

Review Article

Generalized Anhidrosis: A Rare Dermatological Condition

L Khondker¹, A Sultana²**Abstract:**

Anhidrosis is the inability of the body to produce and /or deliver eccrine sweat to the skin surface in response to an appropriate stimulus or environment. It may be due to changes in the eccrine sweat gland, alterations in the nervous pathway or due to metabolic, endocrine or systemic disease. Purely psychogenic anhidrosis is also known. Occasionally it may not be possible to identify the cause. It may be localized, generalized, partial or complete. Extensive anhidrosis may impair heat regulation and produce heat exhaustion. The pathogenesis and underlying lesion of acquired idiopathic generalized anhidrosis (AIGA) are apparently heterogeneous. Acquired idiopathic generalized anhidrosis (AIGA) is a rare disorder in which systemic anhidrosis occurs in the absence of any causative skin, metabolic or neurological disorder. Congenital insensitivity to pain and anhidrosis (CIPA), also known as hereditary sensory and autonomic neuropathy type IV, is an autosomal recessive disorder characterized by the congenital lack of pain sensation, inability to sweat, episodes of recurrent hyperpyrexia, mental retardation, and self-mutilating behavior. It is due to mutations in the neurotrophic tyrosine receptor kinase 1 (NTRK1) gene in chromosome.

Key words: Anhidrosis, acquired anhidrosis, congenital anhidrosis.

Introduction:

A reduced level or complete cessation of sweating can be caused by a variety of factors affecting sweat glands directly or indirectly through alterations in their nerve supply. The most common presentation is the syndrome of heat intolerance with or without features of dysautonomia. An acquired idiopathic form of generalized anhidrosis is characterized by loss of sweating in the absence of any neurological features or destruction of sweat glands.¹ Idiopathic acquired generalized anhidrosis is a very rare condition in which the pathogenesis is still unknown.² Cholinergic urticaria has been associated with some cases of this acquired idiopathic form of generalized anhidrosis.³

Acquired idiopathic generalized anhidrosis (AIGA) represents a heterogeneous clinical syndrome including sudomotor neuropathy and failure of the sweat glands. However, most AIGA cases comprise idiopathic pure sudomotor failure (IPSF), a distinct subgroup without sudomotor neuropathy or sweat gland failure.⁴ Congenital insensitivity to pain with anhidrosis is a type IV hereditary sensory and autonomic neuropathy, presenting early in life. This disorder results from defective neural crest differentiation with loss of the first-order afferent system, which is responsible for sensations of pain and temperature; a neuronal loss in the sympathetic ganglia is also present.⁵ Being an autosomal recessive disorder, genetic counseling may be given to discourage consanguineous marriages, especially when there is a positive family history for CIPA.⁶

Discussion:

Nair et al stated that a 30-year old, a woodcutter man was seen for acquired anhidrosis. He came for complaints of absence of sweating and extreme intolerance to heat since 3 years. The body temperature rose to 40 0 C. Starch iodine test was positive in these sites. Neostigmine tablets 15mg four times a day failed to produce sweating. The disorder is likely to be due to a peripheral dysautonomia selectively affecting the sudomotor function.⁷ Murakami et al presents a 28-year-old Japanese man with acquired generalized anhidrosis. Although the body temperature rose to 38 degrees C, the patient did not sweat. Atrophy and degeneration of the sweat glands, as well as infiltration by lymphocytes and mast cells around the sweat glands, were observed in skin biopsies. Anhidrosis in this patient was suggested to be the result of reduced function of the sweat glands themselves with possible underlying immune-mediated basis.⁸

Ogino et al reported a 25-year-old man with acquired generalized anhidrosis due to occlusion of the coiled ducts. He did not have sweat secretion over the entire surface of the body, including the palms and soles. Electron microscopy demonstrated that the coiled ducts were completely occluded by an amorphous substance. The substance occluding the coiled ducts contained fibrous structures. These findings suggested that the acquired generalized anhidrosis in this patient was caused by occlusion of the coiled ducts by a PAS-positive substance probably derived from dark cell granules.⁹ Tsuji et al stated that a patient developed generalized anhidrosis, probably following sunstroke. Light microscopy showed an atrophic, deeply lobulated or elongated configuration of the eccrine sweat glands, most of them containing many vacuoles that possessed strong acid phosphatase activity. Electron microscopy revealed that the vacuoles showed areas of

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autolysis and were classified in the group of lysosomes. In addition, it is postulated that the anhidrosis resulted from a critical rise in body temperature with subsequent changes in the secretory cells.¹⁰

Terui et al reported a patient with idiopathic acquired generalized anhidrosis with the results of electron-microscopic and immuno-histochemical studies as well as an analysis of the lectin-binding pattern of the cell membrane.¹¹ Muta et al reported a 37-year-old man with acquired generalized anhidrosis but without other autonomic or somatic abnormalities (idiopathic acquired generalized anhidrosis) with special reference to histologic and morphometric findings of the eccrine gland and its nerve terminals and unmyelinated axons.¹² Shimizu et al reported a 61-year-old man who had a 10-year history of anhidrosis was found to have idiopathic diabetes insipidus. Within a month of desmopressin treatment for diabetes insipidus, sweating and skin sympathetic nerve activity returned.¹³ Ando et al reported a 40-year-old male developed complete absence of sweating except for slight sweating in the axillar region. Immunohistochemical staining using anti-CD3, CD4, and CD8 antibodies revealed that CD3 positive cells were dominant in the lesion. After intensive glucocorticoid treatment, generalized sweating was almost completely recovered.¹⁴ Dann et al reported a young Caucasian male, otherwise healthy, had generalized anhidrosis since early childhood. During heat stress tests (40 degrees C, 40% relative humidity), he was found to be heat intolerant since sweat was not apparent. This was the first reported case of familial generalized anhidrosis without anatomopathological lesions affecting sweat glands.¹⁵ Blanc et al reported a one month old girl with sudden attacks of fever and dehydration was found to have anhidrosis. Histologically the sweat glands were normal but the electrical resistance of the skin showed there was an abnormality of sweating. The cause of the disorder is unknown but could be due to a defect in the secretion of acetyl choline by the post ganglionic sympathetic nerves or due to a receptor defect in the sweat gland. This new syndrome should be considered in all infants with a high fever of unknown origin.¹⁶

Low et al describe the cases of eight patients with chronic idiopathic anhidrosis. These patients were heat intolerant and became hot, flushed, dizzy, dyspneic, and weak but did not sweat when the ambient temperature was high or when they exercised.¹⁷ Faden et al stated a patient had progressive segmental anhidrosis, which proved after extensive neurologic and autonomic workup to be an isolated abnormality. The alteration of the sudomotor response to intradermal acetylcholine during the course of the illness suggests that the diagnostic utility of pharmacologic sweat tests may be time dependent, limited to a relatively early period following onset of anhidrosis.¹⁸

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