Hypohidrotic Ectodermal Dysplasia of Sisters in a Family

Ectodermal Dysplasia (ED) is a group of rare disorder that affects various tissues of ectodermal origin. The most common form is hypohidrotic (anhydrotic) ED of X-linked recessive type affecting only males.

Two daughters one four years and another one year of age from a consanguineous married muslim family were admitted in the pediatric department of Burdwan Medical College, West Bengal with history of high grade fever and cough of short duration with a background history of frequent rise of body temperature since early infancy. Absence of sweating even in hot summer, lack of hair and abnormal dentition were associated complaints. Family history revealed healthy parents with these two daughters without history of similar illness in relatives. Clinical examination showed IAP grade III PEM without edema, occasional thin strands of silky hair giving the appearance of total baldness, absent eyelash, frontal bossing, depressed nasal bridge and dry skin but normal nails in both of them. Elder sister had only two conical upper incisor teeth but the younger had none. Others systemic examinations revealed no abnormality except few crackles in lungs. Palmer skin biopsy of both the sisters showed thinned out epidermis, absence of sweat glands and pilosebacious unit in dermis suggestive of hypohidrotic (anhidrotic) ED. Other investigations were inconclusive. ARI responded to oral cotrimoxazole.

The ED is a heterogeneous group of disorders by constellation of findings involving defects of two or more of the following the teeth, skin and appendageal structures including hair, nails and eccrine and sebaceous gland(1). This disorder was first described by Thurunam in 1848(2). The incidence has been reported to be 1 per 10,000 to 1 per 100,000 live births(3). Clinically, ED are of two types: hypohydrotic (anhydrotic) and hydrotic. Hypohidrotic ED is manifested as a triad of defects: partial or complete absence of sweat glands, anomalous dentition and hypotrichosis, anodontia or hypodontia with widely spaced, conical teeth is a constant feature(1). Episodes of high fever in warm environment, dry skin, sparse hypopigmented hair, frontal bossing, flattened nasal bridge are some of other features, Poor development of mucous glands in the respiratory tract may result in increased susceptibility to respiratory infection.

In our presenting case both the sisters satisfied most of the clinical features of hypohidrotic (anhidrotic) ED which was confirmed by palmar skin biopsy.

The mode of inheritance of ED is X-linked recessive with full expression only in males; however, an autosomal recessive mode of inheritance may be operative in some families(1). Monoz, *et al*. in 1997(4) suggested that heterozygous females of unaffected mother having variable clinical features of ED occur because of random X- inactivation (Lyon hypothesis) and mosaic of functionally normal and abnormal cells. The inheritance of presenting case may be autosomal recessive or random X-inactivation. However, genetic analysis was not possible in our working set up.

S. Ghosh,
M.K. Das,
Department of Pediatrics,
BMC, Burdwan, West Bengal, India.

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Mosquito Coil (Allethrin) Poisoining in Two Brothers

The pattern of accidental poisoning in children is changing. There is an increasing incidence of poisoning with common house hold insecticides(1,2). Mosquito (Allethrin) coil, a type of pyrethroid is almost ubiquitous in modern households. We report two brothers brought in unconscious state with alleged history of mosquito coil ingestion.

Two cousin brothers 4 yrs and 3 yrs were brought in department of emergency medicine with history of one episode of generalized tonic-clonic convulsion followed by un-consciousness and violent behaviour. Parents suspected the intake of mosquito coils (they were not sure of quantity of coils ingested). There was no history of pica in the children. At admission both were comatosed ($E_2V_2M_4$) with normal pupillary size and reaction. Vitals were normal. There were no meningeal signs or focal neurological deficits. Routine hematological studies, serum bilirubin, ALT, AST and blood sugar were within normal limits. Stomach wash revealed clear fluid. The older child had two episodes of generalized convulsions, which were managed with intravenous diazepam. Both children regained consciousness in 36 hours with supportive management. They accepted ingestion of one mosquito coil each. On follow up after two weeks both were neurologically normal.

Pyrethroids are common insecticides. There are only few reports of pyrethroid poisoning from India(2,3). Pyrethroids are of two types. Allethrin, present in mosquito coil is a type I pyrethroid which lacks a cyano group and causes repetitive discharges in nerve fibres (by acting on sodium channels) leading to hyperexcitation as compared to Type II pyrethroids (deltamethrin and fenvalerate) which causes nerve membrane depolarization and block leading to paralysis. Allethrin poisoning thus leads to ataxia, loss of coordination, hyperexcitation and convulsions. Recently calcium channels have been also found to be another primary target for allethrin(4). Pyrethroids are generally considered to be safe insecticides in humans because of their rapid biotransformation by ester hydrolysis and hydroxylation to their inactive acids and alcohol components. The fatal dose for allethrin is not known. In our case ingestion of one mosquito coil was sufficient to lead to symptoms. Most cases of acute pyrethroid poisoning recover within 1-6 days with normal neurological out come on follow up. Deaths have been reported in cases where convulsions increase in duration and frequency and do not stop within 2-3