CLINICAL CASE

Fetal cervical rhabdomyosarcoma
Rabdomiosarcoma cervical fetal

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ABSTRACT

We present the case of a pregnant woman in her third trimester who was referred for management of a fetal cervical tumor with a risk of airway obstruction during delivery. The importance of prenatal referral for fetal cervical tumors and the need for a multidisciplinary approach to ensure optimal management were vital in this case. Fetal magnetic resonance imaging (MRI) plays a crucial role in assessing tumor extent and surgical planning. Although fetal cervical rhabdomyosarcoma is rare, its inclusion in the differential diagnosis is essential for appropriate treatment. Successful management of fetal cervical tumors that threaten the airways requires timely and coordinated intervention, with priority given to referring these cases to centers with medical teams trained to perform procedures such as EXIT surgery and chemotherapy, which contributes to a better prognosis for both the mother and the newborn.

Keywords: Rhabdomyosarcoma; embryonal rhabdomyosarcoma; fetal diagnosis; referral and consultation (source: MeSH NLM).

RESUMEN

Se presenta el caso de una gestante en el tercer trimestre de gestación referida para manejo de un tumor cervical fetal con riesgo de obstrucción de la vía aérea durante el parto. La importancia de la referencia prenatal de tumores cervicales fetales y la necesidad de un enfoque multidisciplinario para asegurar un manejo óptimo fueron vitales en este caso. La resonancia magnética fetal desempeña un papel crucial en la evaluación de la extensión del tumor y en la planificación quirúrgica. Aunque el rabdomiosarcoma cervical fetal es poco común, su inclusión en el diagnóstico diferencial es esencial para un tratamiento adecuado. El manejo exitoso de las tumoraciones cervicales fetales que amenazan las vías respiratorias requiere una intervención oportuna y coordinada, siendo prioritaria la necesidad de referir estos casos a centros con equipos médicos capacitados para llevar a cabo procedimientos como la cirugía EXIT y la quimioterapia, lo que contribuye a un mejor pronóstico tanto para la madre como para el neonato.

Palabras clave: Rabdomiosarcoma; rabdomiosarcoma embrionario; diagnóstico fetal; referencia y consulta (fuente: DeCS BIREME).

Introduction

Solid masses in the fetal cervical region are rare anomalies that can obstruct the airways, with high risks of neonatal morbidity and mortality[1]. In some cases, their excessive growth can block the airway externally, increasing the danger to the newborn[2]. The difficulty in securing the airway at birth represents a significant clinical challenge and often leads to problems of perinatal asphyxia due to failure of orotracheal intubation.

Rhabdomyosarcoma (RMS) is the most prevalent malignant neoplasm in pediatric soft tissues, although its presentation in the neonatal or fetal period is extremely rare[1]. It is important to diagnose these tumors or masses prenatally, especially in high-risk pregnancies, in order to adequately plan prenatal care and perinatal management[3]. Tumors in the head and neck can be detected by two-dimensional ultrasound, providing information on tumor volume. However, magnetic resonance imaging (MRI) may surpass ultrasound in identifying possible tumor spread[3]. Fetal magnetic resonance imaging can help select fetuses that will require upper airway management during delivery through the ex utero intrapartum treatment (EXIT) procedure at a higher-complexity center[3].

We present a case of fetal cervical tumor confirmed by magnetic resonance imaging performed in utero. In addition, we present specific images of the diagnosed fetal cervical tumor. This case highlights the importance of coordinating accurate and timely referral to centers of greater complexity.

Case report

We present the case of a 34-year-old multiparous woman, G6 P4014, who was admitted to Ayacucho Regional Hospital at 36 weeks and 3 days of gestation due to the presence of a fetal cervical mass and a high risk of airway obstruction in the fetus.

An ultrasound identified a solid fetal cervical mass with suspected airway obstruction, which was confirmed by fetal magnetic resonance imaging. The latter showed a solid tumor measuring 146 x 96 x 93 mm extending from the right nasogenian and fetal cervical region to the upper third of the thorax, without compromising the upper airways of the fetus (Figure 1A, 1B).

Following a multidisciplinary assessment and review by a medical board, it was decided to refer the patient to a more complex center for timely management with a multidisciplinary approach, considering the difficulty of managing the fetal airways. Despite the difficulties inherent in the cervical mass, the multidisciplinary team successfully accessed the airway via endotracheal intubation to perform emergency EXIT surgery, optimally stabilizing the newborn.

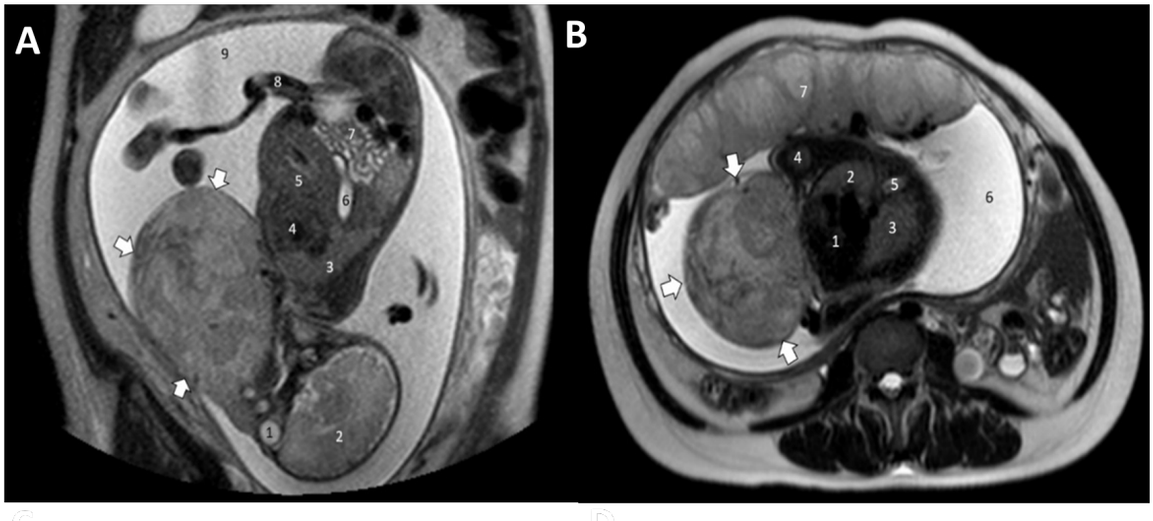
The newborn female infant was transferred to the Neonatal Intensive Care Unit after the airway management described (Figure 2A). After 11 days, she underwent excision surgery plus radical resection by Head and Neck Surgery, achieving favorable surgical results (Figure 2B). However, the pathological anatomy result was consistent with embryonal rhabdomyosarcoma (Figure 3A, 4B). Therefore, she was referred to pediatric oncohematology and is currently undergoing chemotherapy with a favorable response, with case follow-up conducted up to 2 years of age. (Figure 4A, 4B).

Discussion

Fetal cervical tumors are a rare condition that can cause severe airway obstruction with fatal consequences for an otherwise healthy newborn. Once diagnosed, it is crucial to determine the extent of the mass and the level of airway compromise using an ultrasound performed by experts and, if possible, supplemented with fetal magnetic resonance imaging, as in our case[1].

Although the characteristics of rhabdomyosarcomas in magnetic resonance imaging are somewhat nonspecific[4], this technique allows for more detailed characterization of the tumor and surrounding tissue[5], making it the modality of choice[6].

Figure 1: (1A) Sagittal magnetic resonance imaging (MRI) scan showing a large heterogeneous mass at the facial, cervical, and thoracic levels of the fetus (arrows), measuring 14 cm in length. (1B) Axial MRI scan showing a large heterogeneous mass at the thoracic level of the fetus (arrows).



ure 2: (2A) Newborn hospitalized in the neonatal intensive care unit. (2B) Newborn post-surgery for cervical tumor excision.

Teratoma is the most common cause of solid mass in the fetal neck, and the incidence of fetal teratoma varies between 1 in 20,000 and 40,000 live births. Cervical teratomas causing airway obstruction have been successfully managed by EXIT surgery[2, 8]. Unlike teratomas, congenital rhabdomyosarcoma is extremely rare, although rhabdomyosarcoma is the most common soft tissue tumor in children. Embryonal rhabdomyosarcoma, the most common type of rhabdomyosarcoma, responds well to chemotherapy, allowing for less aggressive surgery if necessary[7].

Although there are few reports of rhabdomyosarcoma being treated with EXIT, there is one case report in which an EXIT procedure was planned for the treatment of one of the fetuses in a twin pregnancy with cervical rhabdomyosarcoma before fetal hydrops developed; however, the parents refused to give consent due to concerns about the delivery of the normal fetus[9].

In the 1960s, less than one-third of children with rhabdomyosarcoma survived, but cure rates are now approximately 70%. Survival has improved over the past 40 years, especially for patients with localized disease[10]. Embryonal rhabdomyosarcoma, the most common type of rhabdomyosarcoma, generally responds very well to chemotherapy. According to the Intergroup Rhabdomyosarcoma Study IV, patients with resectable tumors and positive lymph nodes showed the greatest benefit from treatment, with a disease-free survival rate that increased from 72% to 92% over a three-year period[11]. In the case we present, the tumor was removed along with a radical lymph node dissection, followed by chemotherapy, achieving a favorable outcome up to two years of follow-up. This highlights the importance of performing surgi-

Figure 3: (3A) Histopathology with hematoxylin and eosin (HE) staining revealed embryonal rhabdomyosarcoma with pleomorphic tumor cells accompanying rhabdomyoblasts in areas of infiltration into smooth muscle. (3B, 3C) Immunohistochemistry showed that the tumor was positive for desmin and myogenin (arrows).

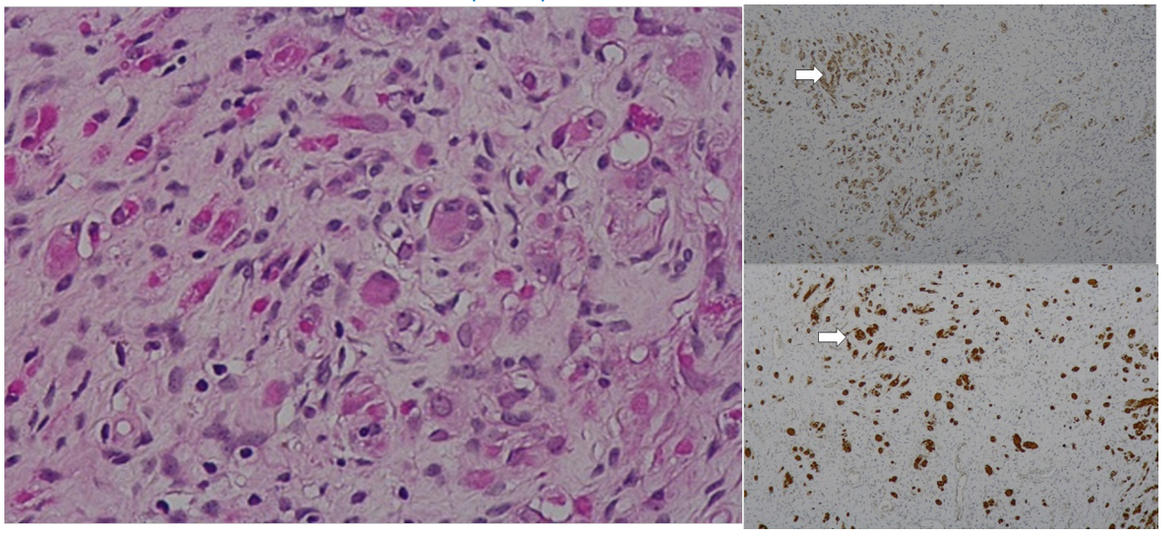


Figure 4: (4A) Newborn undergoing chemotherapy sessions. (4B) Favorable response to date.

cal intervention at the appropriate time and under the correct indications to maximize survival chances and minimize morbidity.

The EXIT procedure is a surgical technique that maintains maternal-fetal circulation (uteroplacental bypass) during cesarean delivery, allowing sufficient time to secure an airway through procedures ranging from orotracheal intubation to surgical excision of the obstructive lesion[12]. It is important to note that, as this procedure is specific to a high-capacity center, the most appropriate referral was decided upon, considering the advanced gestational age[13].

When managing a congenital tumor that compromises airway access, it is essential to have a multidisciplinary team and the necessary materials in place to avoid the morbidity and mortality associated with this scenario[14]. Although the patient was referred late, the intervention was possible thanks to the optimal and satisfactory organization of the entire surgical team. These cases should be referred to a center with the capacity to resolve them as soon as the diagnosis is made, in order to implement the appropriate protocol, which includes a complete fetal morphological study, mapping of the extent and invasion of the lesion by ultrasound, and if necessary, magnetic resonance imaging as in our case, and convening the multidisciplinary team to perform a scheduled procedure at a more complex center[15].

Conclusion

The management of fetal cervical tumors that cause airway obstruction requires timely diagnosis and referral to a center with greater capacity, equipped with a multidisciplinary team capable of offering options such as EXIT surgery or other minimally invasive intrauterine management techniques, in order to reduce risks and complications for both the mother and the newborn.

Although extremely rare, fetal cervical rhabdomyosarcoma should be considered in the differential diagnosis of a solid mass in the fetal cervical region, which requires a multidisciplinary approach to obtain better surgical and chemotherapy response outcomes, preferably in a tertiary hospital.

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