*International Journal Of*

**Recent Scientific Research**

**ISSN: 0976-3031**

**Volume: 7(6) June -2016**

# MULTIPLE JAW CYSTS IN ASSOCIATION WITH GORLIN GOLTZ SYNDROME-A CASE REPORT

**Mohammad Shakeel., Mohamad Imran., Azhar Khan and Mir Shayan**

THE OFFICIAL PUBLICATION OF
INTERNATIONAL JOURNAL OF RECENT SCIENTIFIC RESEARCH (IJRSR)
http://www.recentscientific.com/ recentscientific@gmail.com

ISSN: 0976-3031

*Available Online at http://www.recentscientific.com*

*International Journal of Recent Scientific Research*
*Vol. 7, Issue, 6, pp. 11994-11996, June, 2016*

**International Journal of Recent Scientific Research**

**Case Report**

# MULTIPLE JAW CYSTS IN ASSOCIATION WITH GORLIN GOLTZ SYNDROME-A CASE REPORT

**Mohammad Shakeel[1]., Mohamad Imran[2]., Azhar Khan[3] and Mir Shayan[4]**

**ARTICLE INFO**

***Article History:***

Received 05th March, 2016
Received in revised form 08th April, 2016
Accepted 10th May, 2016
Published online 28st June, 2016

***Key Words:***

Real world evidence, Real world data, Registries, Healthcare, Payers, Pharma

**ABSTRACT**

The simultaneous occurrence of multiple cysts in both the jaws of a patient is rare and it usually occurs as the manifestation of a syndrome. Whenever multiple cysts are found, it is necessary to rule out an association with any syndrome, as the chances of recurrence are very high and a periodic follow-up is required for such patients. In our patient, cyst enucleation was done previously, but the association with the Gorlin-Goltz syndrome was missed and no follow-ups were advised.
Gorlin and Goltz first described the spectrum of features which are associated with the Gorlin-Goltz syndrome or the Nevoid Basal cell carcinoma syndrome (NBCCS), in 1960. It is an autosomal dominant disorder with a genetic locus on chromosome subbands and bands 9q22.3-q31.[1]It comprises of skeletal features such as the bifid rib, frontal and parietal bossing and mandibular prognathism and cutaneous abnormalities such as multiple basal cell carcinomas and palmar and plantar keratosis. NBCCS can also include concomitant hypertelorism, mental retardation, strabismus, calcification ofthe falxcerebri and medulloblastomas.[2]
We are discussing here a case of two mandibular cysts withexpression of many of the features of NBCCS and are briefly reviewing the features of the Gorlin-Goltz syndrome.



## INTRODUCTION

A 20 year old male was reffered to our department for evaluation of jaw cysts. As per the medical records, the patient had undergone multiple jaw surgeries forcyst enucleation in the past. On general examination, the patient was found to be well built with significanthypertelorism and prominent supra-orbital ridges (Fig 1).

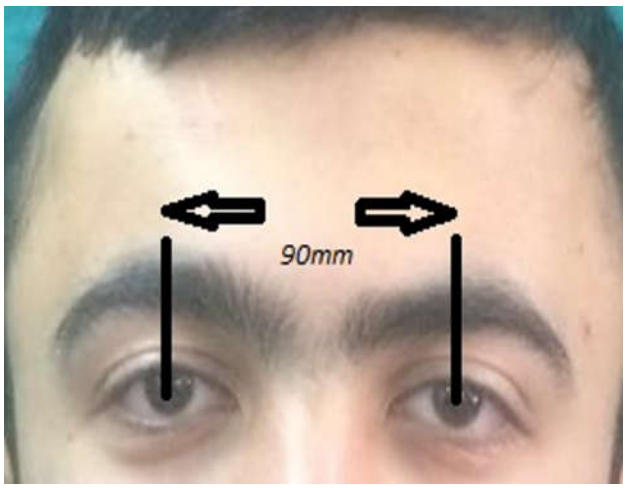


**Fig.1 (a)** Depictiing a significant hypertelorism with interpupillary distance of approx.90mm.

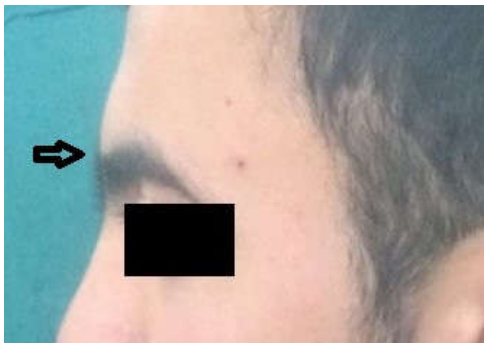

**Fig.1** (b) There are prominent supraorbital ridges seen clearly on lateral view.

There was syndactyly of right foot in relation to second and third phalanges. (fig 2). An Orthopantomogram (OPG) of the patient showed two large cysts in the mandible (Fig 3), the larger one extending from lower left second premolar to the lower right canine and the smaller one lies distal to mandibular second molar involving the lower third of ramus. Considering the possibility of the Gorlin- Goltz syndrome, further evaluation was done with chest radiographs, which revealed a bifidrib (Fig 4), postero anterior skull radiographs which revealed the falxcerebri calcifications (Fig 5). The cysts were enucleated and Cornoy's solution was applied to the bony cavities to prevent recurrence.

*Corresponding author: **Mohammad Shakeel***

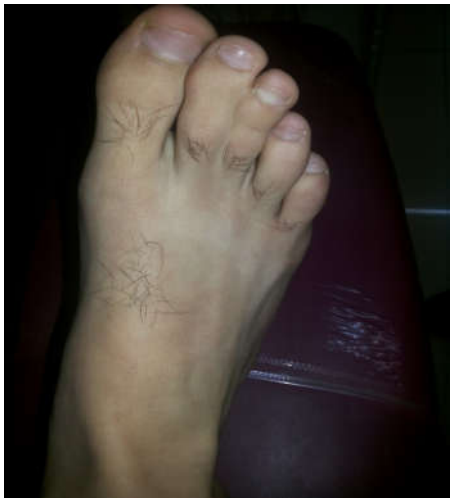

**Fig.2** The second and third phalanges of right foot with syndectyly.

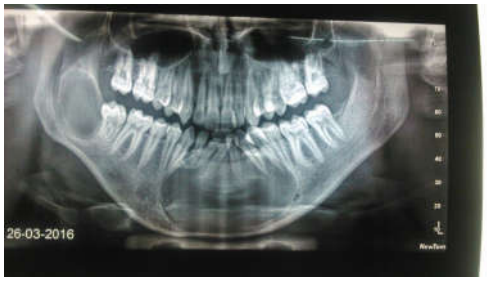


**Fig.3** OPG depicting two well defined cystic radiolucencies in the lower jaw with displacement of roots of teeth.

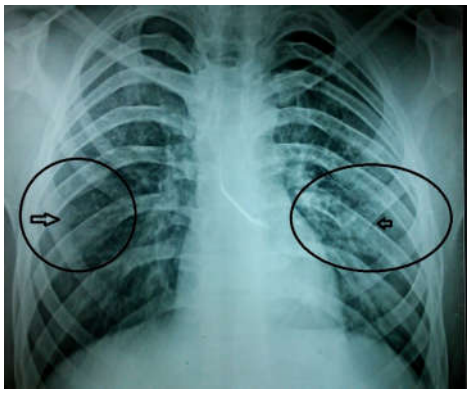

**Fig.4** Chest x ray of the patient with evidence of bifid ribs at encircled areas.

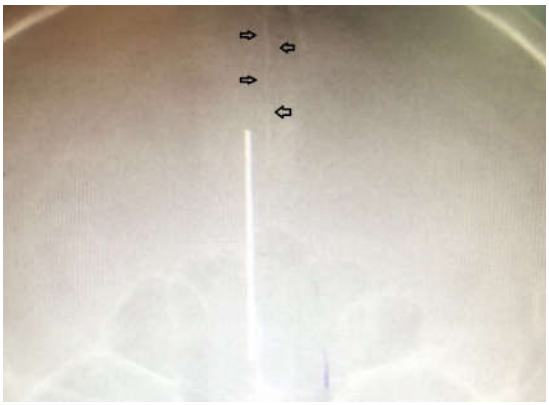

**Fig.5** Postero anterior skull x ray depicting faint calsification of falxcerebri.

The patient was advised a follow up every month in our OPD and periodic checkup in neurology and dermatology departments.

## DISCUSSION

The diagnosis of the Gorlin-Goltz syndrome is made clinically by using the criteria which are suggested by Evans and others.[3] Two major or one major and two minor criteria should be satisfied for a positive diagnosis. Our patient had three major features of NBCCS, namely bifid rib, multiple odontogenickeratocysts in the jaw and lamellar calcification of the falx and minor features such prominent supraorbital ridges and hypertelorism, thus suggesting it to be a case of the Gorlin-Goltzsyndrome. Regarding the site predilection, OKCs which are associated with NBCCS are more common in the mandible with 69% involvement, as compared to 31% in the maxilla. In the mandible, 43% OKCs occurs in the molar ramus region, followed by 18% in the incisor-canine area. In the maxilla, 14% OKCs were found to occur in the incisor-canine area, followed by maxillary tuberosity with 12%, 7% in the mandibular premolar region and 3% in the maxillary premolar region. Regarding the male to female ratio, it was 1:0.62 for OKCs which were not associated with NBCCS and 1:1.22 for OKCs in NBCCS. This shows that simple keratocysts are more common in males, but that more females seem to have NBCCS.[4] Based on histopathological studies, parakeratinization intramural epithelial remnants and satellite cysts were found to be more frequent among the OKCs which were associated with NBCCS thanin the solitary keratocysts.[5] The term "multiple cysts" does not necessarily mean that the patient must have more than one cyst at a given time; rather it refers to the occurrence of cysts over the life time of the patient.[6] There is no specific laboratory test to diagnose NBCCS, although the affected patients may have high levels of cyclic adenosine monophosphate and impaired phosphate diuresis on parathormonechallenge.[7]

The treatment of the GorlinGoltz syndrome is in accordance with the generally accepted rules for the treatment of basal cell carcinomas and keratocysts in other patients. Radiation should be avoided, as it may trigger off the development of other tumors in the adjacent skin areas. Cystectomy, including the removal of the bony walls of the resulting cavity, is an adequate surgical treatment for the odontogenickeratocysts. In the treatment of the recurrent OKCs which are associated with NBCCS, the overlying surface epithelium should be excised along with the cystic lining to preventrecurrences from the residual epithelial islands and microcysts.[8] In addition, the use of Carnoy's solution following cyst enucleation(applied only over the areas where the cyst is attached to the mucosa) and cryosurgery (because of the unique ability of liquidnitrogen to devitalize the bone in situ while leaving the inorganic framework untouched) is advocated to kill the epithelial remnants and the dental lamina within the osseous structures and to thus, prevent recurrences.[9]

## CONCLUSION

Our case highlights the importance of the awareness of this rare syndrome, especially in young patients without any skin lesions. It is useful to keep in mind the existence of this syndrome and to recognize the presence of some major criteria

that are easily recognizable in the CT scan of the head and neck, to thus establish the diagnosis, to offer the opportunity for frequent follow-ups and to therefore, increase the chances for better overall survival rates.[10] In this case, the patient had also undergone surgeries in the past for cyst enucleation, but the possibility of the Gorlin-Goltz syndrome was not considered at that time and therefore, no frequent follow-ups were advised to the patient. It is important to follow up the patients with diagnosed syndromes for the rest of their lives, because they can produce new odontogenic cysts and new basal cell carcinomas almost continuously. Basal Cell Carcinomas require frequent follow-up care, 3-4 times a year (or more), to achieve an early diagnosis and to plan the treatment. In young children whoare at risk of medulloblastomas, necessitate a neurological examinationevery 6 months, and intermittent MRIs should be considered in children who are younger than 7 years of age. Odontogenickeratocystsrequire dental follow-up visits, including a periodic radiographic evaluation every 6 months, especially in childhood and early adolescence.[11]Finally, the whole family of the patients with the Gorlin-Goltz syndrome should be examined and genetic counseling should be offered, as it is inherited as an autosomal dominant disorder.

## References


1. Gailani MR, Bale SJ, Leffell DJ, *et al*. Developmental defects in Gorlin syndrome related to a putative tumor suppressor gene on chromosome. Cell 1992; 69(1):111-7.
2. Lindeboom JA, Kroon FH, de Vires J, van den Akker HP. Multiple recur¬rent and de novo odontogenickeratocysts associated with oral-facial-digital syndrome. OralSurg Oral Med Oral Pathol Oral RadiolEndod 2003; 95(4):458–62.
3. Evans DG, Sims DG, Donnai R. Family implications of neonatal Gorlin′s syndrome. Arch Dis Child 1991; 66:1162-3.
4. Karthiga S Kannan, Sivapatha B Sundharam, RManikandan. Nevoid basal cell carcinoma syndrome. IJDR 2006; 17(1): 50-53.
5. Dominguez FV, Keszler A. Comparative study of keratocysts, associated and non-associated with nevoid basal cell carcinoma syndrome. *J Oral Pathol* 1988; 17(1):39–42.
6. Woolgar JA, Rippin JW, Browne RM. The odontogenickeratocystand its occurrence in the nevoid basal cell carcinoma syndrome. Oral Surg Oral Med Oral Pathol 1987; 64(6):727–30.
7. Bakaeen G, Rajab LD, Sawair FA, Hamdan MA, Dallal ND. Nevoid basal cell carcinoma syndrome: a review of the literature and a report of a case. *Int J Paediatr Dent* 2004; 14(4):279–87.
8. Myoung H, Hong SP, Hong SD, Lee JI, Lim CY, Choung PH, and others. Odontogenickeratocyst: review of 256 cases for recurrence and clinico¬pathologic parameters. Oral Surg Oral Med Oral PatholOral RadiolEndod 2001; 91(3):328–33.
9. Stoelinga PJ. Excision of the overlying, attached mucosa, in conjunction with cyst enucleation and treatment of the bony defect with carnoy solution. Oral MaxillofacSurgClin N Am 2003; 15:407–14.
10. Christina Kaloaeropoulou, PetrosZampakis, SantraKazantzi, PantelisKraniotisand Nicholas S Mastronikolis. Gorlin-Goltz Syndrome – incidental finding on routine CT scan following car accident. *Cases Journal* 2009; 2:9087.doi:10.1186/1757-1626-2-9087.
11. Berg Daniel. Nevoid Basal Cell Carcinoma Syndrome: Follow up [Internet] 2010.[Updated 2009 March 12; cited 2010 Jan 20] Available from http:// emedicine.medscape.com/article/1101196- follow- up


*******

**How to cite this article:**

Mohammad Shakeel *et al*.2016, Multiple Jaw Cysts In Association With Gorlin Goltz Syndrome-A Case Report. *Int J Recent Sci Res.* 7(6), pp. 11994-11996.

TSSN 0976-3031