Isolated Simple Anhidrosis: A rare cause of fever of unknown origin

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Abstract

Here we report a one-year old Saudi female with pyrexia of unknown origin (PUO) and isolated simple anhidrosis with normal sweat gland number and morphology was found as a cause of this infant PUO.

Introduction

Anhidrosis is an abnormal absence of perspiration from the surface of the skin in the presence of an appropriate stimulus that usually induces sweat under similar conditions [1]. Anhidrosis or hypohidrosis can cause hyperthermia because of failure to dissipate heat [2]. Isolated simple anhidrosis is very rare condition with only few cases reported. Here we report a one-year old female with pyrexia of unknown origin (PUO) .clinical and laboratory investigations confirmed a diagnosis of isolated simple anhidrosis with normal sweat gland number and morphology as a cause of PUO in this infant.

Skin biopsy was obtained from the sole of the foot. Biopsy was routinely processed and thin sections [8µm] were stained with haemotoxylin and eosin. Photomicrographs of the selected areas were taken.

Case report

A one-year-old Saudi female infant presented with PUO. Further history revealed that since the age of 1 month, the child had had multiple admissions and extensive investigations for episodes of PUO, irritability and decreased feeding but without revealing any etiology. The mother reported that her daughter never sweats, becomes febrile and irritable when exposed to hot weather, and calms down when exposed to cool weather. Family history revealed no consanguinity with healthy parents and no inherited diseases in the family. Her developmental milestones were appropriate for her age. On examination, she was febrile with temperature up to 40 °C, irritable but not ill looking and there were no dysmorphic features. Her weight, height and head circumference were 8.5 kilograms, 72 centimeters and 43 centimeters at the 10th, 25th and 5th centile respectively. Other systemic examination was unremarkable. Laboratory investigations showed leukocytosis of 18.4 X $10^3/mm^3$ with either normal or negative following tests; lactic acid, blood, urine and cerebral spinal fluid cultures, hepatitis A, B, C, brucella, toxoplasma and EBV serology, eelectromyography and chest x-ray. Computed tomography scan of the head, chest and pelvis was normal. Skin biopsy obtained from the sole of the foot showed normal sweat glands in both number and morphology (Figure 1). A stress iodine-starch test was performed under controlled condition with full resuscitation equipments available. Briefly, the girl was put inside an incubator and the temperature was elevated gradually to 40°C. The patient oral temperature

reached 39°C with no sweating observed which confirmed complete anhidrosis. Based on the clinical presentation and laboratory investigations a diagnosis of simple isolated anhidrosis was made in this infant. The family was instructed to minimize exposing the infant to heat and implement cooling measures whenever outdoor in hot climate and to use antipyretics whenever febrile. The infant was discharged a febrile in a stable condition.

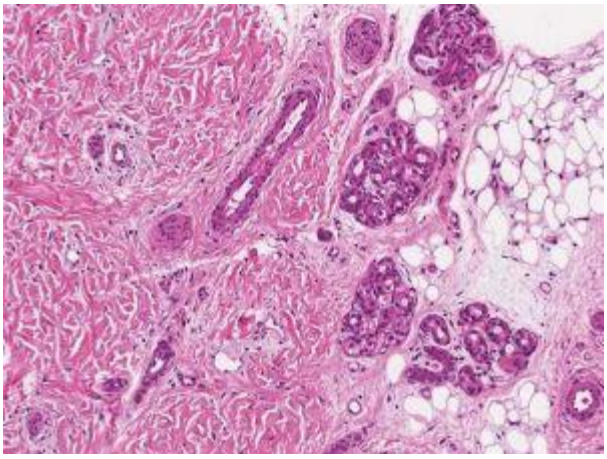

Figure 1: Skin biopsy from the sole of the left foot revealing multiple normal looking sweat glands (Haematoxylin and Eosin stain × 100)

Discussion

Body heat dissipates through evaporation of sweat into ambient air resulting in cooling the skin through evaporation [2]. Anhidrosis is defined as an abnormal absence of perspiration from the surface of the skin in the presence of an appropriate stimulus that usually induces sweat under similar conditions [1]. Anhidrosis or hypohidrosis can cause hyperthermia because of failure to dissipate heat[2].

While local anhidrosis is usually of minor clinical importance, its existence may serve as a clue as in the diagnosis of leprosy[1].The congenital forms of generalized anhidrosis is usually hereditary, as a manifestation of ectodermal dysplasia [1]. In 1950, Shelley et al classified generalized anhidrosis into four main etiologies; a defect in the central nervous system, a defect in sweat glands or their ducts, a defect in intermediate site and idiopathic anhidrosis [1]. The defect in intermediate site is a defect in the neurotransmitters or presynaptic receptors [3].

Literature search indicates the rarity of isolated simple anhidrosis as few cases were reported. In 1917, Lutembacher et al, reported a female with isolated anhidrosis but skin biopsy was not done [4]. In 1967, Mahloudji and Livingston, reported three Iranian siblings with generalized anhidrosis and unremarkable systemic examination Skin biopsies obtained from axillary area revealed complete absence of sweat glands [5]. In our patient we confirmed anhidrosis with normal skin biopsy .Sweat stress test using heater blankets with the elevation of the oral temperatures up to 40°C confirmed complete anhidrosis. Upon reviewing the literature Mahloudji et al named this new syndrome familial and congenital simple anhidrosis [5]. In 1988. Frydman et al reported two siblings with anhidrosis due to paucit y of sw eat glands ducts as seen in skin biopsy [6].

In 1990, Dann et al reported a young Caucasian male suffering from severe heat intolerance with normal physical examination [4]. A heat stress test revealed complete anhidrosis. While skin biopsy revealed normal sweat gland in number and morphology [4]. The patient muscirinic response to pilocarpine introphoresis was 10% of normal [4]. His mother was moderately affected but other family members were normal 4. In an accompanying editorial, Inbger suggested to name this new syndrome Dann-Epstein-Sohar Syndrome [1]. We believe that our patient closely resembles Dann et al patient as proven by absence of sweating despite normal skin biopsy.

Other causes of anhidrosis include congenial and acquired causes [1]. Ectodermal dysplasia is usually apparent by finding of abnormal formation of ectodermal structures These include fine, sparse hair, anodontia or hypodontia with conical teeth [7]. Congenital insensitivity to pain with anhidrosis (type IV) presents early with repeated high fevers due to absence of sweating [8]. Decreased pain sensation and absence of unmyelinated fibres in the sural nerve confirm

the diagnosis8. Fabry's disease is an uncommon X-linked recessive disease with childhood onset of chronic acral paresthesias, heat intolerance, lack of sweating, and angiokeratoma corporis diffisum [9].

Acquired causes of anhidrosis include idiopathic sudomotor failure with early onset anhidrosis, concomitant sharp pain or cholinergic urticaria over the entire body, absence of other autonomic dysfunction, elevated serum IgE levels and marked response to glucocorticoids [10]. Ross Syndrome comprises a triad of segmental anhidrosis, tonic pupils and hyporeflexia [11].

In conclusion, we report a Saudi female who presented with PUO which was due to simple generalized anhidrosis. This entity seems to be very rare. Treatment is largely supportive with avoidance of heat exposure.

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