Coc Cer Derg /Turkish J Ped Surg 2022;36(2): 50-54
doi: 10.29228/JTAPS.56818

Case Report

Çocuk Cerrahisi Dergisi
Turkish Journal of Pediatric Surgery
Volum 36
Number 2
2022

Approach To Prenatal Diagnosed Giant Cervical Mass With Extrauterine İntrapartum Treatment (EXIT) Method

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Received / Geliş: 08.01.2022
Accepted / Kabul: 17.03.2022
Published Date: 01.08.2022

ABSTRACT

A patient with prenatal diagnosis of a cervical mass compressing the trachea managed by the the ex-utero intrapartum treatment (EXIT) at birth was detailed. The patient had presented with polyhydramnios and a cervical mass during the prenatal period. At the thirty-seventh gestational week, decision was given to perform the delivery with the EXIT procedure. The baby was delivered by cesarean section under general anesthesia, with the entire team ready to be sterile intraoperatively. The baby was intubated oropharyngeally without clamping the cord to preserve uteroplacental and placento-fetal circulation. The cord was then clamped and the patient taken to the neonatal intensive care unit under stable condition. On the 2nd postnatal day, cervical CT showed a 65x65x55 mm solid mass lesion, which was totally excised on the 3rd postnatal day. Histopathological examination revealed an immature teratoma. The patient is well and without any clinical problem in the 3th postoperative year of follow-up. The EXIT procedure is an effective treatment option in cervical masses that cause severe airway obstruction during the prenatal period, as it preserves placentofetal circulation until safe airway is achieved. Multidisciplinary approach and planning is required for safe performance of this advanced perinatal resuscitation technique.

Key Words: fetus, neck mass, airways obstruction, Ex-Utero Intrapartum Treatment

Cite as: Rahşan Özcan, Ayşe Karagöz, Ali Ekber Hakalmaz, İbrahim Adaletli, Rıza Madazlı, Fatiş Altındaş, Gonca Topuzlu Tekant. Approach to prenatal diagnosed giant cervical mass with extrauterine intrapartum treatment (EXIT) method. Coc Cer Derg /Turkish J Ped Surg 2022;36(2): 50-54. doi: 10.29228/JTAPS.56818

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Introduction

In congenital mass lesions of the neck, face and mediastinum, it is vital to have an open airway and oxygenation immediately after birth. In these cases, in whom teratomas and lymphangiomas constitute the majority of the lesions, recognition during the antenatal period, revealing the relationship of the lesion with airway structures, making preparations for extrauterine intrapartum treatment (ex utero intrapartum treatment-EXIT) and performing a planned cesarean delivery reduces mortality and morbidity [1]. The aim of this report is to present the treatment process of a prenatally diagnosed cervical teratoma with the EXIT procedure and surgical excision to emphasize the importance of multidisciplinary approach in these cases.

Case Presentation

Twenty-nine years old, ASA I, nullipar, 35 weeks pregnant female presented to the perinatology outpatient clinic due to a cervical mass and polyhydramnios detected in the fetus at 33 weeks of gestation. In ultrasonography and fetal magnetic resonance imaging (MRI) examinations, there was a mass in the right half of the neck, containing solid-cystic areas, 64x55 mm in size, causing complete obstruction in the trachea and polyhydramnios (Figure 1). The case was discussed in the council consisting of perinatology, anesthesiology, neonatal and pediatric surgery specialists and decision was given to perform the EXIT procedure during delivery at the 37th gestational week in concordance with the indications mentioned in the literature, as the mass was larger then 5 cm, solid in character, causing

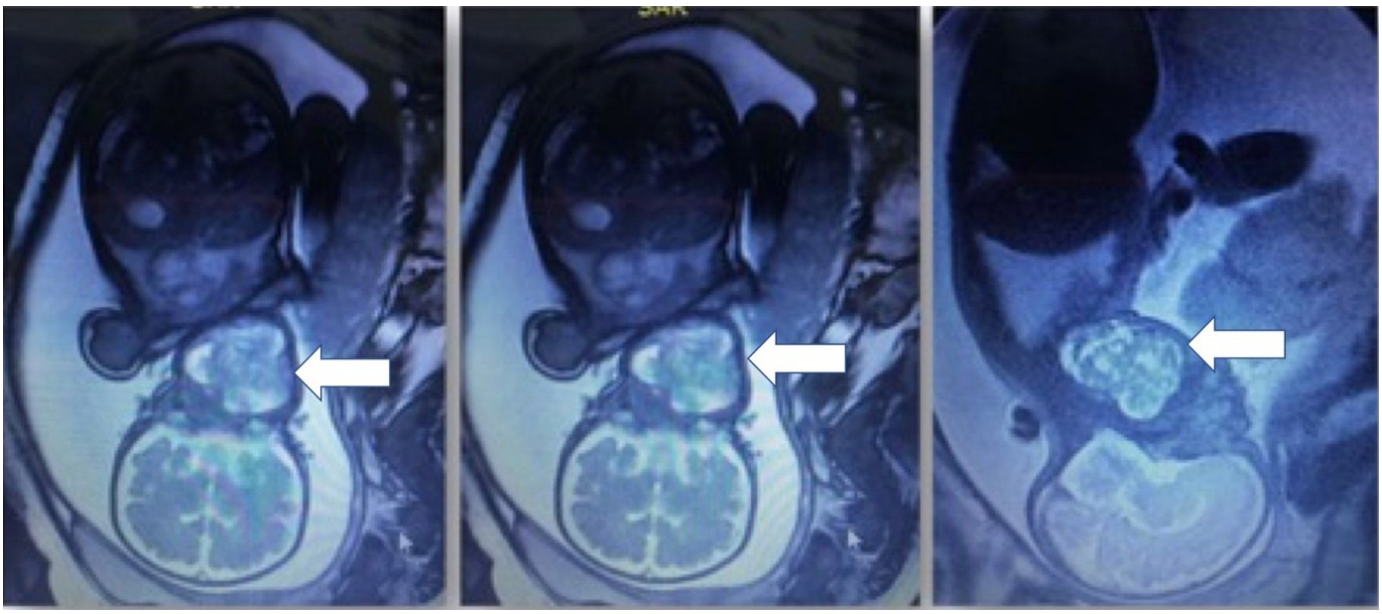

Figure 1. Right cervical mass compressing the trachea in fetal MRI (showed with arrows)

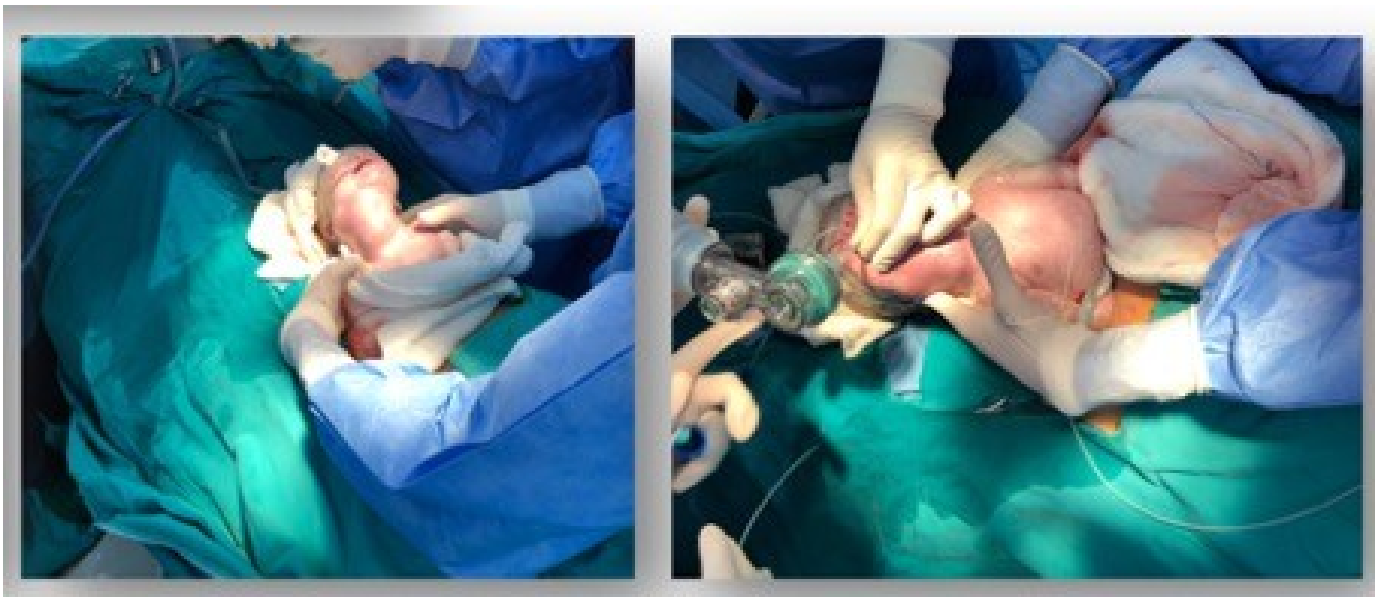

Figure 2. Orotracheal intubation of the baby with EXIT

complete tracheal compression and accompanied by polyhydramnios [2].

Preparations for laryngoscopy, video-laryngoscopy, bronchoscopy and surgical interventions were completed, and general anesthesia was provided with remifentanil and 2-3% sevoflurane (SEVORANE®) after standard monitoring and anesthesia induction of the pregnant patient. The baby's head and arms were taken out of the uterus by performing a hysterotomy. While oxygenation was maintained by the uteroplacental circulation, laryngoscopy was performed at the 5th minute and the baby was intubated with a size 2.5 uncuffed orotracheal tube (Figure 2). After ensuring airway safety and breathing, the umbilical cord was clamped. The baby's Apgar score was 6/8, and she was taken to the neonatal intensive care unit after the procedure. During the procedure, the mother's hemodynamics was stable and no complications were encountered after delivery. On the postnatal 2nd day, IV contrast-enhanced cervical computerized tomography (CT) revealed a 65x65x55mm solid mass lesion covering the right half of the neck, crossing the midline and extending to the pterygopalatine fossa, with diffuse vascular areas and a smooth contour. The mass was totally excised by cervical incision on the 3rd postnatal day and the baby was extubated on the 6th postnatal day (Figure 3). Histopathological examination of the mass was revealed an immature teratoma. Due to clean surgical margins and the immature component comprising less than 20% of the lesion, a decision was given by the oncology council for follow-up without chemotherapy. The patient was without any clinical problem during the 3rd postoperative year.

Figure 3. Right cervical solid mass with surgical excision in the postnatal period

Discussion

Developments in fetal imaging methods have enabled congenital anomalies to be recognized in the intrauterine period and be treated prenatally to avoid complications that may occur during and after delivery. Especially, high resolution ultrasonography and fetal MRI have been very helpful in determining severe congenital malformations and to plan their treatment [3,4]. In parallel with the advances in fetal imaging, the EXIT method, developed to control respiratory tract-related anomalies with high mortality risk during delivery, provides safe cardiopulmonary support while preserving placental circulation [5]. This method was first applied in patients with congenital diaphragmatic hernia and cystic hygroma in the second half of the 1990's [6,7]. Over time, especially with surgical techniques, anesthetic and tocolytic applications used in fetal interventions during EXIT, intervention time and the range of applications expanded [8].

Many congenital airway malformations such as the Congenital High Airway Obstruction Syndrome (tracheal atresia, laryngeal atresia etc.), congenital diaphragmatic hernia, head-neck-mediastinum teratomas, lymphangiomas, epulis, ranula cysts, branchial sinus cysts are associated with serious airway complications during delivery. In the past, mortality rates of up to 80-100 % have been reported in these type of malformations [1]. The primary goal is to provide a safe airway in the neck, face and mediastinum pathologies that cause full obstruction in the early postnatal period by damaging or compressing the airway structures. The use of the EXIT procedure to facilitate airway safety in a controlled manner while placental circulation continues, has been proposed as an optimal administration strategy for patients with such pathologies [9-15]. With EXIT procedure, the reported survival becomes 50% for tracheal atresia and 94% for laryngeal atresia. Cervical teratoma which is one of the extrinsic causes of fetal tracheolaryngeal obstruction, have a mortality rate of 80% if untreated and a mortality rate of 23-36% if treated with EXIT procedure.[5,16] The procedure includes intubation with direct laryngoscopy or videolaryngoscopy, bronchoscopy, tracheostomy, surgical excision, tracheoplasty operations, and more than one of these procedures may be required in severe airway malformations [17].

Although EXIT is life-saving in selected cases, it contains some risks for mother and baby. Although maternal death has not been reported in the literature, 13% of mothers may require blood transfusion due to excessive postpartum hemorrhage, and postoperative wound complications may be 10 times higher than in standard cesarean deliveries [18,19]. In the literature, in head and neck masses where airway and oxygenation cannot be provided, the mortality risk due to the EXIT procedure in the baby varies between 8-57 % [20]. In addition, emergency surgical resection may be required when airway cannot be established with minimally invasive procedures. Complications such as nerve-vascular injuries, traumatic facial nerve palsy, hypothyroidism, hypoparathyroidism may be encountered in these cases due to the limited surgical resection time and anatomical structures damaged by the mass [21]. In the later period, conditions such as feeding difficulties, obstructive sleep apnea and delay in speech development can be seen. However, it is still difficult to distinguish whether these late-period problems are directly caused by the pathology or the procedure itself [20].

Considering the risks that may arise for the mother and the baby, one should be selective about the indication and these decisions should be given by multidisciplinary councils after discussions with the parents in detail. Findings of compression on the trachea and airways acquired by ultrasonography performed in the second trimester of pregnancy should be revealed more clearly with fetal MRI. Current indications for the EXIT procedure are the presence of a solid mass larger than 5 cm in the head and neck region, compression of the trachea and airways causing tracheal deviation and polyhydramnios [20-22]. In a study of 109 cases by Jiang et al., it was shown that the presence of a solid mass and polyhydramnios were closely related with the need for the EXIT procedure at birth [22]. In addition, it is known that choosing well-equipped centers with experienced teams in disciplines that may be needed for these cases reduces the risk of morbidity and mortality [3,17,22]. Therefore, the key step is completing all necessary planning and preparations during the prenatal period. The rates of stillbirth, mortality and morbidity are quite high in cases not diagnosed prenatally or in cases where delivery occurs in unequipped centers [24,25]. Our case was evaluated by ultrasonography and fetal MRI in the prenatal period, discussed in the perinatology council with the findings obtained and the decision was taken that the EXIT procedure was suitable and needed in this patient. Airway control was provided by intubation under direct laryngoscopy with the EXIT at birth, and the procedure could be safely completed without complications in both the mother and the baby. No bleeding requiring blood transfusion was encountered in the mother.

Conclusion

In conclusion, EXIT is an important method that provides advanced resuscitation and intervention opportunity in giant neck masses with high mortality and morbidity and severe airway obstruction. The EXIT procedures performed in well-equipped experienced centers with prenatal diagnosis and multidisciplinary treatment planning are life-saving. However, in these high-risk cases, determining the correct indications and patient selection should be done in multidisciplinary councils that also involve the parents.

Conflict of Interest: The authors declare no conflict of interest.

Funding: None.

Informed Consent: Informed consent was obtained from parents for the publication of this case report.

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