Idiopathic generalized anhidrosis: A feature of panautonomic failure

Sir,

Anhidrosis refers to the absence of sweating in the presence of appropriate stimuli. Anhidrosis may result from abnormalities of sweat glands or from autonomic dysfunction.[1] It can be both congenital or acquired, and can be generalized or localized in nature. Chronic idiopathic anhidrosis has also been described as a forme fruste of acute panautonomic neuropathy.[2] We are reporting here a case of panautonomic dysfunction in a patient who presented with idiopathic generalized anhidrosis.

A 40 year-old woman presented with a history of recurrent bouts of high fever for the preceding 15 years. There was a generalized lack of sweating since childhood, except over the left half of her face, which also eventually became anhidrotic for the last 15 years. On presentation, she had a total absence of sweating, heat intolerance, as well as fecal and urinary urgency. Cutaneous examination showed shiny, smooth, hairless, dry skin. No sweating was visible even on provocation (with vigorous exercise and by taking hot drinks) and the result of the iodine-starch test was negative. Systemic examination revealed that the right pupil was irregularly dilated, the light reflex was absent in both the eyes although the accommodation reflex was intact; the extraocular muscle function was normal. In addition, she had postural hypotension. There was evidence of cardiac dysautonomia in the form of a decrease in heart rate variability

with deep breathing and an abnormal Valsalva response. Higher mental functions, muscle tone and power, as well as both superficial and deep reflexes were normal. Routine investigations (including blood sugar), ultrasonography of the abdomen, electrocardiogram, magnetic resonance imaging of the brain, and nerve conduction velocity studies of the peripheral nerves did not reveal any abnormalities. Cutaneous histopathology was normal with preservation of the sweat glands.

A large number of cutaneous or systemic conditions may underlie anhidrosis. These include neurologic disorders such as Guillain-Barre syndrome, heatstroke, diabetes, congenital disorders including ectodermal dysplasia, drugs, autonomic neuropathy, infections of or trauma to the sweat glands, burns, and excessive dehydration.

Localized anhidrosis is of limited clinical importance apart from its diagnostic value in leprosy. On the other hand, generalized or extensive anhidrosis (such as in the case of anhidrotic ectodermal dysplasia) may lead to hyperpyrexia with its associated complications.[1] Disorders of the autonomic nervous system may result from pathology of either the central or peripheral nervous systems.[3] Pure pandysautonomia is clinically characterized by some combinations of anhidrosis, orthostatic hypotension, paralysis of papillary reflexes, loss of lacrimation and salivation, impotence, impaired bladder and bowel function, flushing, and heat intolerance. Somatosensory and reflex functions are usually spared.[4] This patient showed an absence of sweating along with other features of autonomic failure in the form of urinary and fecal urgency, postural hypotension, asymmetry of the pupils with an absence of a light reflex, as well as an abnormal Valsalva response. Moreover, a histopathological examination of the skin did not reveal any abnormalities, indicating that the anhidrosis was due to autonomic failure. The neuroimaging and nerve conduction velocity were normal; hence, a diagnosis of idiopathic generalized anhidrosis was made.

Patients with generalized anhidrosis have a dangerous inability to tolerate heat. Therefore, when the weather is dry and hot, the inability to sweat can be life-threatening due to the potential to develop heat stroke. Anhidrosis may go unrecognized until a substantial amount of heat or exertion fails to cause sweating. This forms the background of the hyperpyrexia seen in our patient. In rare cases, family members of such patients may have anhidrosis and anisocoria,[5] but our patient did not have any such family history. In conclusion, anhidrosis can be present in some stages of autonomic failure, and a dermatologist must be aware of this fact so as to be able to routinely rule out the possibility of autonomic dysfunction when dealing with a case of anhidrosis.

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