aged >20 years or, 2 minor criteria; 1) characteristic histological changes in non-lesional skin and 2) history of PXE in first degree relatives. Management includes timely photocoagulation of retinal hemorrhage to prevent choroiditis and visual loss, prevention and management of coronary occlusion, gastrointestinal or cerebral bleeds which may end fatally otherwise, cosmetic improvement and genetic counseling. Identification of mutations in the ABCC6 gene (chromosome locus 16q13.1) encoding MRP6 protein provides prenatal and presymptomatic testing in families at risk.

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A five-year old boy presented with symptoms of itching and pain in the phallus, especially at night, for the last 3 months. The caregivers had noticed a progressively developing scab located just proximal to the glans. The child had been seen by several practitioners who gave local antibiotic ointments. Since the last two weeks, the child had been passing urine partly from the affected area. On examination, there was no penile edema or discharge, and only a fibrotic groove proximal to the glans with a scab. Once the scab was elevated, hair coil strangulation of the penis with the offending hair could be seen (Fig. 1). There was also a partial disruption of the urethra.

Hair coil strangulation or penile tourniquet injury is a rare and potentially devastating condition that has been reported mainly in circumcised children. The hair is thought to belong to the patient, and a majority of cases are believed to be accidental although in select cases, child abuse should be suspected. Hair is extremely thin, has high tensile strength and is easily overlooked especially in the presence of a foreign body reaction. Moreover, hair stretches when wet and shortens when dried, which makes it an efficient tourniquet. Both superficial, and more commonly, deeper injuries such as transection of the urethra and partial or complete amputation of the glans has been reported, necessitating repair of varying degrees of complexity. Awareness of the condition, with early detection and removal of the offending coil of hair can prevent these complications.

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Fig. 1 Hair coil injury with partial transaction of the urethra.