CASE REPORT

Exit : A Salvage Procedure for Intraoral Teratoma

Masaany Binti Mansor, MSO HNS (ORL) UKM, Ida Sadja'ah Sachlin, (MSO HNS, USM), Norleza Ahmad Norhan, (MSO HNS, UKM), Norzi Binti Gazali, (MSO HNS, USM), Siti Sabzah Mohd Hashim, (MSO HNS, UKM)

ORL Specialist, Hospital Sultanah Bahiyah, Otorhinolaryngology, KM 6, Jalan Langgar, Alor Setar, Kedah 05460 Malaysia

SUMMARY

Teratomas arising from the palate or pharynx may cause immediate life-threatening airway obstruction to the newborn. Early diagnosis via antenatal ultrasound enables the treating team to perform an ex utero intrapartum treatment (EXIT) procedure to secure an airway and perform subsequent tumour resection. We present the first EXIT procedure done in Malaysia. A 34 year old , gravida 4, para 3, her unborn child diagnosed at 24 weeks of gestation to have a large oropharyngeal mass. Upper airway obstruction was anticipated. Orchestration of multiple specialities was executed to properly plan and perform the EXIT procedure. The fetus was delivered at 33 weeks of gestation and managed to be intubated. The extraoral portion of the multilobular mass originating from the palate was resected. Complete resection of the intraoral teratoma was successfully done at day 22 of life.

INTRODUCTION

Ex-utero intrapartum treatment (EXIT) is a technique designed to allow partial fetal delivery via caesarean section while a safe fetal airway is established. Upon delivery of the fetal head, neck and one or both upper limbs, a stepwise, sequential attempt to sequre the neonates airway is performed via laryngoscopy, bronchoscopy, tracheostomy or even resection of the obstructing mass. The placenta remains in utero to maintain feto-placental circulation and subsequently the continuance of fetal oxygenation. Once the newborn's airway is secured, the umbilical cord is clamped and delivery of the infant completed. First described in 1990 by Zerella and Finberg [1] to improve the survival of fetuses with head and neck teratomas and also by Levine et al for epignathus [1], the use of EXIT has proven to be effective for the management of fetal airway obstruction due to a variety of other causes including congenital high airway obstruction syndrome (CHAOS), intrathoracic masses and congenital diaphragmatic hernias.

CASE REPORT

A 34 year old , gravida 4, para 3 was referred to our centre at 34 weeks of gestation. The reason for referral was that the prenatal ultrasound revealed the patient had extensive polyhydramnios and the unborn fetus had a multicystic perioral tumour. The patient was also having premature contractions 1 in every 5 minutes. Neonatal upper airway obstruction was anticipated .The case was referred to the ear nose and throat surgery (ENT) and paediatrics teams. An EXIT procedure was planned. Emergency lower segment caeserian section was performed on that day itself. Upon delivery of the baby's head, a mixed solid cystic multilobular intraoral mass approximately 7cm by 8 cm was seen protruding extraorally .The neonate could only be intubated by the ENT team using the Parson direct laryngoscope. An endotracheal tube of size 2.5 was inserted. The newborn then moved to another theatre. Further examination revealed the mass to have a broad base with the stalk attached to the palate. It filled up the oral cavity and further extended down until the base of tongue and supraglottic region. The rest of the laryngeal structures were normal. The main extraoral mass was removed. Using a size 2.5 bronchoscope, examination of the subglottis, trachea , left and right main bronchus revealed no abnormality. A nasogastric tube of size 6 was inserted with the aid of a flexible nasopharyngolaryngoscope. Both nasal cavities were normal, the choana both patent but the nasopharynx was filled up by the mass. Histologically, the excised extraoral portion of the mass was found to be immature teratoma grade 1.

Postoperatively, the newborn was ventilated in the neonatal intensive care unit. However, he developed bilateral tension pneumothorax and was treated. On day 4 of life, the neonate was diagnosed to have hydrocephalus secondary to prematuraty. He was however active, afebrile and stable.

A second operation to remove the remaining intraoral part of the teratoma was successfully done on day 19 of life. Postoperatively he was well until day 26 of life when he started vomiting. The hydrocephalus had worsened causing macrocephaly. The baby was immediately referred to the neurosurgical team in another tertiary hospital. Unfortunately, a week later the child succumbed to pneumonia and septicaemic shock despite all measures taken.

DISCUSSION

The term teratoma, introduced by Virchow in 1863, is derived from the Greek word "teraton" which means monster. Teratomas can be generally classified into 4 types. The commonest type is the dermoids, also known as hair polyps, consists of epidermal and mesodermal elements. The teratoid type consists of poorly differentiated elements of ectoderm, mesoderm and endoderm. True teratomas contain all 3 germ cell layers similar to the teratoid but differentiates into more recognizable tissues or organs. Histologically, an epignathus

This article was accepted: 24 April 2012
Corresponding Author: Masaany Binti Mansor, ORL Specialist, Hospital Sultanah Bahiyah, Otorhinolaryngology, KM 6, Jalan Langgar, Alor Setar, Kedah 05460 Malaysia Email: masaanymansor@yahoo.com

is defined as a highly differentiated teratoma that has organized into recognizable organs or limbs [2]. However, etymologically, the term "epignathus tumour" applies to teratomas of the oropharyngeal cavity in neonates without any specific site of origin [2]. The incidence of oropharyngeal teratomas varies between one in 35,000 to one in 200,000 live births [2]. Head and neck teratomas only account for less than 5% of all teratomas, with cervical teratomas being more common than oropharyngeal teratomas [2]. In our case, the oropharyngeal teratoma arose from the palate. Histopathological report showed the tissues to be mature derivatives from ectoderm, mesoderm and endoderm but the presence of neuroectodermal elements made it an immature teratoma Grade 1. 90% of childhood teratomas contain derivatives from all 3 embryonic germ cell layers. 75% to 85% of the head and neck teratomas usually show both mature and immature characteristics [2].

Advances in prenatal diagnosis enabling anticipation of neonatal upper airway obstruction and orchestration of delivery via the EXIT has helped improve the prognosis and survival of many potentially life threatening fetal anomalies. During the EXIT, intensive maternal-fetal monitoring, hemostatic hysterectomy with maximal uterine relaxation to maintain a good feto-placental circulation is needed to provide a controlled environment and sufficient time to secure the neonatal airway. Hirose S et al reported an average operating time on placental support to be 45+/- 25 minutes. Their longest duration was 2.5 hours [1]. The EXIT however carries its own risks. Before the procedure is done, localization of the placenta based on ultrasound is needed in order to avoid placental involvement in the hysterotomy incision site. Polyhydramnios is common in parturients with fetal anomalies. The excessive liquor can compress the edge of the placenta and subsequently obscure ultrasound assessment. In general, there is no difference in short-term maternal outcome between the conventional Caeserean section and the EXIT procedure. Our maternal intraoperative blood loss was only 300 ml. The mother had an uneventful postoperative period and was discharged well on day 2 of the EXIT. A retrospective review of 52 patients who had undergone the EXIT showed average maternal blood loss to be 970+/- 510 ml [3]. In order to avoid heamorrhage from the hysterotomy site, uterine stapling device together with good coordination between surgeon and anaesthesiologist to reduce the concentration of inhalational anaesthetics simultaneous with administration of oxytocin before umbilical cord ligation is necessary. However, Butwick A et al reported massive obstetric haemorrhage during an EXIT despite all the above steps executed.

CONCLUSION

The EXIT is an excellent strategy to establish an airway in a controlled manner, avoiding traumatic intubation or tracheostomy in a newborn child yet causing no additional maternal morbidity. The success of the EXIT however, depends on meticulous preoperative planning, discussion , and close collaboration among the multidisciplinary teams involved.

REFERENCES

1. Hirose S, Farmer DL, Lee H, Nobuhara KK, Harrison MR. The ex utero intrapartum treatment procedure: Looking back at the EXIT. J Pediatric Surg 2004 Mar; 39(3): 375-80.
2. Ajaz S, Suhail L, Irshad A, Altaf H.M. Head and Neck Teraromas. J Maxillofac Oral Surg 2009; 8(1): 60-3.
3. Butwick A, Aleshi P, Yamout I. Obstetric Haemorrhage During An Exit Procedure For Severe Fetal Airway Obstruction. Can J Anaesth. 2009 June; 56(6): 437-42.