LETTER TO EDITOR (VIEWER'S CHOICE)

GINGIVAL HYPERPLASIA, HYPERTRICHOSIS AND A CHARACTERISTIC FACIES: A CASE REPORT

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A 6 year old boy was seen in the Outpatients Department with the complaints of gingival hyperplasia which was first noticed at 6 months of age and had been increasing gradually since. It was now causing difficulty in eating. Parents noticed at birth that the child had excessive hair. His growth, development and intellect were normal for age. There was no history of any drug administration, convulsions or any other neurological symptoms. There is no consanguinity. He has 2 siblings who are well. On examination, the child had a coarse facies with a prominent and broad forehead, a very broad nose, large ears, full lips and a prominent jaw. The palate was high arched with marked gingival hyperplasia. He had abundant facial hair, thick eyebrows and eyelashes and generalised hirsuitism with normal hair texture. (Figure 1, 2, 3 &4). He had a short neck and short broad hands. His weight was 15 kg (25th percentile), height was 110 cms (50th percentile) and head circumference was 49 cms (50th percentile). His IQ on assessment was within normal range. Dental examination revealed presence of marked fibrous overgrowth of gingival tissues covering all the erupted deciduous teeth. (Figure 5) Hand X-rays showed broad first metacarpal, Skull X-ray showed prominent maxilla and mandible, but no osteochondrodysplasia. The karyotype, 46 XY at 500 band resolution was normal. Bilateral gingivectomy was done, histopathology revealed parakeratinised epithelium and dense connective tissue. Following the gingivectomy there was eruption of full complement of deciduous teeth. He has come back for follow up and is now able to eat better. Further plan is to monitor him and watch for any complications.

Gingival hyperplasia with hypertrichosis has been well described in literature. The facial features have been described as mostly normal. (1-4) Canun from

Fig 1

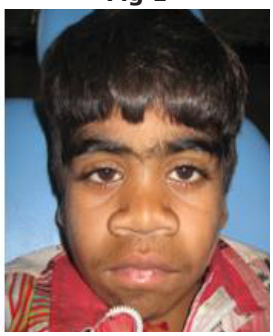


Fig 2

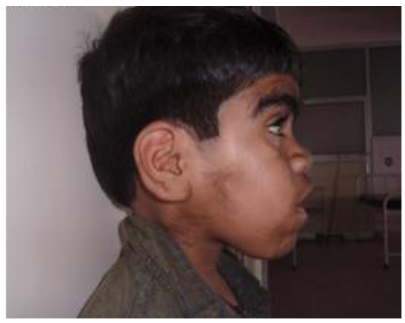


Fig 3

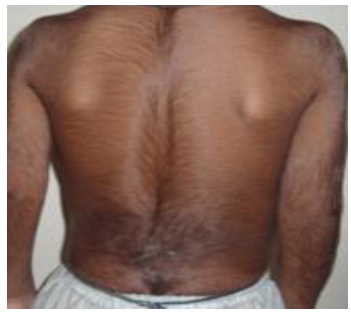


Fig 4

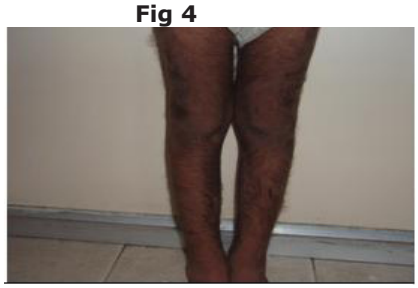
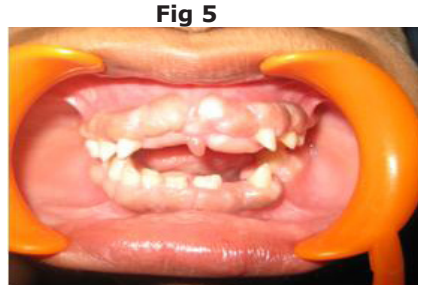


Fig 5



Mexico described an 11 years old girl who had gingival hyperplasia, hypertrichosis with a coarse and typical facies and proposed that this was a separate entity. (5)

This has syndromic associations namely Ambras which is associated with paracentric inversion (8) (q12;q22). (7) and Cantus syndrome which has associated osteochondrodysplasia and cardiomegaly. (8) Gingival hyperplasia, hypertrichosis, with Mental retardation and epilepsy was reported by Anavi (9). Only Bondeson (10) and Canun (7) have described the facial features in detail. Their patients had coarse craniofacial features very similar to ours. Their patients were of Mexican origin and to the best of our knowledge our patient is the first reported one from India. Canun et al proposed that this association of hypertrichosis, gingival hyperplasia and a characteristic facies is a separate entity. We report this first case from India and reinforce that this is a separate entity. Further studies are required to establish the etiology and pathogenesis of this condition.

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E-published: 1st October 2010 **Art**#61