Clinical Reports

Chronic idiopathic anhidrosis—a rare cause of heat stroke

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Summary: A 27 year old man presented with heat stroke following exposure to a humid, hot environment in the absence of physical exertion. Investigation revealed the presence of generalized anhidrosis without evidence of an associated disease. Although chronic idiopathic anhidrosis is rare, this entity should be considered in cases of unexplained heat intolerance and heat stroke.

Introduction

Heat stroke results from an imbalance between heat generation and heat dissipation.¹ It may occur following exercise in a hot, humid environment² or in the presence of various dermatological and neurological diseases that impair sweating.^{3,4} Idiopathic anhidrosis was first described in 1917 and since then only four other publications have reported this entity. We report a case of chronic idiopathic anhidrosis in which the initial clinical manifestation was heat stroke.

Case report

A 27 year old Caucasian male was admitted to our hospital in a deep coma (Glasgow coma scale 4). He had no previous illnesses and had been taking no drugs. On the day of admission he had taken a 3 hour drive in a car and had thereafter spent 2 hours in the sun (ambient temperature 38.6°C, 90% humidity, 28.8 discomfort index – equivalent to heavy heat burden). Two hours later he complained of fatigue, gradually became stuporotic and had a generalized convulsion. On admission his pulse rate was 140 per minute, systolic blood pressure was 75 mmHg and his rectal temperature was 40.8°C. His pupils were midposition, equal and reacted normally to light. There was no papilloedema. He had marked bilateral spasticity with clonus and did not respond to painful stimuli. Babinski's sign was not elicited. Blood tests for glucose, sodium, potassium, calcium, phosphate, urea, white cell count and haemoglobin were within normal limits. Chest X-ray and urinalysis were also normal.

The patient had a second generalized seizure in the emergency room. Endotracheal intubation was performed and he was treated with intravenous fluids, diazepam and phenylhydantoin. He was aggressively cooled with alcohol sponging and paracetamol suppositories. His temperature gradually fell to 37.0°C but he developed a generalized coagulopathy with frank haematuria and bloody diarrhoea. Prothrombin time was 31%, partial thromboplastin time was two times normal and repeat platelet count was 20,000/mm³. A computed tomographic (CT) scan of the brain showed generalized oedema. Lumbar puncture was completely normal. Blood, cerebrospinal fluid and urine cultures were negative. Creatine kinase was 157 U (normal < 100). Thyroid function test and cholinesterase levels were within normal limits. Within 12 hours of admission the patient became responsive to pain and within 10 days he had regained normal neurological function.

Two weeks after admission sweat tests were performed. Sweat glands in a 4–5 cm² area of skin on both arms were stimulated by pilocarpine iontophoresis for 5 minutes. Thereafter sweat was collected over 30 minutes on a weighted filter paper.⁵ The patient produced 15 mg of sweat on the left arm and 12 mg on the right arm (normal 120 ± 50 mg). Repeat testing after intravenous hydration yielded 20 mg of sweat on both arms. A heat tolerance test was also performed – the patient exercised by stepping on and off a 30 cm high bench at a rate of 12 times per minute. Ambient temperature was maintained at 40.0°C with a humidity of 40%.⁶ Rectal temperature rose to 38.9°C within 30 minutes but no sweating was noted. These results are compatible with severe heat intolerance. Punch biopsy of the skin showed intact eccrine sweat glands.

Follow-up neurological examination, including tests of autonomic function, were normal. The

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patient has subsequently avoided exposure to hot environments and has had no further episodes of heat stroke. Repeat heat tolerance tests 6 and 18 months later again revealed evidence of severe heat intolerance. There is no history of similar episodes in family members and heat intolerance tests performed on two siblings were normal.

Discussion

Heat intolerance is a condition in which the subject is unable to adapt physiologically to heat load.¹ It manifests clinically as malaise, fatigue and discomfort in a hot, humid environment and may progress to flushing, palpitations and even to hyperpyrexia (temperature $>40.0^{\circ}\text{C}$), convulsions and coma (heat stroke). If untreated, death follows soon thereafter. Heat stroke invariably occurs following physical exertion.

An impaired ability to dissipate heat may be due to reduced transfer of heat from the body core to the skin such as in low cardiac output states⁴ or impaired transfer of heat from the skin to the environment. The latter may occur in normal individuals wearing impermeable clothing⁷ or on exposure to extreme heat without adequate acclimatization.^{2,8} Various drugs such as tricyclic antidepressants and diuretics also predispose to heat intolerance.⁹

Hypohydrosis and anhidrosis refer to an impaired ability to sweat (partial or complete) and result in failure to dissipate heat adequately and therefore to heat intolerance. The condition may be localized to only part of the skin or may be generalized.

Most cases of anhidrosis are secondary to an underlying disease affecting the sweating apparatus either at a preganglionic level, as in autonomic neuropathy,³ or at a postganglionic level (distal to the nerve-sweat gland junction). The latter occurs in various dermatological disorders, such as Fabry's disease,¹⁰ congenital ectodermal dysplasia,^{11,12} scleroderma, miliaria rubra¹³ and secondary to radiation-induced injury to the skin.

In a small number of patients with anhidrosis, no underlying disease can be identified and the

condition is then referred to as idiopathic. The pathological features on skin biopsies from these patients are variable, ranging from normal to complete absence of sweat glands.

A review of the literature on patients with idiopathic or isolated anhidrosis revealed very little. In 1917, Lutembacher¹⁴ described a female with generalized anhidrosis in the absence of associated disease. In this case, however, skin biopsy was not performed. Mahloudji and Livingstone¹⁵ describe a family with three siblings who suffered from anhidrosis but in whom skin biopsies showed complete absence of sweat glands. Tsuji and Yamamoto¹⁶ report a case of anhidrosis with atrophic sweat glands on skin biopsy. The only reported series of patients with idiopathic anhidrosis is that of Low *et al.*¹⁷ who describe eight cases with this condition. Most of these patients had mild accompanying neurological deficits and in two of their cases the anhidrosis was congenital. In four of the cases the defect was postganglionic and in four preganglionic. Skin biopsy was performed in only one of their patients and was reported as normal. One of the authors has previously described a case of familial anhidrosis.¹⁸ Skin biopsy in this patient was also normal.

Our patient is only the fourth case of idiopathic anhidrosis reported in the English medical literature in whom skin biopsy was performed and found to be normal. He is the first patient described in whom heat stroke in the absence of physical exertion was the presenting manifestation of this disorder.

Pilocarpine iontophoresis revealed a markedly reduced response to muscarinic stimulation suggesting that the sweating defect in this patient is postganglionic.

In conclusion, anhidrosis should be considered in any patient presenting with heat stroke in whom environmental factors and workload do not adequately explain the condition. This entity is of special relevance to medical personnel dealing with army recruits.

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