Images in Clinical Practice

'Custard apple' scalp

A 29-year-old man presented with gradually progressive, asymptomatic folds of his scalp skin for the past 10 years. It started as 'doughy feeling' of the skin over his vertex at puberty which gradually progressed to form alternate areas of convexity and deep concavity involving the same area. There was no disfigurement of face or extremities and neither was any family history of similar disorder.

On examination of the patient's head with cropped hair, there were deep furrows in the anteroposterior direction involving the whole vertex. On application of centripetal pressure on the scalp by placing examiner's hands at both fronto-parietal regions, the skin folds attained a criss-cross pattern simulating the outer surface of custard apple fruit [Figure 1]. Other areas of scalp were looking normal and there was no alteration of hair density. His facial configuration was normal and lines of expressions were appropriate to his age. Hands and feet were normal in configuration and feel. Increased seborrhea and palmoplantar hyperhidrosis were not present. Ophthalmological examination was within normal limits. Patient's mental development was appropriate to his age.

A clinical diagnosis of 'primary essential cutis verticis gyrata' was done. Moreover, a skin biopsy was taken from the involved scalp. H and E stained tissue section showed hyperkeratosis and focal parakeratosis. Dermis was thickened with thick fibrous bands extending into deeper tissues. There were increased collagen bundles, fibroblasts and ground substance. Focal aggregates of lymphocytes were present. The histopathological features were consistent with cutis verticis gyrata. The patient was assured of the benign nature of the condition and maintenance of scalp hygiene was advised.

Figure 1: Scalp skin simulating the surface of custard apple

Cutis verticis gyrata (CVG) is characterized by folded hyperplasia of the scalp resulting in a cerebriform appearance. In primary essential form of CVG, gyrate folds on the scalp are the presenting feature, without associated anomalies. In the primary non-essential form, various mental, cerebral and ophthalmological abnormalities are present. Secondary form of CVG is associated either with nevoid malformations of the scalp or part of genetic disorders like pachydermoperiostosis, Rosenthal-Kloepfer syndrome, Turner's syndrome or endocrinological disorders like acromegaly, myxedema, insulin resistance, or a paraneoplastic disease.

Primary essential CVG typically starts at puberty and males are the common sufferers. Majority of the cases are sporadic but other family members are often affected. Typically, folds are present over vertex and/or occipital areas; rarely entire scalp may be involved. The condition is asymptomatic except lack of adequate cleaning of the invaginated areas may give rise to foul odor due to collection of debris. Histopathology may be normal in most cases of primary CVG.

It is important to rule out any underlying disorder in all cases of CVG, before labeling it as the primary essential variant. Management includes counseling regarding benign nature of the condition as well as education regarding maintenance of scalp hygiene.

Surgical correction of the deformity may be helpful in selected cases.

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The appearance of the scalp skin akin to the surface of 'custard apple' in this patient was due to the centripetal pressure exerted by examiner's hands resulting in accentuation of the existing folds as well as appearance of inapparent folds.

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