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Original Research Paper

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CONGENITAL RANULA PRESENTING AS SEVERE ACUTE AIRWAY OBSTRUCTION AT BIRTH- A CASE REPORT

Sidharth Nayyar	Neonatology resident KEM Hospital, Pune
Arup Ratan Mondal*	Neonatology resident KEM Hospital, Pune *Corresponding Author
Dhyey Pandya	Neonatology resident KEM Hospital, Pune
Umesh Vaidya	Consultant neonatology, KEM Hospital, Pune

ABSTRACT Congenital ranula is very rare. Case reports published worldwide have been very few. They are formed either as retention cyst or as pseudocyst due to extravasation of mucus in the surrounding tissue. We report the case of a full term female neonate with a congenital ranula in the floor of mouth. A well-planned immediate postpartum treatment procedure for aspiration and decompression of the cystic lesion is simple, efficacious and effective in securing a patent airway, and may prevent lifelong neurodevelopmental complications associated with perinatal asphyxia in such cases.

KEYWORDS :

INTRODUCTION

The incidence of congenital ranula is estimated to be about 0.7% in newborn infants [1]. It is a retention cyst situated in the floor of the mouth caused by atresia or failure of canalization of the salivary ducts of the sublingual or submandibular gland. At the initial stage of ranula formation, the excretory duct ruptures, and leads to extravasation and the accumulation of mucinous secretion in the surrounding tissues. The high protein content of the secretion triggers an intense inflammatory reaction and results in the formation of a pseudocyst. Very rarely such a cystic lesion may grow so large that it completely occupies the entire oral cavity and obstructs the upper airway at birth [2,3].

Ranula is classified as being simple or plunging. A simple ranula involves the sublingual space only whereas a plunging ranula extends posterior to the mylohyoid muscle into the neck. Simple ranulas are mostly asymptomatic but can lead to airway obstruction We recently encountered such a scenario of a congenital giant ranula diagnosed antenatally by ultrasound.

CASE REPORT

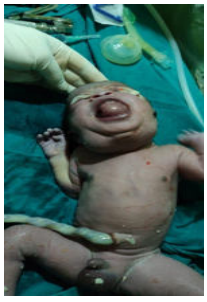
A male full term neonate with birth weight 2.5 kg was born to a apparently healthy 25 yrs old G2P0A1 mother at the obstetrics operation theatre of K.E.M Hospital, Pune by lower segment caesarean section. There was a history of spontaneous abortion at 3 months of pregnancy one year ago. The pregnancy was relatively uncomplicated and there were no other specific antenatal risk factors. Family history was not significant.

Antenatal USG showed a giant congenital ranula arising from the floor of the mouth measuring approximately 3 cm x 4 cm along with polyhydramnios.

In the anticipation of severe airway obstruction at birth due to the large ranula, the caesarean section was planned and attended by a team consisting of a Neonatologist, ENT surgeon, Paediatric surgeon and Anaesthesiologist. A proper resuscitation kit was prepared as the baby may require immediate aspiration of the contents of the ranula or if the structure is vascular then the baby may require immediate tracheostomy at birth to facilitate and maintain a patent airway.

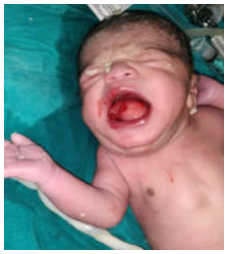
Picture 1. The resuscitation kit

Baby was delivered with a large cystic lesion filling the entire oral cavity pushing the tongue backwards making it difficult for the baby to maintain the patency of the airway.



Picture 2- A Large Cystic Ranula obstructing the entire oral cavity causing acute upper airway obstruction can bee seen.

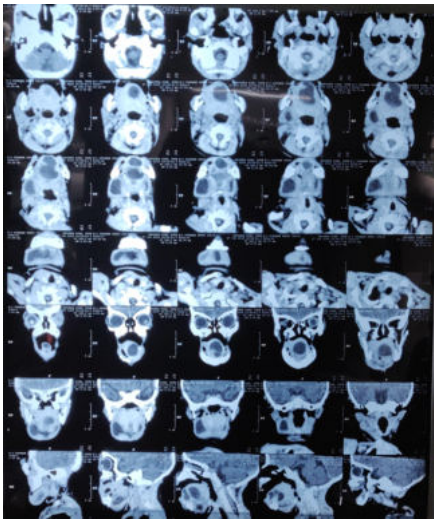
Baby was shifted immediately underneath a preheated warmer after the delivery. The baby showed good spontaneous respiratory efforts and cry activity was fair. A local examination was performed which was suggestive of a large bluish, translucent swelling measuring approximately 4 cm x 4 cm ,non pedunculated, round smooth surfaced, non pulsatile, avascular, appearing to be arising from the floor of the mouth and uplifting the tongue and pushing it backwards causing difficulty in breathing seconds after the baby was born. Under supervision of paediatric and ENT surgeon , the cyst was aspirated with 18 G needle under all aseptic precautions in the OT immediately. A total of 15 cc clear aspirate followed by another 8 cc haemorrhagic aspirate was obtained and the cyst size reduced to approximately 1.5 cm x 1.5 cm. Baby started breathing comfortably after the intervention and was shifted to NICU for stabilization and further course of action.



Picture 3- Post aspitration in the OT, size ot the ranula has decreased

Baby maintained saturation on room air with no respiratory distress. He was kept NBM and with IV fluid for a period of 6 hrs and after that he was started on Full Orogastric feeds. On day 3rd of life he was

started on Oral feed which he was taking adequately without any respiratory distress. The size of the ranula remained constant and there was no need of further aspiration of the cyst. CT Scan of the cyst was done on day 4th and showed a well defined cystic lesion in sublingual space in region of the floor of mouth extending inferiorly B/L into submandibular space suggestive of a Plunging Ranula.



Picture 4- CT Scan showing B/L Plunging ranula

Baby was discharged from NICU at 10th days of his life after securing proper oral feeding and adequate weight gaining. Definitive corrective surgery was planned after 1 month in consultation with a pediatric surgeon as an early surgery would lead onto superimposed infections of the ranula and the surrounding neck tissues inturn leading onto the compromise of the airway due to edema and abscess formation subsequently

DISCISSION AND CONCLUSION

A Giant Congenital ranula is a very rare presentation at birth (incidence 0.7%) [1]. A giant ranula like this may cause a severe life threatening obstruction of the upper airway at the level of the oral cavity and needs immediate decompression or drainage to prevent the adverse consequences of asphyxiating birth injury to the brain and other vital organs.

An EXIT Procedure (*Ex Utero* Intrapartum treatment) has been described by Chan *et al* in 2006 wherein they aspirated and decompressed the cystic lesion of the oral cavity in securing a patent airway before the fetus is completely delivered [2].

Onderglue et al. reported a case of congenital ranula in a developing fetus displacing tongue antero superiorly. Anticipating airway obstruction, an ex utero intra partum treatment (EXIT) procedure was done at 38 weeks gestation and management by simple aspiration of the cyst fluid prior to ligation of umbilical cord was done, with no recurrence at 6 months follow up.

There are however potential adverse effects and limitations associated with the EXIT procedure. This technique is not widely available in the majority of OT's and specialized tertiary centres with a specialized team is required for the procedure to complete within a short period of time. Secondly careful selection of the appropriate medical conditions and timely transfer of patients are important for successful management of this infant. Thirdly vasoactive tocolytic drugs and high doses of inhaled halogenated agents are required for deep general anaesthesia and uterine relaxation, which is essential for maintaining the feto-placental circulation for which mother are at an increased risk of developing uterine haemorrhage and hemodynamic complications. Thus a very close monitoring of foetal and maternal cardio-respiratory parameters, blood loss are mandatory during the procedure. In the present case report, a full term male baby presenting with a giant ranula completely obstructing the upper airway, diagnosed as a plunging ranula by CT scan later on, was treated with aspiration of the cyst in the OT immediately after the baby was born. All the resuscitation equipment needs to be made available in a resource limited setup to prevent mortality or long term morbidity in such cases as a consequence of birth asphyxia. Precious time would be lost in trying to establish a patent airway, and the infant might suffer from hypoxic-ischaemic brain insult, resulting in lifelong neurodevelopmental deficit. In contrast, the procedure of aspirationduring the immediate postpartum period in the Operation theatre or the delivery room performed by skilful hands is a highly structured and coordinated operation which offers genuine hope to mothers and emergency rescue to fetuses with life-threatening upper airway obstruction. In particular, the intervention of congenital giant ranula or other oral cystic lesions is simple and efficacious.

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