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Case Report

Gorlin Goltz Syndrome: A Case Report

Dr.Gagandeep Singh[1], Dr. Taranjot Kaur[2], Gulsheen Kaur Kochhar[3], Dr. Banmeet Singh Uppal[4]

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Affiliations:

1. Private Practioner, Department of Oral & Maxillofacial Surgery, Chandigarh, India
2. Reader, Department Of Pedodontics & Preventive Dentistry, Swami Devi Dyal Hospital and Dental College, Barwala, Panchkula, Haryana, India.
3. Senior Lecturer, Department Of Pedodontics & Preventive Dentistry,Swami Devi Dyal Hospital and Dental College, Barwala, Panchkula, Haryana,India
4. Medical Officer, Mini PHC, Surtapur, Punjab.

Corresponding Address:

Dr.Gulsheen Kaur Kochhar
Department of Pedodontics & Preventive Dentistry
Swami Devi Dyal Hospital and Dental College
Barwala, Panchkula,Haryana
Email- gulsheenuppal@gmail.com

ABSTRACT

Gorlin Goltz syndrome is a rare autosomal dominant disorder which includes a variety of possible abnormalities namely cutaneous, dental, osseous, ophthalmologic, neurologic and sexual disorders. One such case of a north Indian female patient, affected with this rare syndrome with minimal expression of the anomalies is presented along with a brief account of review of literature. We also focussed on the etiology and the various treatment modalities of multiple odontogenic keratocysts present in the maxilla and mandible.

Keywords- Gorlin Goltz syndrome, Odontogenic Keratocysts.

INTRODUCTION

Gorlin Goltz syndrome has received several names such as "basal cell nevus syndrome", "nevoid basal cell carcinomas syndrome", "multiple basal epithelioma", "jaw cysts" and "bifid rib syndrome" and was first described in 1960.[1] This syndrome may be manifested by some or all of the following clinical features; odontogenic keratocyst, multiple basal cell carcinomas of the skin, rib and vertebrae anomalies, intracranial calcification, skeletal abnormalities such as bifid ribs, kyphoscoliosis, calcification of falx cerebri, distinct facies presenting as frontal and temporo-parietal bossing, hypertelorism, and mandibular prognathism. It affects male and female equally and is more commonly seen during the first to third decades of life.[2]

Multiple odontogenic keratocysts are seen in 75% of patients affected with *Gorlin Goltz syndrome* and are the most common finding.[3] They may be found on incidental radiographic examination or the patient may present with symptoms in case of secondary infection. They can appear as both multilocular and unilocular radiolucencies of the posterior body, angle or ramus of the mandible. Evidence of displacement of developing teeth, root