Anesthetic management for Ex-utero Intrapartum Treatment (EXIT) of fetus with Epignathus tumor.

Lucia Caroline Schons Correa[1], Mayra Lima Brunelli[2], Carlos Darcy Bersot A[3], Rafael Mercante Linhares[4], Jose Eduardo Guimaraes Pereira[5]*, Marilia SF Macedo[1]

[1]Department of Anesthesiology, Hospital Universitario Pedro Ernesto, HUPE/UERJ, Brazil
[2]Deparment of Anesthesiology, Hospital Naval Marcilio Dias, Rio de Janeiro, Brazil
[3]Department of Anesthesiology, Hospital Federal Lagoa, Rio de Janeiro, Brazil
[4]Department of Anesthesiology, Hospital Municipal Miguel Couto. Rio de Janeiro, Brazil
[5]Department of Anesthesiology, Hospital Unimed Volta Redonda, Rio de Janeiro, Brazil

Abstract

Epignathus is a rare benign teratoma tumor that affects the oropharynx and can obstruct the newborn's airways, leading to death immediately after birth. Despite having a bad prognosis, with the advancement of medicine and early diagnosis with ultrasound performed in prenatal care, it may be treated by a multidisciplinary team in the delivery room, including anesthesiologists, pediatricians, obstetricians, radiologists, pediatric surgeons, among others with the EXIT technique (Ex. intrapartum utero treatment) to secure the fetal airway before complete separation of the fetus from the maternal circulation and avoid risk of maternal complications that may occur during prolongation of the intrapartum period. In this case, we present a successful case of a newborn epignathus with a large mass with total airway obstruction.

Keywords: Epignathus tumor, Ex-utero intrapartum treatment, Pediatric surgeons.

Introduction

Epignathus is a very rare congenital teratoma of the oropharynx composed of cells from ectodermal, mesodermal and endodermal layers, corresponding to approximately 2% of fetal tumors [1]. It originates from the skull base, usually in the hard palate or mandible, specifically in the Rathke pouch region. Diagnosis is performed by means of ultrasonography during pregnancy and its management should be carried out by a multidisciplinary team [2].

Epignathus that arise from the palate or pharynx and protrude from the mouth results in life-threatening airway obstruction and usually causes asphyxia soon after birth. The prognosis depends on the size and location of the lesion, its growth rate, the involvement of intracranial structures and adequate tumor resection [3]. Approximately 6% of patients have associated congenital malformations. It is a rare condition and the largest case series reported to date is from. Who described 3 cases, and in all of them there was an association with other malformations such as Pierre-Robin syndrome and meningoencephalocele. Once the diagnosis is made, the fetus must be stabilized for surgical repair, which is mainly securing the airway [4]. Studies show that better results are obtained when the fetus is stabilized with the umbilical cord intact to provide oxygen to the fetus in case of airway obstruction, which consists of the EXIT technique, which the most is, recommended perinatal treatment for babies with airway obstruction. This treatment is important in cases where tracheostomy is necessary to allow air to enter the lungs [5]. In this report, we present a case of anesthetic management for the EXIT technique for a 34-week pregnant woman with a fetus diagnosed with Epignathus.

Case Report

Patient ASNO, 27 years old, with no previous comorbidities, ASA 2, first pregnancy, 34 weeks, presenting a fetus with an intrauterine diagnosis of congenital oropharyngeal teratoma, diagnosed by ultrasonography in the 20th week of pregnancy, with surgical treatment being proposed in childbirth with a fetus still connected the placenta (EXIT technique: ex utero intrapartum treatment) [6]. The patient was admitted to the operating room and monitored with cardioscopy, pulse oximetry, non-invasive blood pressure monitoring and analysis of the Bi Spectral Index (BSI). Peripheral venous catheter was inserted and general anesthesia was performed in rapid sequence with remifentanil, lidocaine, propofol and rocuronium. Orotracheal intubation was performed by direct laryngoscopy and placement of a 6.5 mm tube and maintenance of hypnosis with 3% Sevoflurane [7]. Then, the patient was placed in the left lateral decubitus position and an epidural was performed at L3-L4 using a 16 G Tuohy needle and epidural catheter was threaded. Catheterization of the left radial artery was performed for monitoring blood pressure.

***Correspondence to:** Jose Eduardo Guimaraes Pereira, Department of Anaesthesiology, Hospital Unimed Volta Redonda, Volta Redonda RJ, Brazil, E-mail:dudunest@gmail.com

***Received:** 06-Jul-2022, Manuscript No. AAAA-22-68663; **Editor assigned:** 08-Jul-2022, PreQC No. AAAA-22-68663 (PQ); **Reviewed:** 22-Jul-2022, QC No. AAAA -22-68663; **Revised**: 12-Aug-2022, Manuscript No. AAAA -22-68663(R); **Published**: 20-Aug-2022, DOI:10.35841/aaaa-4.4.116*

***Citation:** Schons Correa LC, Brunelli ML, Bersot CDA, et al. Anesthetic management for Ex-utero Intrapartum Treatment (EXIT) of fetus with Epignathus tumor. J Anesthetic Anesthesiol. 2022;4(4):116*

During surgery, nitroglycerin infusion in a continuous infusion pump (BIC) was maintained for uterine relaxation and maintenance of uterine volume with warmed lactated Ringer's infusion at 42 ml/h rate [8].

After the cesarean section incision, and after exposing the fetus still connected to the placenta by the umbilical cord, an attempt of orotracheal intubation was performed, but with no success. Therefore, a tracheostomy was performed at birth with maternal-fetal blood flow still intact through the umbilical cord. After securing the fetus' airway, the umbilical cord was clamped and cut, with the birth of a female fetus, which was immediately assisted by the neonatal pediatrics team [9].

After birth, the infusion of nitroglycerin was interrupted, the administration of sevoflurane was interrupted and the infusion of 1 g tranexamic acid, 15 IU of oxytocin in titrated form and controlled target propofol was carried out, in addition to analgesia by administering 1 mg of morphine and 0.2 ropivacaine % 20 ml through the epidural catheter [10]. At the end of the procedure, the patient was extubated and sent to the intensive care unit, presenting a nine point score at the Aldrete-Kroulik scale.

Discussion

Congenital oropharyngeal tumors derived from embryonic germ cells, teratomas, are extremely rare. Epignathus teratomas are intraoral malformations of unknown etiology that can cause respiratory tract obstruction immediately after birth [11].

Epignathus occurs in 1:35000 to 1:200000 live births, corresponding to 2 to 9% of all teratomas. These tumors may be associated with concomitant fetal malformations, with cleft palate being the most common. Other associated malformations are the presence of a bifid tongue and/or nose. Polyhydramnios is usually associated with teratoma epignathus, since the fetus has a delayed swallowing reflex and may be associated with a large neck mass [12,13].

Prenatal diagnosis is essential for perioperative care and management. Recent advances in diagnostic imaging, especially ultrasound, have enabled the early detection of tumors that obstruct the airways of fetuses, allowing planning strategies for securing their airways before birth

12. In the present case, the patient was diagnosed in the 20th week of pregnancy, by means of an ultrasound examination and had polyhydramnios throughout her pregnancy, undergoing weekly amniotic drainage [14].

The EXIT procedure is the ideal delivery strategy for fetuses with oropharyngeal tumors diagnosed in the prenatal period and possible airway obstruction at birth. Fetuses with large cervical or oral tumors obstructing the airways, who are born by conventional cesarean section present higher mortality rate in the immediate postpartum period due to difficulties in ventilation, intubation and tracheostomy. The EXIT procedure consists of maintaining the fetoplacental circulation until the fetus' airway is secure [15]. In this procedure, the anesthesia technique must ensure maternal and fetal anesthesia, with adequate uterine relaxation.

General anesthesia with halogenated anesthetics has been shown to be ideal for fetal surgery in the EXIT procedure. An alternative would be neuraxial anesthesia with intravenous injection of nitroglycerin for uterine relaxation; however such a method may cause clinical significant hypotension. With general anesthesia, the passage of anesthetics through the placenta is guaranteed, which promotes anesthesia and fetal immobility [16]. In addition, the administration of halogenated anesthetics promotes the uterine relaxation required by the EXIT technique. Studies show that administration of isoflurane or sevoflurane up to 2 MAC promotes adequate uterine relaxation, without causing significant maternal hypotension. Concentrations above 2 MAC may result in hypotension, reduced placental blood flow, hypoxia and fetal acidosis [17]. For the anesthesia of the patient in this case, we opted for the association of balanced general anesthesia with the administration of sevoflurane and neuroanalgesia through the epidural for the postoperative period. To ensure adequate uterine relaxation, continuous infusion of nitroglycerin was also used.

The use of tocolytic agents such as nitroglycerin and magnesium sulfate is beneficial in association with halogenated anesthetics to ensure uterine relaxation. Nitroglycerin is the most used agent and has the best pharmacodynamic profile, as its rapid onset of action and short half-life allow the return of uterine tone as soon as its infusion is interrupted. As unwanted effects, nitroglycerin can cause hypotension, which must be promptly corrected with ephedrine or phenylephrine to maintain uterine perfusion. Magnesium sulfate may also be an option, but may prolong the effects of neuromuscular blockers and cause respiratory and cardiovascular depression.

During the entire procedure, a professional experienced in fiberoptic bronchoscopy was present to carry out the intubation by fibroscopy, if necessary. The management of the airway in this case is a challenge and the primary approach by fibroscopy must be considered, and the team must be prepared to perform a tracheostomy as well, as it was chosen in this case.

After clamping the umbilical cord it is necessary that the once high concentrations of halogenated anesthetics to be reduced, thus avoiding uterine atony. The reduction of halogenated anesthetics and the intravenous infusion of oxytocin are capable of reversing uterine relaxation. Thus maternal anesthesia must be complemented with the administration of other anesthetic agents, such as the propofol, which was the choice in this case, in addition to the analgesia promoted by the administration of local anesthetic through the epidural catheter [18].

Conclusion

Fetal surgery is a rapidly evolving area and requires the participation of multidisciplinary teams, in which the anesthesiologist plays an important role. The anesthetic procedure is directed to the maternal-fetal binomial and must be well planned to warrant the safety of both. In this context, the anesthesiologist must not only promote anesthesia, unconsciousness and analgesia of the maternal-fetus binomial, but also the maintenance of uterine relaxation,

without compromising maternal-fetal hemodynamics. Dialogue, integration between the teams and the continuous development, learning and updating of the professionals involved are paramount.

References

1. Chaves YS, Sousa JS, Feldner Jr PC, et al. Teratoma congenito de orofaringe: relato de caso. Revista Brasileira de Ginecologia e Obstetricia. 2005;27:86-91.
2. Chattopadhyay A, Patra R. Oral tumors in newborn. Indian J Pediatr. 2003;70(7):587-8.
3. Gull I, Wolman I, Har-Toov J, et al. Antenatal sonographic diagnosis of epignathus at 15 weeks of pregnancy. Ultrasound in Obstetrics and Gynecology: The Official Ultrasound Obstet Gynecol. 1999;13(4):271-3.
4. Oliveira-Filho AG, Carvalho MH, Bustorff-Silva JM, et al. Epignathus: report of a case with successful outcome. J Pediatr Surg. 1998;33(3):520-1.
5. Goraib JA, Cabral JA, Nogueira AR, et al. Giant epignathus in the newborn: report of a case and review of the literature. Jornal de pediatria. 1995;71(1):41-4.
6. Vandenhaute B, Leteurtre E, Lecomte-Houcke M, et al. Epignathus teratoma: report of three cases with a review of the literature. Cleft Palate Craniofac J. 2000;37(1):83-91.
7. Oliveira-Filho AG, Carvalho MH, Bustorff-Silva JM, et al. Epignathus: report of a case with successful outcome. J Pediatr Surg. 1998;33(3):520-1.
8. Cohen J. Syndromes with cleft lip and cleft palate. Cleft Palate J. 1978;15(4):306-28.
9. Liang CC, Lai JP, Lui CC. Cleft palate with congenital midline teratoma. Ann Plast Surg. 2003;50(5):550.
10. Celik M, Akkaya H, Arda IS, et al. Congenital teratoma of the tongue: a case report and review of the literature. J Pediatr Surg. 2006;41(11):e25-8.
11. Izadi K, Smith M, Askari M, et al. A patient with an epignathus: management of a large oropharyngeal teratoma in a newborn. J Craniofac Surg. 2003;14(4):468-72.
12. Laje P, Howell LJ, Johnson MP, et al. Perinatal management of congenital oropharyngeal tumors: the ex utero intrapartum treatment (EXIT) approach. J Pediatr Surg. 2013;48(10):2005-10.
13. Tanaka M, Sato S, Naito H, et al. Anaesthetic management of a neonate with prenatally diagnosed cervical tumour and upper airway obstruction. Can J Anaesth. 1994;41(3):236-40.
14. Myers LB, Cohen D, Galinkin J et al. Anaesthesia for fetal surgery. Paediatr Anaesth, 2002;12:569-578.
15. George RB, Melnick AH, Rose EC, et al. Case series: Combined spinal epidural anesthesia for Cesarean delivery andex utero intrapartum treatment procedure. Can J Anaesth. 2007;54(3):218-22.
16. Palahniuk RJ, Shnider SM. Maternal and fetal cardiovascular and acid–base changes during halothane and isoflurane anesthesia in the pregnant ewe. The Journal of the American Society of Anesthesiologists. 1974;41(5):462-71.
17. Gaiser RR, Cheek TG, Kurth CD. Anesthetic management of cesarean delivery complicated by ex utero intrapartum treatment of the fetus. Anesth. Analg. 1997;84(5):1150-3.
18. Gaiser RR, Kurth CD, Cohen D, et al. The cesarean delivery of a twin gestation under 2 minimum alveolar anesthetic concentration isoflurane: one normal and one with a large neck mass. Anesth. Analg. 1999;88(3):584-6.