A Rare Case of Gingival Fibromatosis Associated with Hypertrichosis and a Dysmorphic Face

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Several forms of hypertrichosis have been described with and without gingival hyperplasia; some of them are recognized as genetic disorder and associated with syndromes. In all reported cases the most striking differences from other are the craniofacial features. We present a case of a 6-year-old boy with hypertrichosis associated with gingival hyperplasia and a characteristic, coarse face and we consider this case to be a distinctive entity.

Keywords: *congenital hypertrichosis, gingival fibromatosis, children*
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INTRODUCTION

Gingival hyperplasia is frequently an isolated disorder, but in some cases, it is associated with other symptoms that characterize certain syndromes. Bondenson and Miles (1993)[2] described a Mexican woman with a very distinctive "ape-like" appearance, hypertrichosis and over- development of the jaws. This woman and her son had coarse faces, congenital and universal hypertrichosis, terminal hairy growth and gingival hyperplasia. The researchers proposed this as a separate entity. Gingival hyperplasia with hypertrichosis has been well described in the literature.[3-5] These conditions syndromic associations, namely with Ambras syndrome and Cantu syndrome.[6-10]

We studied a 6-year-old boy with hypertrichosis associated with gingival hyperplasia and a coarse face. His case was similar to the cases reported by Canun,[11] and by Bondenson and Miles.[2]

Case report

A 6-year-old boy was first brought to the Department of Pediatric and Preventive Dentistry because of gingival over-

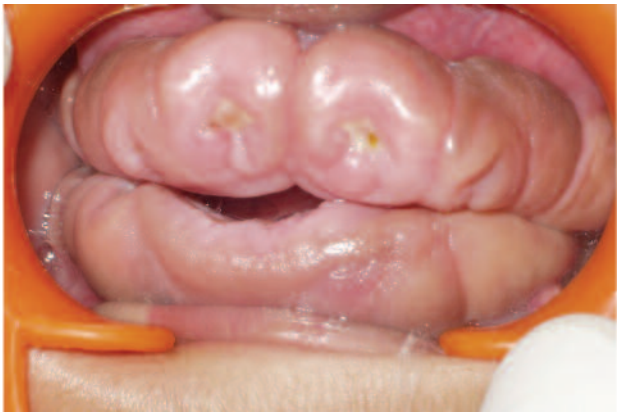

Figure 1. Generalized gingival hyperplasia covering deciduous erupted teeth

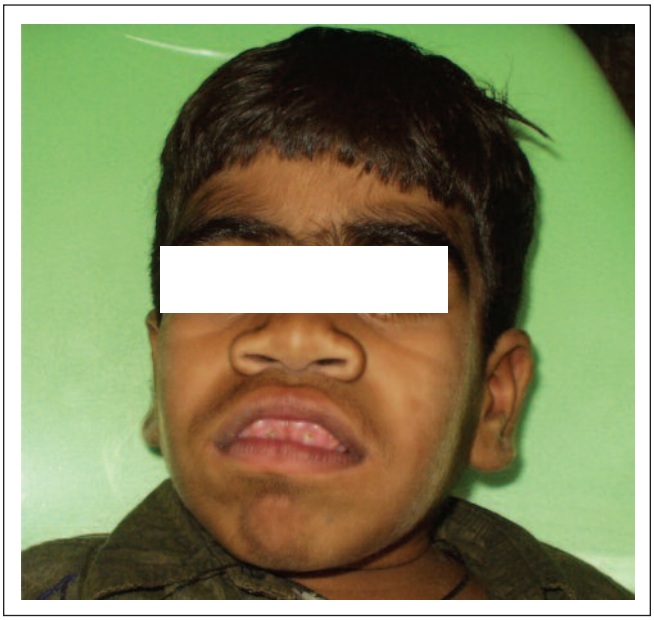

Figure 2. Hypertricosis with coarse facies

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growth, which was causing him difficulty in eating (Figs.1 and 2). He was the first son of unrelated parents; the pregnancy and delivery were uneventful. His parents and two younger siblings, a 4-year-old sister and a 2-year-old brother, showed no sign of gingival overgrowth and hypertrichosis. There was no history of any drug administration. At the examination, the patient presented with hypertrichosis and generalized gingival hyperplasia covering all primary teeth. He had dysmorphic facial features, which included a prominent and broad forehead, thick and abundant eye lashes, a broad nose with depressed nasal bridge and labial fullness with a prominent jaw. The palate had a high arch and both of the dental arches were square. He had a large amount of facial hair and generalized hypertrichosis with normal hair texture and pigmentation. He had a short neck and short, broad hands (Figs. 3 and 4).

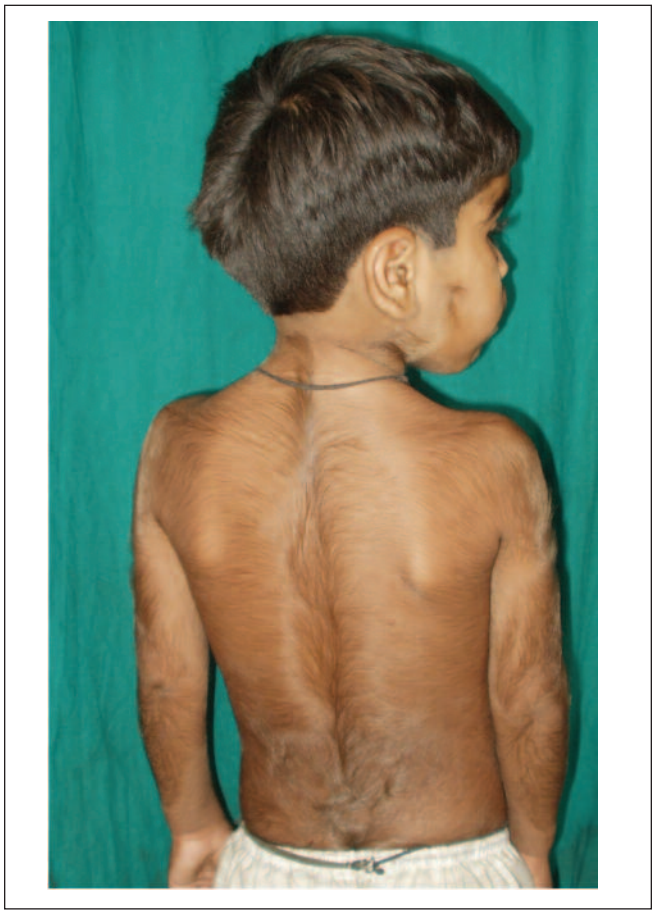

Figure 3. Generalized hypertrichosis covering back and arms

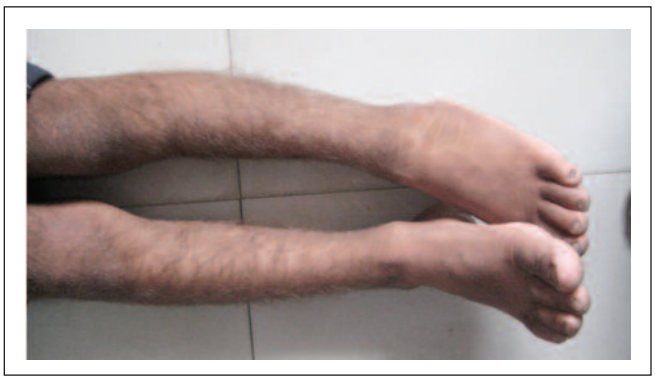

Figure 4. Generalized hypertrichosis covering legs

The anthropometric measurements were as follows:- Weight 15kg (25th percentile), height 127cm (5th percentile) and head circumference 40cm (50th percentile). The upper segment was 73 cm and the lower segment was 54cm. No organomegaly was found. The patient had a clear chest and normal heart sounds. His IQ was found to be within the normal range. Laboratory and radiographic tests were performed to evaluate any associated abnormalities. The results of routine blood tests, the blood chemistry panel and the liver function tests were normal. The radiographic findings of the anteroposterior view of the skull were normal, whereas in the lateral view, a vertical growth pattern was seen (Fig. 7). The posteroanterior chest, spine and hand-wrist radiographs were all normal. His karyotype was normal at a 500 band resolution (46XY).

The patient underwent a gingivectomy under conscious sedation in two sittings. During the procedure, all mobile primary teeth were extracted. Following the gingivectomy, all primary teeth were clinically erupted, and the masticatory and aesthetic profile was enhanced (Fig. 5). The post-

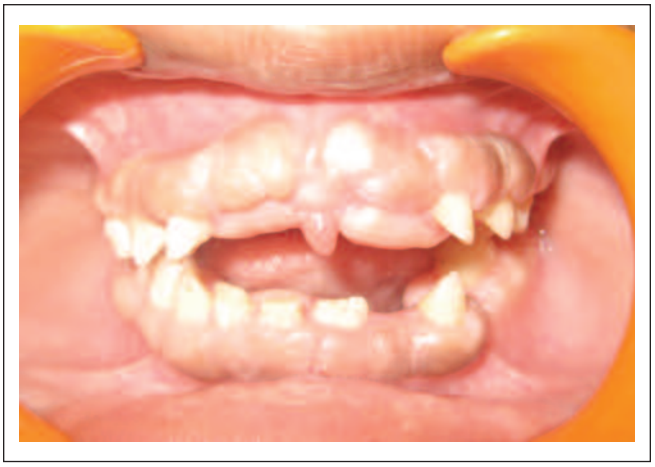

Figure 5. Post surgical Intraoral view

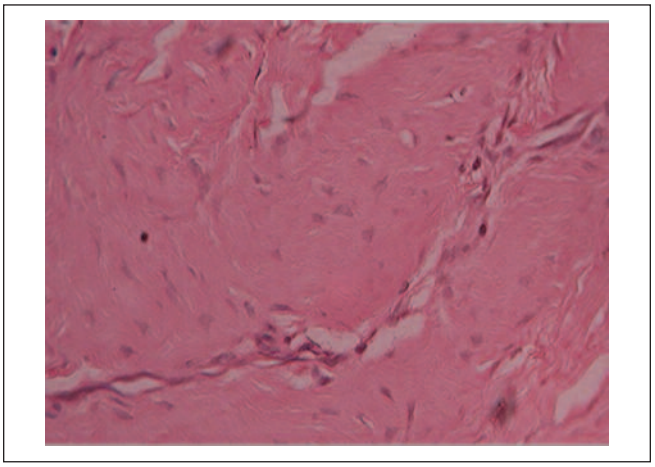

Figure 6. Photomicrograph showing connective tissue with dense collagen fibres and extravasated RBC's.

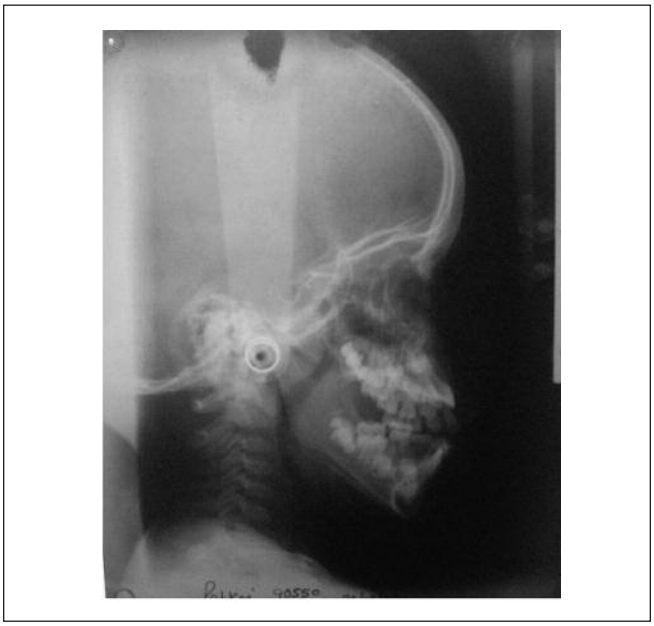

Figure 7. Lateral cephalometric radiograph showing vertical growth

operative healing was uneventful. Seven days after the gingivectomy, the sutures were removed, and oral hygiene instructions were given to the patient. The histopathological examination of the excised tissues revealed parakeratinized stratified squamous epithelium that was hyperplastic in the focal areas. The connective tissue was fibrocellular in nature with dense collagen fibers running in different directions. Mixed inflammatory infiltration with extravasated red blood cells (RBCs) were seen (Fig. 6). These features were suggestive of gingival fibromatosis. After two months of surgery, a mild gingival hyperplasia recurrence was noted and the patient was put on recall for every month.

DISCUSSION

Although the association of gingival hyperplasia and hypertrichosis has been reported on more than 50 occasions[5] several types of universal hypertrichosis have been described without gingival hyperplasia. Some of them are also recognized as genetic disorders (Table 1). Gingival hyperplasia has been reported as a symptom of a variety of syndromes. Usually, genetic expression is considered to be autosomal

Table 1.

	Ambras syndrome	Cantu syndrome	GF with distinct facial syndrome	GF with hypertrichosis syndrome	Bondenson and Miles (1993)	Canun (2003)	Sun et al. (2009)		Index case (2010)
Origin	Germany /Italy	Mexico			Mexico	Mexico	China	China	India
Inheritance		AR	AR	AD	AD	AD	AD	AD	-
FEATURES									
HAIR:									
.Congenital	+	+		+	+	+	+	+	+
.Universal	+				+	+	+	+	+
.Dark				+	+	+	+	+	+
.Light	+								
.Terminalis thick					+	+	+	+	+
.Straight									
.Long									
.Lanuga							+	+	
.Curly	+	+		+					
FACE									
.Normal									
.Dysmorphic	+		+		+	+			
.Coarse		+			+	+	+	+	+
.Abundant eyebrows		+	+		+	+	+	+	+
.Curly eyelashes									
.Long eyelashes					+	+			
.Abundant eyelashes					+	+			+
.Epicanthal folds				+					
.Hypoplastic nose									
.Broad nose									
.Anteverted nostrils					+	+			
.Thick lips	+				+	+	+	+	+
.Prognathism					+	+			
.Gingival hypertrophy	+				+	+	+	+	+
.Osteochondro-dysplasia		+	+	+	+	+		+	+
IQ		low			normal	normal	normal	normal	normal
OTHERS									
.Accessory nipple	+								
.Polydactyle									
.Cardio megaly	+	+							

dominant, and sporadic cases without family history are probably due to genetic mutation[11] as in our case.

Several types of hypertrichosis are reported, based on the distribution of excess hair and the supposed etiology. The pattern of hairiness corresponds to the condition seen in congenital hypertrichosis lanuginosa or Ambras syndrome. Hypertrichosis *universalis* congenita[13,14] (generalized congenital hypertrichosis) was identified as an x-linked entity in a very large family.[15] Hypertrichosis associated with osteochondrodysplasia has an autosomal recessive pattern of transmission and is identified as Cantu syndrome.[9,10]

In our patient, the main feature was generalized hypertrichosis with severe gingival fibromatosis. There was no indication of the particular cause or anomaly, and his family pedigree did not suggest an inherited condition.

A literature review revealed many cases of gingival hyperplasia accompanied by characteristic hypertrichosis. Therefore, it was reasonable for us to relate hairiness and gingival hypertrophy[16-21] and report a close relationship between the two phenomena.

Many cases with normal facial features have been reported in the literature.[22,23] Ambras type dysmorphic features[6,24] and a even Coarse facial appearance have been reported.[2,25]

The most striking differences from other forms of hypertrichosis are the craniofacial features. The inherent nature of our patient and the one described by Canun *et al,*[11] are similar:

a) Universal distribution of congenital terminal hypertrichosis
b) Gingival hyperplasia
c) very characteristic, coarse craniofacial appearance.

Most of the cases mentioned were of Mexican origin. To the best of our knowledge this is the first reported case from India. Further research is required to determine the etiopathogenesis of this condition.

CONCLUSION

The main feature of this patient was generalized hypertrichosis with gingival fibromatosis without reference to a particular cause or accompanying anomaly and no credible family pedigree to suggest an inherited condition.

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