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IDIOPATHIC GINGIVAL ENLARGEMENT IN A FAMILY WITH AUTOSOMAL DOMINANT INHERITANCE: A CASE REPORT

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Abstract:

Idiopathic Gingival Enlargement is a hereditary condition; it can be expressed in both Autosomal Recessive and Autosomal Dominant inheritance. Here a case of Idiopathic gingival enlargement has been reported in a family with an Autosomal Dominant inheritance with other clinical features like massive hair distribution over the body of a female, bimaxillary protrusion and enlarged palm and fingers with flat feet. The atypical massive hair distribution all over the body and face making this case to **simulate (mimicking) a missing link between primates and homosapiens. This clinical condition has never been reported before.**

Introduction

Idiopathic Gingival Enlargement is a very rare variety of Gingival Enlargement which is a hereditary disorder, regarding which data has been already reported several times. It has been reported that it can be expressed as one Autosomal Recessive inheritance and one Autosomal Dominant inheritance. It has also been reported that, Very often Idiopathic Gingival Enlargement associated with Hypertrichosis for example; Lab and syndrome, Rutherford syndrome, Cross syndrome and Ramon syndrome. All these syndromes are having one common character i.e. Idiopathic Gingival Enlargement but not to the extent that we are going to report it here. Here in this case the characteristic atypical hair

distribution over face , bimaxillary protrusion and a typical shape of palm and feet with massive hair distribution over the body is not matching to any of these already existing syndromes. Those syndromes are associated with Corneal fibromatosis, Cherubism, Epilepsy, Mental retardation, Syndactyl, Nail hypoplasia and Hypersplenism. Here the enlargement is so massive which is causing difficulty in chewing, swallowing and causing ultimately nutrition problem..

Case History :

A fourteen year old female presented with symptoms of swelling of both maxilla and mandible to such an extent which simulate (mimic) a missing link between primates and homosapiens, Vide fig-1. The growth of maxilla and mandible was noticeable since she was three year age as reported by her parent. The massive abnormal growth of gingiva leads to difficulty in chewing, swallowing, speaking and unsociable appearance. Her lip is incompetent and whole body and face is covered with hair. She had been consulted to many doctors but due to the rarity of this condition and diagnosis problem she was not guided properly. Her parent could not feel to contact Doctors for last 11 years because of both lack of facility , knowledge and prejudice problem. They think their problem to be a curse of God as they belong to a tribal community, residing in remote part of Orissa inside one sanctuary known as sata kosia sanctuary andThat tribal community is called KANDHA community. So at the age of 14 years she reached the Dental Wing , SCB Medical College and referred to the department of Periodontology for need full action.

Family History :

One of her brother was also suffering from the similar condition and died because of nutrition problem as reported in his death certificate. Her father is also suffering from the same idiopathic gingival enlargement but degree of expression is much less and hair distribution over his face is not similar to that of his daughter as shown in fig- . Her mother and another brother on the other hand carry no such clinical features and they are absolutely normal

Psychological History:

Her psychological health is absolutely normal with normal IQ; she could go up to Std. V because of facilities and social problem that was because of hesitation of other children to sit with her in the school.

Menstrual History

Her menstrual cycle is also normal.

General Examination:

Massive Atypical hair distribution through out the body and face. Normal and good body build. No pallor, no icterus, no cyanosis, no clubbing, no lymphadenopathy with all other normal vital signs.

ON LOCAL EXAMINATION:

Extra Oral:

Hair through out face Incompetent lip due to Protrusion of the enlarged gingivae, Skin normal dark complexion.

Dimensional Analysis of Face:

Dimension of Ala tragus to right labial fold = 16cm

Dimension of Ala tragus to left labial fold = 16cm

Length of lip right corner to left corner = 11cm

Chin to tip of nose = 12cm

Right ala of nose to left = 7cm

Chin to Glabella = 15cm

Glabella to apex of skull = 10cm

Medial Canthus to Medial Canthus = 4cm

Medial Canthus lateral Canthus = 4cm

Intraoral Examination :

She has no visible teeth Mucosa pink color The Gingival growth is firm non tender non inflammatory. Attached gingiva so massively over grown which covered the whole of the jaw and it is pink, firm, leathery in consistency with lobulated pebbled surface. The jaw is distorted due to the bulbous enlargement. Teeth are almost completely covered by about 1-2 1/2 inches of the grown tissue of attached gingiva as shown in Tongue is slightly enlarged.

Systemic Evaluation:

Systemic Evaluation suggests no internal organ anomaly, no cardiac and respiratory anomaly, and no endocrine disorders.

Blood investigation:

she has normal serum Na⁺, K⁺, Normal LFT with other findings also normal, Except haemoglobin level 8gm%.

Radiological Investigation:

Show the presence of all the permanent teeth. Some retained deciduous teeth all are completely covered by the enlarged mass shown in the lateral cephalogram i.e. fig no-10

Histopathological Study:

Show hypercellular, densely arranged collagenous bundles of fibers
Long thin tubular rete pegs .

Discussion:

This case of Idiopathic Gingival Enlargement with severe Hypertrichosis, associated with other clinical feature such as bimaxillary protrusion, flat feet, comparatively enlarged palm and fingers not matching to any of the mentioned syndromes those have been already reported, there fore this is a new phenotype expression. Even if Labands syndrome carries similar clinical features here this case is differentiated from this syndrome because of association of 1- bimaxillary protrusion, 2- massive hair distribution over the entire body with classic hair distribution over the face. 3- Comparatively enlarged palm and fingers. This case is clearly a case of autosomal dominant inheritance as in this family we have found, out of three children from a parent constituting one affected phenotypic male and a normal unaffected phenotypic female, when one male child and another female child are affected and third male child is absolutely normal with out any clinical feature like her mother. So the 50% affected children going absolutely in favor of AUTOSOMAL DOMINANT INHERITANCE. Bimaxillary protrusion is evident from lateral cephalogram .The histopathological features are going in favor of Idiopathic gingival enlargement.

Treatment and Management:

Because of Patient's convenience, massiveness of the enlargement and the risk of bleeding from the vessel like incisive vessel and Grater palatine vessel the management can be done in phase wise manner, With an objective of restoring normal contour of gingiva, exposure of the teeth and bringing back the normal facial profile.

Conclusion:

This case of Idiopathic Gingival Enlargement is attributed to a hereditary disorder reflected as Autosomal Dominant inheritance. This massiveness of enlargement and atypical hair distribution over face and body and other associated clinical feature simulating (mimicking) a missing link between primates and homosapiens that is not matching to any of the previous syndromes that has already been reported with more or less similar clinical features. This case of idiopathic gingival enlargement will be in need of the detection of risk allele responsible for this phenotype expression in further research. The future research could be directed towards the therapeutic that could change dramatically by one of the recombinant DNA and monoclonal antibody technology strategy to embarrass the approach are inevitable so gene therapy has been champion as a new and exiting approach for the prevention of such diseases.

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Figure Legends

- Figure 1 - patients suffering from idiopathic gingival enlargement
Figure 2 - massive enlargement causing problems during chewing
Figure 3 - massive idiopathic gingival enlargement of upper and lower jaw
Figure 4 - Patient showing normal IQ and massive hair distribution over hands
Figure 5 - posterior teeth embedded 3[1].5-4cm below the massive
Figure 6 - patient father showing protrusion of face
Figure 7 - patient's father lateral view showing protruded face due to enlargement.
Figure 8 - patient's father showing similar idiopathic enlargement of attached gingival
Figure 9 - Histopathological slide showing long slender tube shaped rete pegs with densely arranged collagen bundles
Figure 10 - Lateral cephalogram showing bimaxillary protrusion with over lying soft tissue



Figure 1



Figure 2



Figure 3



Figure 4



Figure 5



Figure 6



Figure 7



Figure 8



Figure 9



Figure 10