately 30–60% of patients complain of tingling sensations and/or wheals under circumstances that would normally induce sweating. Additionally, patients may experience flushing, hyperthermia, weakness, fatigue, facial erythema, nausea, vomiting, headache, dizziness, or palpitations.[8] Some patients may develop heat stroke. Stinging pain and the cutaneous eruption of cholinergic urticaria are also commonly observed.[3,6]

The diagnosis of AIGA is based on clinical and functional criteria. It is defined by generalized hypohidrosis or anhidrosis affecting more than 25% of the body surface area, in the absence of dysautonomia or any generalized disorder other than sudomotor dysfunction.[7,10] The thermoregulatory sweat test constitutes the cornerstone of diagnosis and was employed in our patient.

Currently, no specific treatment algorithm exists for this condition, and therapeutic options remain limited. Reported treatments include corticosteroids, pilocarpine, intravenous immunoglobulin, and cyclosporine, with variable results. Several studies have reported successful outcomes using high-dose systemic corticosteroids in selected patients. Although steroid pulse therapy has been adopted for the treatment of AIGA, evidence supporting its efficacy remains limited, resulting in only moderate recommendations regarding therapeutic regimens.[4,8] Several reports have demonstrated substantial efficacy of corticosteroids in improving sweat gland function; however, complete treatment failure and high recurrence rates have also been documented in some patients.[5]

The efficacy of oral cyclosporine has been described in the treatment of refractory AIGA. Multiple case reports and case series have reported favorable clinical response rates in a significant proportion of patients, particularly when treatment is initiated during the early stages of the disease.[7]

Pilocarpine has been evaluated in small case series as an adjunct to immunosuppressive therapy (corticosteroids or cyclosporine) or in patients with mild-to-moderate AIGA. Findings from some studies suggest that steroid pulse therapy and oral pilocarpine improve AIGA through different mechanisms: corticosteroids increase both the extent and amount of sweating, whereas pilocarpine increases sweat production only in areas with preserved hydrotic function.[10]

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