

Pseudoxanthoma Elasticum

A 15-year-old boy of non-consanguineous parentage had progressively increasing loose folded skin of three year duration. Skin over neck, shoulders, chest and axillary folds showed yellowish pebbly plucked-chicken skin appearance and large well-defined atrophic plaques over neck having erythematous and ragged margins studded with discrete keratotic papules which would bleed on removal (**Fig 1**). The trunk skin was soft, lax and had hanging redundant folds (**Fig 1**). Hair, nails, mucous membranes and other systemic examination were normal, and ophthalmoscopy showed no angioid streaks. Laboratory investigations including hemogram, serum biochemistry, urinalysis, chest X-ray, ECG, echocardiography and doppler studies were normal. Histology showed normal epidermis, swollen, irregularly clumped degenerated faintly basophilic and calcified dermal elastic fibers, and mixed inflammatory cells. He was diagnosed having pseudoxanthoma elasticum.

Pseudoxanthoma elasticum (PXE) is an uncommon autosomal recessive disorder of generalized elastorrhexis and calcification of elastic fibers in the dermis, Bruch's membrane and arterial lamina. Spontaneous perforating skin lesions with transepidermal elimination of fragmented elastic fibers may develop manifesting as hyperkeratotic papules. PXE needs to be differentiated from yellow-orange plaques of localized or generalized plane xanthomas seen associated with abnormalities of lipid metabolism, and juvenile elastoma wherein skin lesions show thickened elastic fibers histologically. A PXE-like clinicohistopathologic syndrome in patients with β -thalassemia, sickle cell anemia or sickle thalassemia has late onset and is acquired as consequences of the primary disease. In view of clinical heterogeneity, the diagnosis of PXE requires all three major criteria; 1) The characteristic "plucked-chicken-skin" appearance that becomes evident by second decade and progressively becomes lax and redundant, 2) characteristic histology and 3) angioid streaks, symmetric tears in the Bruch's membrane, in adults

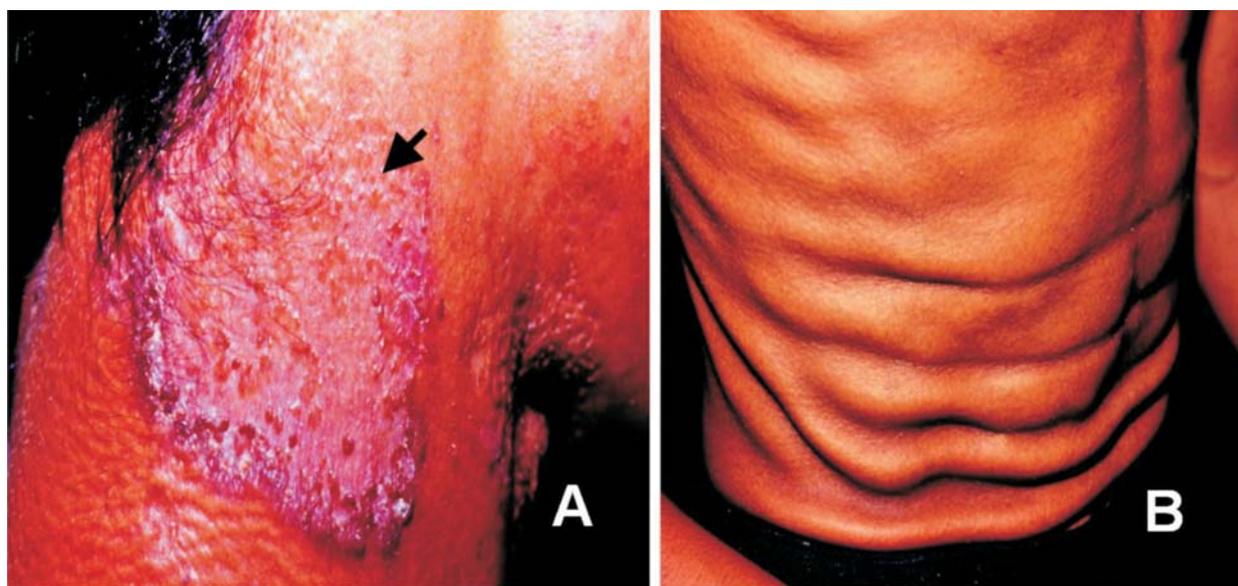


FIG.1 (a) Perforating lesion of pseudoxanthoma elasticum (arrow) showing central atrophy and discrete keratotic papules over the erythematous margins and central atrophic skin. (b) Folded, lax, redundant abdominal skin.

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 pre-symptomatic testing in families at risk.

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Hair Coil Strangulation of the Penis

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



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

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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