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Research Article

GORLIN GOLTZ SYNDROME – AN ATYPICAL CASE REPORT

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ABSTRACT

Occurrence of cysts and osteomas in the maxillofacial region is quite rare and occurs as a manifestation of some syndrome. The diagnosis of such syndrome can be easily missed which lead to poor patient follow up. We report a case of 30 year-old male presenting with this syndrome presenting withGorlin-Goltz syndrome with unusual oral manifestations.

INTRODUCTION

Gorlin – Goltz Syndrome is inherited as autosomal dominant disorder. Numerous Basal cell carcinomas, musculoskeletal deformation and multiple keratocysts are frequent characteristics of this syndrome (2).Jarish and White first described this syndrome in 1894 (7-8). In common population this syndrome occurs approximately in 1in 50000 to 150000 (4). Several names have been used to describe this syndrome such as, Gorlin syndrome, Jaw – cyst-Basal Cell Nevus Bifid Rib syndrome, Multiple Basilomasyndrome, Fifth Phacomatosis Nevoid Basal cell carcinoma syndrome and Basal Cell Nevus Syndrome.(9)

Case Report

A 30 year old male patient came to the Department of Oral and Maxillofacial surgery, Faculty of Dental Sciences, Sri Ramachandra University with a chief complaint of missing tooth in the upper front region of the jaw. Patient gave a history of road traffic accident one year ago following which he lost his upper front teeth. Patient was provided with a temporary partial denture by a local dental surgeon during that period. Patient has no preexisting medical illness. On general examination, his vitals were stable.

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There is no signs of pallor, icterus, cyanosis, clubbing or pedal edema .Intra oral examination revealed missing right central incisor. No evidence of swelling or tenderness present in the maxilla and mandible. Patient's occlusion was stable .Mouth opening and lateral temporomandibular movements were satisfactory.Orthopantomogram was taken for implant planning, which surprisingly revealed impacted supernumerary teeth with a radiolucent lesion (A) surrounded by a well-defined sclerotic border in relation to 11 and 21 region. There is also radioopaque structure (B) evidentin relation to 44 and 45 region as shown in Figure 1. Considering the size of the cystic lesion in the maxilla and the radio opaque lesion in the mandible, surgical enucleation of the cystic lesion and removal of mesiodens with simultaneous removal of the radio opaque mass in the mandible was planned under general anesthesia. A crevicular incision was placed from 12 to 23 regions. Mucoperiosteal flap was elevated, bone guttering was done in relation to 21 to 22 region using a No 6 round bur. On enucleation of the cyst, to our surprise there was evidence of dirty white cheesy material from the cystic lesion. Based on the clinical finding, an immediate chemical cauterization was done with Cornoy's Solution with a suspicion of odontogenic keratocyst. Once hemostasis was achieved, closure of the wound was done with resorbable sutures. A vestibular incision was placed from 43 to 46 region, mucoperiosteal flap elevated, bone guttering was done to reach to the radio opaque mass .A surgical excision of the radio opaque mass was done and closure was done with

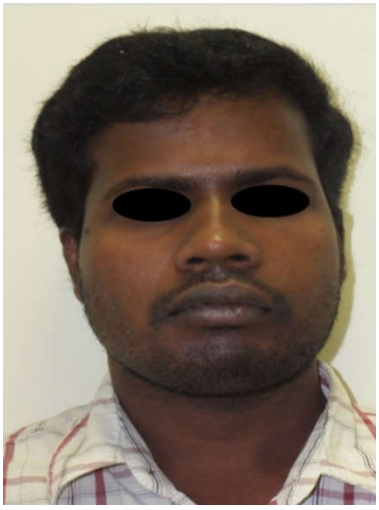
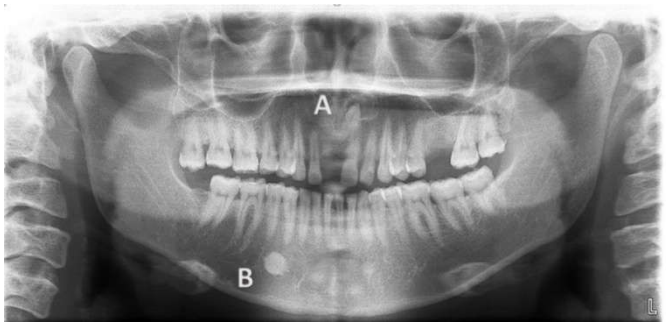


Figure 1. Patient Photo



Figures 2. Pre-Operative Orthopantomogram showing the impacted supernumerary tooth in relation to 21 region (A) and radio opaque mass in relation to 44 and 45 region (B)

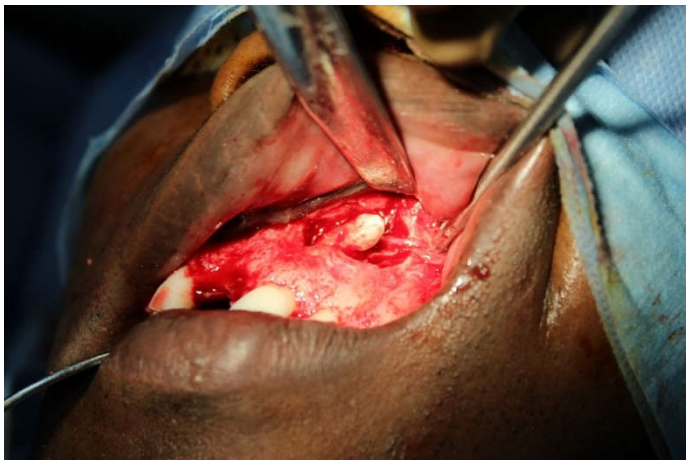


Figure 3. Intra Operative View – Impacted Supernumerary tooth with cystic lesion

Resorbable suture. The cystic lesion, tooth and the bony mass was sent for histopathological examination. Histopathology showed the cystic lumen lined by thin corrugated parakeratotic stratified squamous epithelium .Connective tissue wall shows cholesterol clefts with calcification suggestive of odontogenic keratocyst with inflammation. The decalcified section of the radio opaque mass is suggestive of compact osteoma.The Chest X Ray which was taken revealed bifid third rib on the right side.

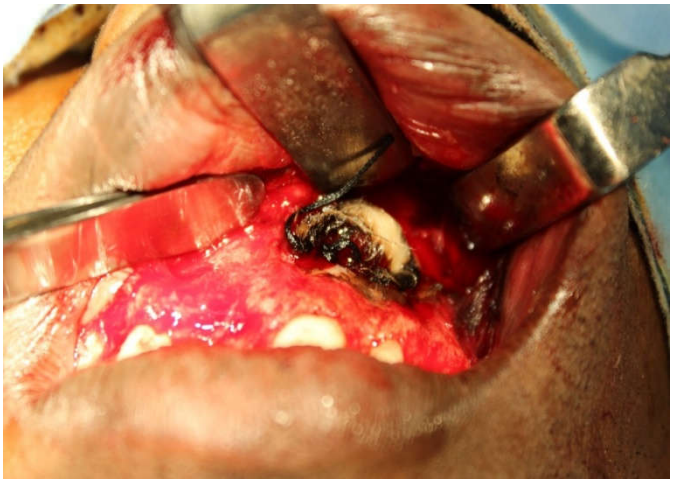


Figure 4. Chemical Cauterization with Carnoy's Solution

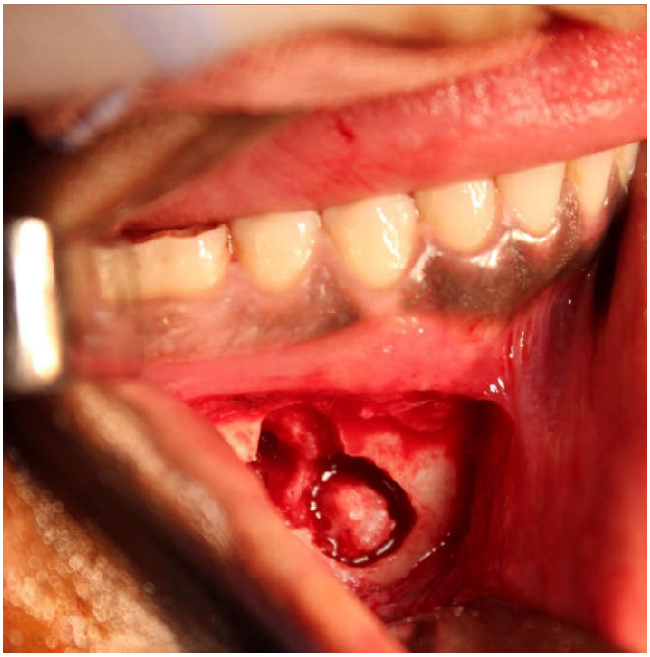


Figure 5. Intraoperative View – Osteoma in relation to right lower premolars

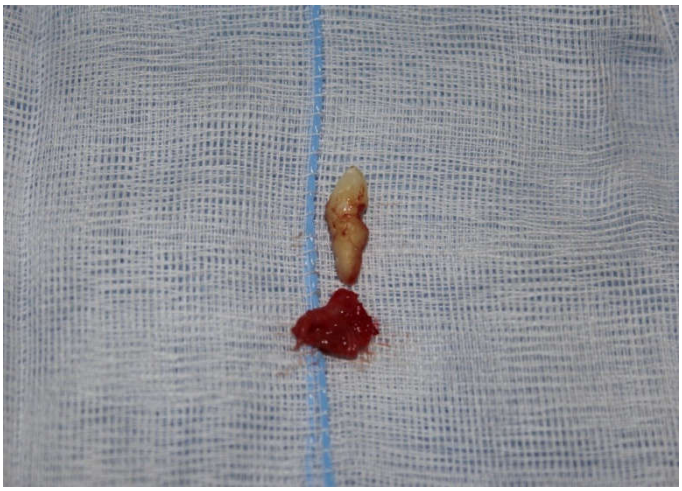


Figure 6. Removed Mesiodens & Cystic Lining

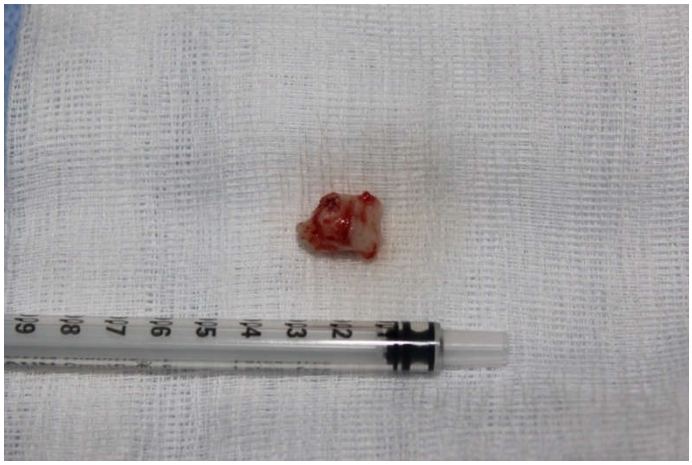


Figure 7. Removed Osteoma mass

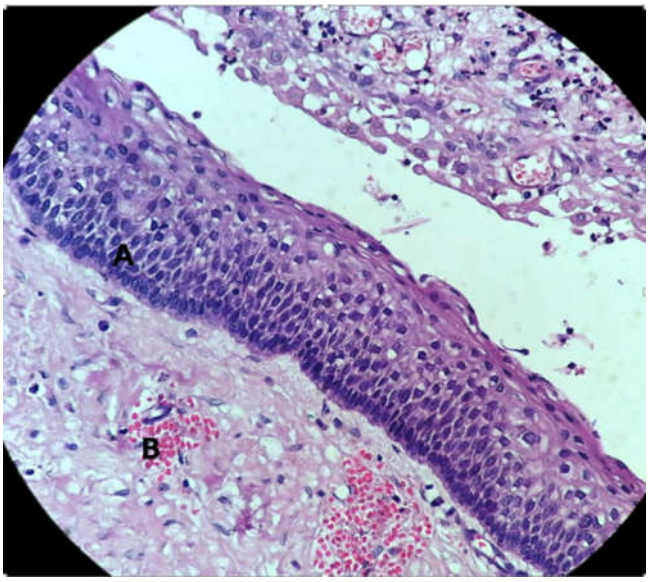


Figure 8. Histopathological specimen showing Cystic lumen lined by corrugated parakeratinized stratified squamous epithelium with characteristic tomb stone appearance of the basal cell layer with reversal polarity (A0.The fibrous capsule shows few inflammatory cells and extravasated red blood cells (B)-40x

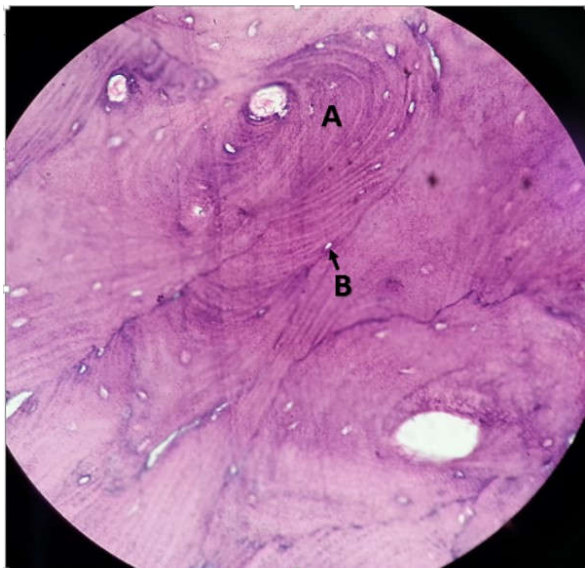


Figure 9. Histopathological specimen showing compact osteoid tissue (A) with osteocytes(B)present within the laminae -40 X

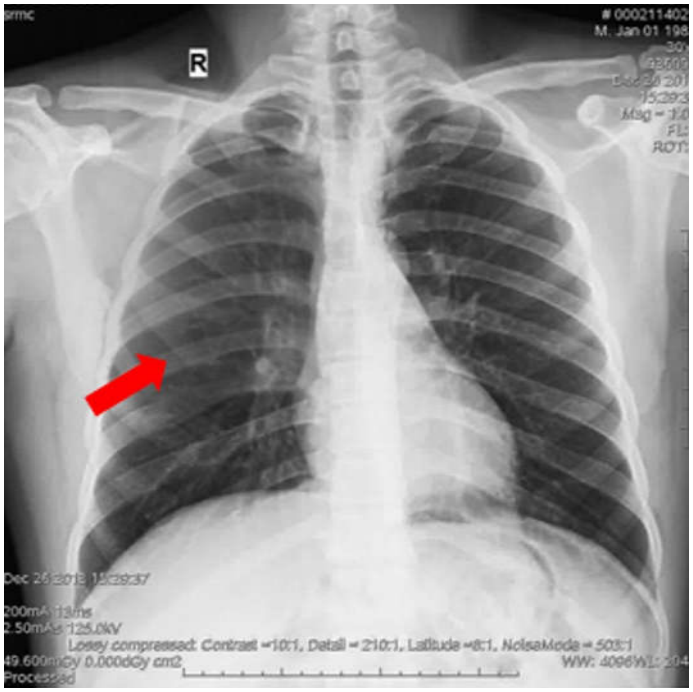


Figure 10. Chest radiograph showing bifid right third rib

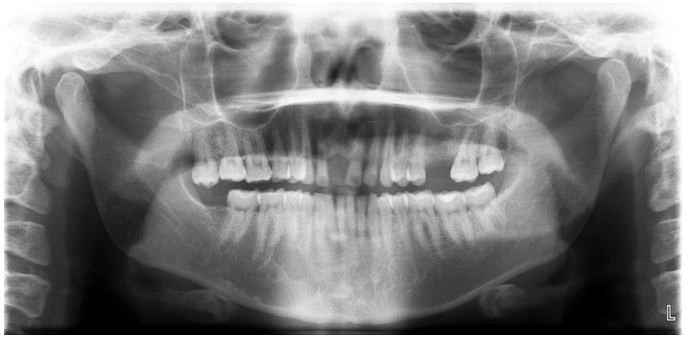


Figure 11. Post-Operative Orthopantomogram showing complete removal of the impacted supernumerary tooth and osteoma removal

DISCUSSION

GorlinGoltz Syndrome is an inherited disorder occurs due to mutations in the PTCH1 (Patched 1) gene(4).To arrive at a diagnosis of Gorlin-Goltz syndrome, a patient should atleast present with 2 major or one major and two minor criteria.(10). The major criteria includes histologically proven any odontogenic keratocyst, ectopic calcifications, basal cell carcinomas, palmar or plantar pits. Minor criteria includes Cleft lip and palate, medulloblastoma, impacted teeth and/agenesis cardiac or ovarian fibroma, congenital skeletal anomalies, frontal bossing, dental ectopic position, fibrosarcoma of the jaws. Our patient had odontogenic keratocyst, impacted tooth, bifid rib and osteoma of mandible which were cardinal signs of this syndrome, presenting with two major and one minor criteria thereby confirming to have Gorlin-Gotlz syndrome.

Conclusion

Diagnosis of this syndrome requires a multiunit approach which comprises of removal of cysts and tumors .Protection from sun is important and counseling has to be given for the patients so

that further manifestations of disorders can be minimized. Patients with Gorlin-Gotlz syndrome requires constant follow – up and protection from sun due the increased risk of basal cell carcinoma. Genetic risks should be counselled.

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