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Gorlin-Goltz syndrome – a case report

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Abstract: *Gorlin-Goltz syndrome or basal cell nevus bifid rib syndrome is a genetic disorder characterized by the presence of multiple odontogenic keratocysts. Odontogenic keratocyst (OKC) is a developmental cyst of epithelial origin associated with high rate of recurrence. This case report elaborates a case of gorlin-goltz syndrome in 10 year old boy treated by enucleation with chemical cauterisation and stresses the need of systemic survey in all patients diagnosed with multiple OKC to rule out gorlin-goltz syndrome.*

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I. Introduction

Gorlin-Goltz syndrome is an autosomal dominant inherited disorder[1] characterized by the presence of multiple odontogenic keratocysts along with various cutaneous, dental, osseous, ophthalmic, neurological, and sex organ abnormalities.[2, 3]

Early diagnosis is essential as it may progress to aggressive basal cell carcinomas and neoplasias. Odontogenic keratocyst (OKC, currently designated by the World Health Organization as a keratocystic odontogenic tumor) is a locally aggressive, cystic jaw lesion with a putative high-growth potential and propensity for recurrence.

II. Case Report

A 9 yr old male patient presented with a swelling in relation to right side of his face for past 3 weeks, with a gradual increase in size.

On examination the swelling was diffuse, roughly ovoid, 3cm×2cm in size and extending superoinferiorly from lower margin of orbit to the angle of mouth and anteroposteriorly from ala of nose to zygomatic arch. The skin over the swelling was normal. On palpation, the swelling was non-tender, bony hard in consistency and there was no evidence of associated lymphadenopathy. (Fig 1) The swelling extended intraorally from 12 to 16 regions obliterating the buccal vestibule. (Fig 2)

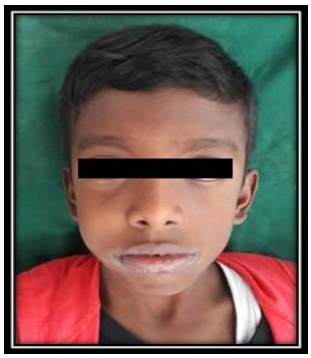

Fig 1: Preoperative clinical picture

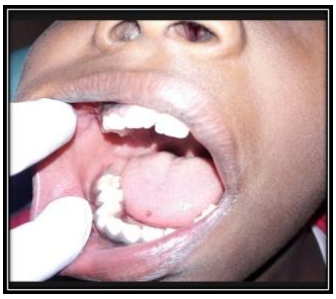

Fig 2: Preoperative intraoral picture

OPG revealed loss of sinus architecture in right maxilla with displacement of 17 to infra orbital margin, pericornal radiolucencies were also seen in relation to 27, 43, 46, and 47 region with displacement of involved and the adjacent teeth. (Fig 3) The radiographic findings were confirmed in the CBCT. (Fig 4 &5)
The radiolucencies where aspirated under LA and they yielded a dirty white fluid with keratin flakes suggestive of OKC.

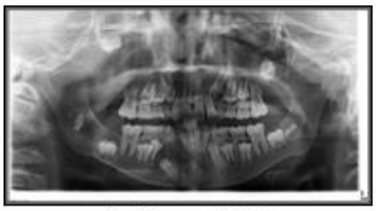

Fig 3: Preoperative OPG

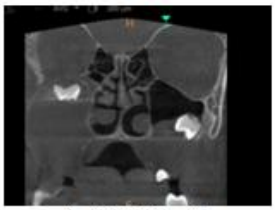

Fig 4: CBCT Maxilla

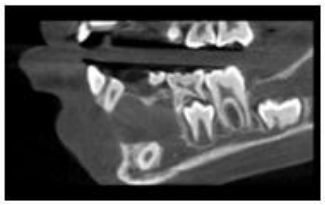

Fig 5: CBCT Mandible

Since the case was reported with multiple cystic lesions, a systemic survey was carried out which revealed mild frontal bossing and hypertelorism, palmar and plantar pitting, bifid 5th rib and bridging of sella turcica, all suggestive of gorlin-goltz syndrome. (fig 6, 7, 8, 9)

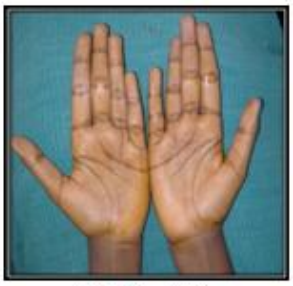

Fig 6: Palmar Pitting

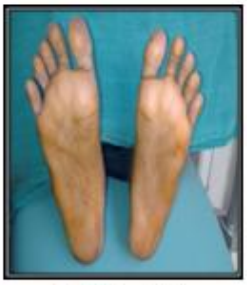

Fig 7: Palmar Pitting

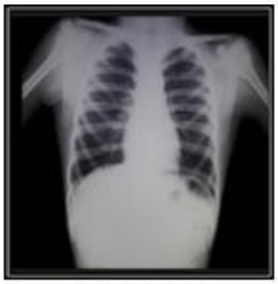

Fig 8: Bifid 5th rib

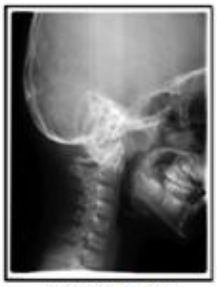

Fig 9: Bridging Sella

The case was posted under general anaesthesia, crevicular incision were placed over the involved teeth with anterior and posterior releasing incision wherever deemed necessary, mucoperiosteal flaps were raised, bony windows were created and the cystic lesions were enucleated in toto along with the impacted and the involved teeth. The Surgical beds were treated with carnoy's solution with due care to adjacent vital structures. (fig10, 11, 12)

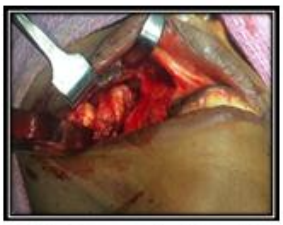

Fig 10: Intraoperative right maxilla

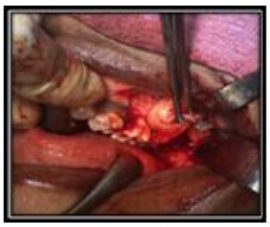

Fig 11: Intraoperative left maxilla

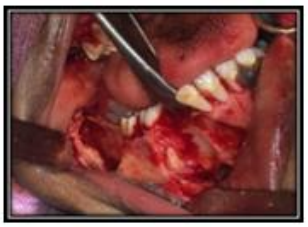

Fig 12: Intraoperative right mandible

The postoperative outcome was better with all surgical sites healing well without any complications. (Fig13, 14)

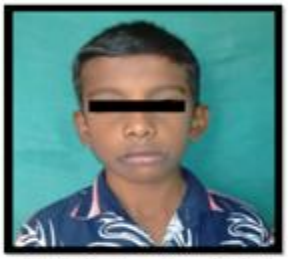

Fig 13: Postoperative clinical picture

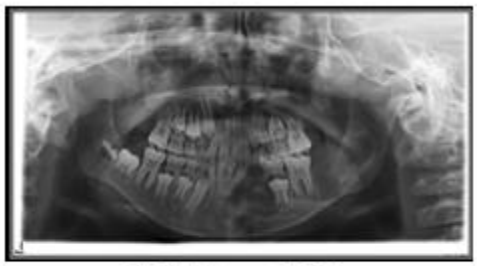

Fig 14: Postoperative OPG

III. Discussion

The incidence of OKC varies from 1 in 50,000 to 1 in 150,000. It has equal gender predilection. Peak incidence of occurrence is 10-40 years and has greater predilections for the mandible than the maxilla. In the present case the age of occurrence is 10 yrs and the lesions were present both in maxilla and mandible.

The presence of multiple OKC of the jaws must alert a clinician as to the possibility of Gorlin-Goltz syndrome. The syndrome was 1st reported by Jarisch and White in 1894, and later in detail by Gorlin and Goltz. The pathogenesis of the syndrome is attributed to abnormalities in the long arm of chromosome 9(q22.3–q31) and loss of / mutations of human patched gene (PTCH1 gene) [4].

Diagnosis of Gorlin-Goltz syndrome can be established when two major or one major and two minor criteria are present.[5, 6]

The major criteria are:

- Multiple basal cell carcinomas or one occurring under the age of 20 years.
- Histologically proven OKCs of the jaws.
- Palmar or plantar pits (three or more).
- Bilamellar calcification of the falx cerebri.
- Bifid, fused or markedly splayed ribs.
- First-degree relative with Nevoid Basal Cell Carcinoma syndrome

The minor criteria are:

- Macrocephaly (adjusted for height).
- Congenital malformation: cleft lip or palate, frontal bossing, coarse face, moderate or severe hypertelorism.
- Other skeletal abnormalities: sprengel deformity, marked pectus deformity, marked syndactily of the digits.
- Radiological abnormalities: bridging of the sella tursica, vertebral anomalies such as hemivertebrae, fusion or elongation of the vertebral bodies, modeling defects of the hands and feet or flame shaped hands or feet.
- Ovarian fibroma.
- Medulloblastoma.

In the described case, 3 major criteria and 2 minor criteria were fulfilled thus establishing it as Gorlin-Goltz syndrome.

OKC is always associated with high rate of recurrence, Brannon, in 1976, suggested incomplete removal of the cyst lining, growth of a new KCOT from satellite cysts (or odontogenic rests left behind after surgery) and the development of a new KCOT in an adjacent area as possible mechanisms of recurrence[7]. It is also based on the great mitotic activity and growth potential found in the cystic epithelium as well as to the remnants of dental lamina and epithelial islands.

Most clinicians favour “conservative” therapy, while others advocate more “aggressive” forms of treatment. Meiselman et al. consider “enucleation, curettage and marsupialization” as conservative therapies. Aggressive treatment addresses the “neoplastic nature” of KCOT and includes peripheral ostectomy, chemical curettage with Carnoy’s solution or en bloc/segmental resection. All patients need frequent follow up and monitoring at regular intervals.

Stoelinga (2005) recommended treating the bone defect around the cyst with carnoy’s solution and excising the overlying, attached mucosa when cortices are perforated[8, 10]. Carnoy’s solution eradicates epithelial rests from the cyst wall but its major drawbacks include caustic effect that may damage adjacent tissues and nerves and possible systemic toxicity.

IV. Conclusion

All cases of multiple OKC should undergo a systemic survey to rule out Gorlin-Goltz syndrome. The surgical bed after enucleation of the cyst should chemically cauterised with carnoy's solution to prevent recurrence and all patients require frequent follow up and monitoring at regular intervals.

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