Cutis Verticis Gyrata

A female infant, born of non-consanguineous marriage, presented with edema of hands and feet and loose folds of skin on the vault of skull which her parents noticed from birth. The baby was a term normal delivery. Her birth and developmental history was otherwise uneventful. On head to foot examination, the baby presented with 6-8 loose puckered folds of skin, extending from anterior to posterior in the saggital plane, spanning the vertex and occipital region. The folds were spongy, non tender with overlying hair being sparse. Besides, she had loose folds of skin at the nape of her neck and non-pitting edema of her hands and feet. G-Band karyotype revealed 45 XO. Cutis Verticis Gyrata (CVG) is a rare entity with an estimated prevalence of 1 in 100,000 with an estimated male to female ratio of 6:1. CVG exists in primary and secondary forms. Primary forms exist in primary essential (in which no other abnormality is found) and primary non essential (that can be associated with cerebral palsy, epilepsy, and cranial or ophthalmologic abnormality. Secondary forms are associated with pachydermatosis, melanocytic nevi, dermatofibroma and some syndromic associations like Noonan syndrome, Turner syndrome and many more. In our case, the baby had Turner syndrome, as evident by the karyotyping report. Hence this was a secondary cause of CVG. The close differentials are acromegaly, cutis laxa (elastolysis), cylindroma, and pachydermo-periostosis.

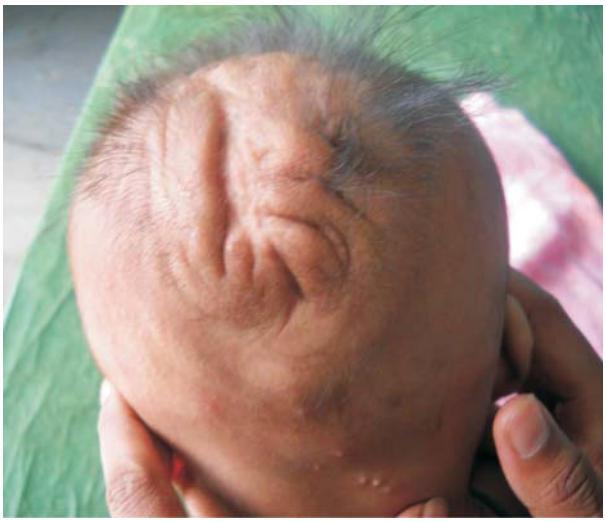

FIG.1 *Cutis Verticis Gyrata.*

The course of the disease, in majority of cases is benign except in primary cases caused due to cerebriform melanocytic naevus, close follow-up is required to detect early malignant transformation. Treatment includes maintenance of optimum hygiene of the skin folds and surgical correction of these folds by laser.

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Erythema Multiforme

An 11-year-old boy presented with slightly itchy skin eruption on his palms for the preceding 5 days. Examination revealed multiple circular plaques with central dusky coloration, bullae formation, and peripheral erythematous rings on his palms. The central bullae or dusky coloration with surrounding concentric rings resemble the appearance of a 'target'. A few similar skin lesions were also seen on the other areas of his body. There was no mucosal lesion. Based on the distinctive clinical feature, a diagnosis of Herpes simplex-associated erythema multiforme (EM) was made (**Fig. 1**).

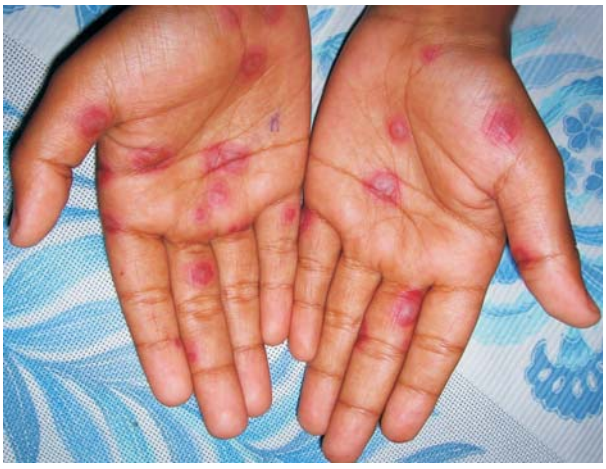

FIG.1 *Erythema multiforme*

Erythema multiforme is a cutaneous reaction pattern precipitated mainly by various infections and drugs. Herpes simplex virus (HSV) 1 and 2, adenovirus, measles, *Mycoplasma*, and *Yersinia* are considered important infectious cause of EM amongst others. Drugs like sulfonamides, penicillin, cephalosporin, and tetracycline may also precipitate EM. Rare causes of EM include malignancies and collagen vascular diseases. However, no underlying cause is found in a number of cases.

Other differential diagnoses that should be considered in the present case are: urticaria, hand foot and mouth disease (HFMD), fixed drug eruption (FDE), vasculitis, and urticarial vasculitis. In urticaria the central zone comprises of normal skin, lesions usually change within hours, associated with swelling, and lesions resolve and re-appear at different sites on daily basis. On the other hand, the central zone of EM is damaged skin, lesions are symmetrical, fixed (at least for some days), and all lesions usually appear within a few days. FDE may resemble EM, but usually the lesions are solitary or a few compared with multiple lesions of EM. FDE usually manifests as round or oval, sharply delinea-ted erythematous plaque the center of which may blister or become necrotic. It gradually fades away with residual hyperpigmentation. Moreover, recurrent lesions usually appear at the same anatomical site. In HFMD, the characteristic rash consist of flat or raised erythematous lesions, sometimes with vesicles with a peri-lesional erythematous halo and are usually located on the palms, soles, knees and buttocks. The lesions on the palms and soles are characteristically elliptical in shape. Associated buccal mucosal lesions are usually present. Vasculitis or urticarial vasculitis may also mimic EM, but the target lesions are usually absent. Finally, histopathological examination of the lesional skin often helps to differentiate EM from other close mimickers.

EM is usually a self-limiting condition and management should focus on treating the underlying infection or immediate withdrawal of the offending drug. Oral acyclovir has been shown to be beneficial in EM caused by HSV and also in suppression of recurrent EM.

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