Gingival Fibromatosis with Hypertrichosis Syndrome: Case Series of Rare Syndrome

Abstract
Gingival fibromatosis with hypertrichosis syndrome is an extremely rare genetic condition characterized by profound overgrowth of hair and gums, as well as other variable features. Gingival fibromatosis is characterized by a large increase in the gingival dimension which extends above the dental crowns, covering them partially or completely. They were found to have a genetic origin, may also occur in isolation or be part of a syndrome, or acquired origin, due to specific drugs administered systemically. Congenital generalized hypertrichosis is a heterogeneous group of diseases with continuing excessive growth of terminal hair without androgenic stimulation. It has informally been called werewolf syndrome because the appearance is similar to that of a werewolf. Various syndromes have been associated with these features such as epilepsy, mental retardation, cardiomegaly, or osteochondrodysplasia. As so far very few cases have been reported in literature, we are reporting a series of three cases with management of the same. The excess gingival tissues, in these cases, were removed by conventional gingivectomy under general anesthesia. The postoperative result was uneventful and the patient’s appearance improved significantly. Good esthetic result was achieved to allow patient to practice oral hygiene measures. Though this is not a serious condition clinically, psychosocial trauma cannot be neglected owing to the cosmetic disfigurement it produces.

Keywords: Gingival fibromatosis, hypertrichosis, werewolf syndrome

Introduction
Gingival fibromatosis with hypertrichosis (GFH) syndrome is an extremely rare condition of genetic origin characterized by the excessive growth of gingival tissue with abnormal facial and body hairs. Gingival diseases of genetic origin are uncommon conditions and their most common clinically evident form is known as hereditary gingival fibromatosis (HGF). It may occur singly or in association with other inherited syndromes.[1] This condition is also known as elephantiasis gingivae, diffuse fibroma, familial elephantiasis, idiopathic fibromatosis, HGF, and congenital familial fibromatosis. It is inherited as an autosomal dominant trait and has an incidence of 1:350,000.[2,3] Hypertrichosis may be congenital or acquired. People with congenital hypertrichosis were often referred to as “wolf men,” “werewolves,” and “ape-men” in the 19th century, and perhaps even today, they are crowd-drawers at sideshow acts.[4] Various syndromes have been associated with gingival fibromatosis and hypertrichosis condition which have other features such as epilepsy, mental retardation, cardiomegaly, or osteochondrodysplasia.

Case Reports
Case 1
A 10-year-old girl reported with the complaint of everted lips and swollen gums with coarse hair present over the entire body since birth. She also complained of delayed eruption of permanent teeth. There was no medical history and no systemic abnormalities. The girl was born out of nonconsanguineous marriage and she had average growth and achieved normal developmental milestones. Hypertrichosis, with extremely bushy hairs over the chin, cheeks, and also in the mid forehead with retruded nasal bridge, is seen. Profuse growth of dark brown hair was present on both the upper and lower limbs and in the mid back. On intraoral examination, gingival enlargement involving the upper dentition was found. Teeth were completely covered with gingival overgrowth and lips were incompetent due to abnormality in thickness. In both the jaws, dentition appeared to be normal, as it led to

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functional disturbances while eating and chewing and esthetic concern. The treatment plan was to do debulking of the lips and gingivectomy procedures [Figure 1a-d].

**Surgical procedure**

Under general anesthesia and nasotracheal intubation, lidocaine gel was applied 30–45 min before the procedure. The lip was held taut between the thumb and forefinger. Using a number 15 blade, incisions were first made along the newly outlined margin, excess tissue was removed and suturing was done. Generally, gingivectomy is done when there is moderate gingival enlargement or when aberrations are present or when there is asymmetrical or unesthetic gingival topography. Pocket depth was measured by inducing bleeding points. Initial incision was placed apical to the bleeding point and 45° bevel was given to the root. The gingival tissues covering the teeth were removed placing an incision leaving a 2 mm gingival margin around the teeth and were excised. Excess gingival tissue was excised and zinc oxide eugenol pack was placed. After 5–14 days, surface epithelialization was completed.

Immediate result of the treatment was a dramatic cosmetic improvement in her outlook and socialization also greatly improved [Figures 2-3].

**Case 2**

A 6-year-old girl presented with the chief complaint of gum enlargement over the upper and lower teeth regions. The girl was born out of nonconsanguineous marriage and presented with excessive hair on the whole body. She had average growth and achieved normal developmental milestones. Clinical examination showed a mentally alert, well-built girl with hypertrichosis, presenting with extremely bushy eyebrows with downy growth of hair on the cheeks, chin, upper lip, and chin. On intraoral examination, generalized gingival hyperplasia was present covering the anatomical crowns of the deciduous teeth as well as the permanent teeth. Lips were incompetent due to gingival hyperplasia [Figures 4-6] (for surgical procedure, refer Case 1).

**Case 3**

An 8-year-old girl was brought to our hospital for the treatment of overgrown gums in the upper and lower teeth. She was born out of a nonconsanguineous marriage with an uneventful antenatal and postnatal history, as described by the mother. There were no associated systemic complications in the child. On examination, terminal hair had a generalized distribution over the entire body sparing the palms, soles, and the mucosal surfaces.
examination revealed pink, hyperplastic, and stippled gingiva with an altered dental architecture covering almost all teeth except for occlusal surface of all the four molars, and she was planned for gingivectomy procedure (for surgical procedure, refer Case 1).

Discussion

Congenital generalized hypertrichosis (CGH) represents a heterogeneous group of conditions that are phenotypically and genetically distinct; it is characterized by excessive universal hair growth as the hallmark feature that is disproportionate when compared to normal individuals of that age, sex, and race and it is not dependent on androgenic hormones. Various genetic syndromes have been associated with hypertrichosis condition such as Ambras syndrome, Barber Say syndrome, and Cantu syndrome.

Gingival fibromatosis may be familial or idiopathic. The familial variation may occur with a number of other inherited syndromes when it could be associated with some of the following, for example, Zimmermann–Laband syndrome, Murray–Puretic–Drescher (juvenile hyaline fibromatosis), Rutherford, Cross, Cowden syndrome, multiple hamartomas, and tuberous sclerosis.

The gingival overgrowth usually begins at the time of eruption of permanent dentition or less frequently with the eruption of primary dentition. In our cases, case 1 reported that the gingival enlargement started since birth but it almost covered the entire teeth at the time of teeth eruption of permanent dentition, and hence there was delayed eruption of permanent dentition. The extent and severity of fibromatosis in this case has covered almost all teeth, thereby causing difficulty in mastication and speech.

The microscopic features of the present cases were classic of gingival fibromatosis. The tissue showed excess amount of collagen in an avascular corium with the overlying parakeratinized epithelium.

The treatment need varies according to the degree of severity. The relative increase in the gingival mass necessitates the need for surgical intervention owing to the functional and esthetic compromise.

Whenever possible, the treatment should be performed after the complete eruption of permanent dentition. Recurrence is a common feature over varying periods. One report indicated that there is less chance of recurrence if the gingivectomy is delayed until permanent dentition is in place.

Conclusion

GFH syndrome is a rare disorder characterized by varying degrees of attached gingival overgrowth with excessive facial and body terminal hairs. Esthetic and functional compromises of gingival enlargement often demand surgical intervention, albeit recurrence cannot be predicted. Good esthetic result was achieved without the recurrence of gingival overgrowth. After treatment, regular recalls are necessary to evaluate oral hygiene and stability of surgical treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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