

## Subluxation of Eyes in Crouzon Syndrome

A 3-year-old boy, diagnosed case of Crouzon syndrome, presented with sudden protrusion of both eyes out of the orbit during a bout of cough (**Fig. 1**). On examination, vitals were stable. Bilateral subluxated eyes were seen, with normal cornea and anterior chamber. Pupils were bilaterally equal and reacting. Eye ball movements were absent with normal intra-ocular pressure; fundus was normal. Anterior narrowing of the head was present with a normal head circumference. Examination of limbs as well as systemic examination was within normal limits. Eyes were repositioned and emergency lateral tarsorrhaphy was done, following which the child had normal vision with no residual damage. After a month, he underwent surgical correction of craniosynostosis and orbital reconstruction, and there has been no recurrence of subluxation of eyes thereafter.

While subluxation of globe is rare in children, Crouzon syndrome is known to be a predisposing factor. Timely repositioning usually results in a good outcome.



**FIG 1** Subluxation of eyes in a child with Crouzon Syndrome.

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## Genital Herpes Infection in an Adolescent with Diabetes Mellitus

A 14-year-old girl with insulin-dependent diabetes mellitus presented to us with a 3-day history of burning vaginal pain. She had no fever, vomiting, or diarrhea. She had no previous hospital admissions for diabetes, and had no history of any sexually transmitted diseases. Examination showed an erythematous and edematous perineal area dotted with numerous painful vesicles extending over the entire external genital organs, including clitoris, labia majora and the anal region (**Fig. 1**); along with inguinal lymphadenopathy. There was no vaginal discharge. Blood gas analysis revealed metabolic acidosis, and urine examination confirmed presence of glycosuria and ketonuria. We diagnosed her as having diabetic ketoacidosis induced by genital herpes infection. Herpetic serology was initially negative, but seroconversion for herpes simplex virus 2 (IgM) was documented after one month. She was treated with intravenous aciclovir (10 mg/kg/dose 8 hourly) for 10 days along with insulin therapy.



**Fig. 1** Multiple vesicles over genital region in an adolescent girl.

Genital lesions began to improve after two days of treatment.

Genital lesions caused by herpes simplex virus are rare in children and adolescents, who are not sexually active. The main differential diagnosis is vulvo-vaginal candidiasis, which is characterized by the pruritus and raspberry aspect of the inguinal folds surmounted by whitish coating. In the presence of a clump of painful

vesicles associated with inguinal adenopathy, the diagnosis of genital herpes is very likely.

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## Porokeratotic Eccrine Ostial and Dermal Duct Nevus

A 3-year-old girl presented to us for a rough aspect of the trunk. Dermatologic examination showed multiple rough papules in the upper and lower limbs (**Fig. 1a**), extending to the trunk (**Fig. 1b**) and upper limbs with a linear arrangement, and then to the face (**Fig. 1c**). We also found a focal palmoplantar keratoderma distributed in small islands. According to the clinical aspect of the lesions, we suspected porokeratosis that was confirmed by the skin biopsy. She was treated by acitretin (0.25 mg/kg/day) with good evolution within 3 months.

Porokeratotic eccrine ostial and dermal duct nevus (PEODDN) is a rare variant of porokeratosis due to a mutation in *GJB2* gene coding for the connexin 26 junction protein. PEODDN can present as multiple hyperkeratotic or atrophic papules grouped on plaques with a blaszko linear arrangement. These lesions are mainly located at the extremities and are frequently associated with a palmoplantar involvement. Proximal involvement of the extremities, the trunk and the face, as seen in this patient is very rare. The pathognomonic histological findings are characterized by cornoid lamella located in relation of a dilated sweat excretor canals and associated to vacuolated keratinocytes in the epidermic invagination. Differential

diagnoses include Linear porokeratosis, linear lichen planus, linear verrucous epidermal nevus, and linear Darier's disease. PEODDN start at birth or in the early childhood. Clinically it resembles a comedonal nevus, but it occurs on the palms and soles where pilosebaceous follicles are normally absent. Treatment options include



**FIG.1** (a) Multiple papules in the upper limbs with a linear arrangement; (b) Multiple rough papules in the trunk; and (c) Extended lesions to the trunk and the face.

dermocorticoids, topical calcipotriol, cryotherapy, CO<sub>2</sub> laser, photodynamic therapy and retinoids.

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