IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 19, Issue 10 Ser.11 (October. 2020), PP 22-24
www.iosrjournals.org

Airway and anaesthetic management ina rare case of GorlinGoltz Syndrome

Prashant Khadanga[1], Pallavi Gaur[1], Praveen Ubale[2], Sandeep More[1]
[1]Registrar, [2]Associate Professor
Department of Anaesthesia,Topiwala National Medical College and BYL Nair Charitable Hospital,Mumbai,India)
**Corresponding Author: PrashantKhadanga*

Abstract:
Background*:Gorlin-Goltz syndrome (GGS) is a rare autosomal dominant inherited condition characterized by nevoid basal cell carcinomas, odontoid cysts, palmoplantar pits with syndromicfacies. Odontogenickeratocyst makes the airway difficult and associated syndromic features with systemic comorbidities can complicate the overall anaesthetic management. The choice of anaesthetic procedure must be carefully considered given these abnormalities. We report successful anaesthetic management of 15 years old male patient diagnosed with GGS presented for excision of odontogenickeratocyst. The uneventful perioperative course, in this case, was due to thorough systemic evaluation and careful anaesthetic management.*

Key Word*: Gorlin-Goltz syndrome (GGS), Nevoid basal cell Carcinoma Syndrome (NBCCS), Difficult Intubation, Odontogenickeratocyst (OKC)*

Date of Submission: 10-10-2020 Date of Acceptance: 26-10-2020

I. Introduction

Management of syndromic patients with multi-system disorders and craniofacial deformities has always been challenging for the anaesthetist. One among the rare genetic disorders is Gorlin-Goltz syndrome (GGS) caused due to mutations in the patched gene (PTCH1) found on chromosome arm 9q.[1] It shows high penetrance with variable expressivity and characterized by basal cell carcinoma (BCC), odontogenickeratocysts (OKC), palmar and/or plantar pits and musculoskeletal abnormalities.[2,3] The incidence varies from 1 in 57000 to 1 in 256,000 with equal prevalence in males and females.[3] OKC is a major presentation in approximately 75% of younger patients which imposes a greater difficulty in airway management.[4] We present a report of anaesthetic management of one such patient posted for odontogenickeratocystectomy.

II. Case Report

After A 15-year-old male weighing 44kg a diagnosed case of GGS was posted for excision of odontogenickeratocyst under general anaesthesia. The patient had presented with swelling and pain in the lower jaw for one month. On oral examination, it was a large cystic lesion of the mandible. The patient had syndromic features of frontal bossing, hypertelorism and wide nasal bridge, multiple nevi on face with palmar and plantar pits (Figure 1). Intraoral examination revealed a large cystic lesion in left lower jaw. Airway assessment showed inter-incisor gap of 2 cm and Mallampatti class of 2. On further evaluation from computed tomography (CT) scan brain and 3D CT face, there was evidence of multiple odontogenickeratocysts and calcifications involving falxcerebri and tentorium cerebelli suggesting a diagnosis of GGS.

Routine haematological and biochemical investigations were within normal limits. Chest X-ray and 2D-echocardiogram (2D-Echo) were normal. There was no kyphoscoliosis or other vertebral defects. The patient was conscious, calm, and cooperative. There was no history of congenital hydrocephalous, congenital blindness or strabismus, or features suggestive of medulloblastoma. No gonadal abnormalities were noted. With the consideration of anticipated difficult airway and intraoral cystic lesion, we planned for awake nasal intubation followed by general anaesthesia. A well-informed written consent was taken and patient was counselled thoroughly for awakefibreoptic intubation. The Difficult airway cart including fibreoptic bronchoscope (FOB), variable sizes of facemasks, oral airways, nasopharyngeal airways, endotracheal tubes, Proseal laryngeal mask airway and emergency cricothyroidotomy kit were kept ready. In the operating theatre, after attaching full standard monitoring peripheral intravenous access was secured. The patient was premedicated for aspiration prophylaxis with Pantoprazole (0.8mg/kg) and Ondansetron (0.08mg/kg) intravenously (iv). The oropharyngeal and nasal passage was anaesthetized using lignocaine gargles and nasal packing with 2% lignocaine respectively. Xylometazoline (0.05%) was also instilled in both nostrils. Injection

(Inj.) Glycopyrrolate 0.16mg, Inj. Midazolam 1 mg and Inj. Fentanyl 80 µg was administered. FOB was inserted in right nostril and on visualization of vocal cord, the FOB was advanced using the "spray as you go technique" with 4% lignocaine. Fibreoptic intubation was done successfully with 6.0 no. reinforced endotracheal tube (ETT) and tracheal placement was confirmed by end-tidal carbon dioxidetracings on the monitor (Figure 2). General anaesthesia was administered with Inj. propofol 2 mg/kg and Inj. vecuronium 0.1 mg/kg. Throat pack was also inserted. Anaesthesia was maintained with oxygen air mixture (50:50) and sevoflurane with intermittent doses of Inj. vecuronium. Neuromuscular monitoring was done using peripheral nerve stimulator. Hemodynamic and other vital parameters were stable throughout. For pain relief, Inj. Paracetamol 600mg and local infiltration were given. The total duration of surgery was two hours. The patient was reversed with Inj. Glycopyrollate 0.4mg, Inj. Neostigmine 2.5mg and extubated after regular respiration and return of adequate tone, power and reflexes. The patient was transferred in recovery position to the post anaesthesia care unit for further monitoring.

Figure 1: Facial features showing multiple nevi, hypertelorism and frontal bossing

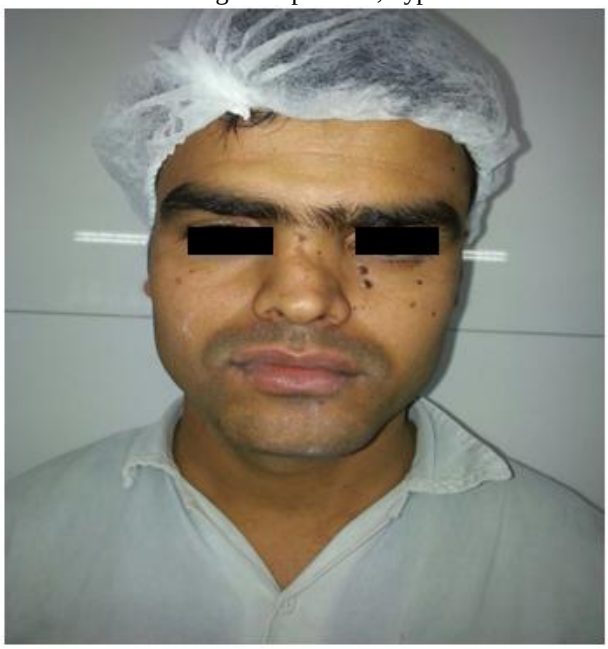
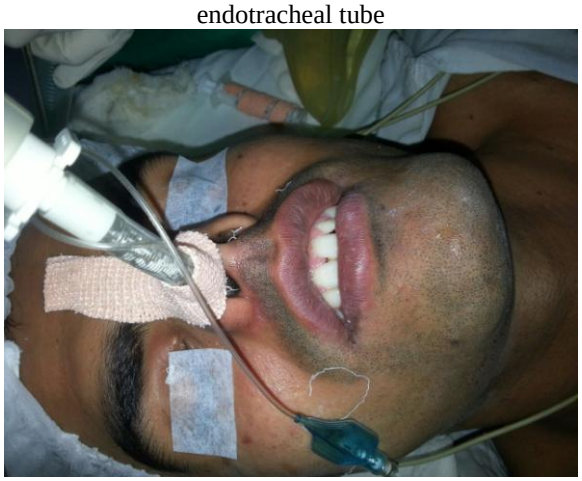


Figure 2: Intraoperative image showing patient intubated through right nostril with No. 6.0 reinforced endotracheal tube



III. Discussion

GGS is a rare autosomal dominant inherited multi-systemic disorder. The main clinical features for diagnosis are BCC, OKC, palmoplantar pits, calcification of the falxcerebri, medulloblastoma and first-degree relative with GGS (table-1).[3] The incidence of this syndrome is variable among different ethnic groups and rarely reported in the Indian population.[1,5]

Table-1: Diagnostic criteria for GorlinGoltz syndrome according to Kimonis *et al*.[6] and consensus statement from the first international colloquium on Gorlin-Goltz syndrome[3] (two major or one major and two minor criteria are present)

Major criteria	Minor Criteria
Multiple basal cell carcinomas or one occurring under the age of 20 years	Macrocephaly (adjusted for height).
Histologically proven OKC of the jaws	Congenital malformation: Cleft lip or palate, frontal bossing, coarse face, moderate or severe hypertelorism
Palmar or plantar pits (three or more)	Other skeletal abnormalities: Sprengel deformity, marked pectus deformity, marked syndactyly of the digits
Bilamellar calcification of the falxcerebri	Radiological abnormalities: Bridging of the sellaturcica, vertebral anomalies such as hemivertebrae, fusion or elongation of the vertebral bodies, modelling defects of the hands and feet or flame-shaped hands or feet
Bifid, fused or markedly splayed ribs	Ovarian fibroma
First-degree relative with Nevoid BCC	Medulloblastoma

In our patient, the diagnosis of the GGS was established by the presence of two major criteria (multiple OKC and calcification of falxcerebri/ tentorium cerebelli) and three minor criteria (frontal bossing, palmoplantar pits and hypertelorism).

Numerous OKC typically occur as a component of Nevoid BCC or GGS, Oro-facial-digital syndrome, Noonan syndrome, Ehler-Danlos syndrome, Simpson-Golabi-Behmel syndrome.[2,7] However, in this case, clinical and radiological features were not in favour of any other syndromes except for GGS. Our case presented with multiple cystic lesions involving the mandible, which was confirmed as keratocysts by radiological and histopathological tests.

Anaesthetic implications in GGS involve multiple system involvement including skeletal and cardiac abnormalities. Our patient had the syndromic facial features such as frontal bossing, wide nasal bridge and hypertelorism along with intraoral OKCs. These features may also impose difficulty in mask ventilation and laryngoscopy and airway obstruction.[1,8,9] Considering the above features and anticipating difficult airway, we decided for awake fibreoptic intubation as the first choice.

Skeletal abnormalities like bifid ribs and kyphoscoliosis if present can compromise pulmonary compliance and increase the risk of perioperative pulmonary complications.[1,3,4] This patient had no such abnormalities. Preoperative 2D-Echo was done to rule out cardiac anomalies, which was normal in our case. The patient was extubated uneventfully after the surgery. The postoperative course was uneventful as well and he was discharged to the ward after adequate monitoring.

IV. Conclusion

Anaesthetic management of GGS is very challenging due to multi-systemic involvement and airway abnormalities. The uneventful course of this case was because of thorough preoperative assessment, detailed investigations, adequate perioperative preparation and management.

References

[1]. Gosavi KS, Mundada SD. Anaesthetic management in Gorlin-Goltz syndrome. *Indian J Anaesth*. 2012;56(4):394-396.
[2]. Mehta D, Raval N, Patadiya H, Tarsariya V. Gorlin-Goltz Syndrome. *Ann Med Health Sci Res*. 2014;4(2):279-282.
[3]. Larsen Anne Kristine, MikkelsenDortheBisgaard, Hertz Jens Michael, BygumAnette. Manifestations of Gorlin-Goltz syndrome. *Dan Med J*. 2014;61(5):1-5.
[4]. Ljubenovic M, Ljubenovic D, Binic I, Jovanovic D, Stanojevic M. *Gorlin-Goltz Syndrome*.2007;16(4):166-169.
[5]. Lata J, Verma N, Kaur A. Gorlin-Goltz syndrome: A case series of 5 patients in North Indian population with comparative analysis of literature. *ContempClin Dent*. 2015;6(Suppl 1):S192-S201.
[6]. Kimonis V, Goldstein A, Pastakia B, Yang M, Kase R. Clinical Manifestations in 105 Persons With Nevoid Basal Cell Carcinoma Syndrome. *Am J Med Genet*. 1997;69(3):299-308.
[7]. Muzio L Lo. Nevoid basal cell carcinoma syndrome (Gorlin syndrome). *Orphanet J Rare Dis*. 2008;3(32):1-16.
[8]. Rhee KY, Baek RM, Ahn KJ. Airway management in a patient with focal dermal hypoplasia [25]. *AnesthAnalg*. 2006;103(5):1342.
[9]. Holzman RS. Airway involvement and anesthetic management in Goltz's syndrome. *J ClinAnesth*. 1991;3(5):422-425.

PrashantKhadanga, et. al. "Airway and anaesthetic management ina rare case of GorlinGoltz Syndrome." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 19(10), 2020, pp. 22-24.