Congenital cervical teratoma – an amalgamation of embryology with clinical findings

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SUMMARY

Teratoma means "A malformed body part of a monster" in Greek. A cervical teratoma is a very rare congenital tumor in the neck. These tumors tend to be large, disfiguring masses – partly solid and partly fluid. They make it impossible for a newborn to breathe upon birth. Knowledge of basic embryology of the brachial apparatus is necessary to understand neck malformations and congenital abnormalities. Here, a recent observation of this exceptional case is described. A pregnant woman, gravida 2, presented with pain abdomen in the 37th week of gestation. A male, living baby was delivered by caesarean section. A well-defined mass was present in the anterior neck region. A diagnosis of giant congenital cervical teratoma with airway compromise was made. The baby was intubated but could not survive. The objective of this study is the significance of a thorough knowledge of anatomy and embryology to prevent any late diagnosis or misdiagnosis. This report aims to create awareness about embryological development of a fetus to enhance the clinical recognition of this rare disorder, to highlight their occurrence in our locale, and to reiterate the associated management challenges in resource-limited settings. The present case highlights the importance of regular antenatal checkups, with timely ultrasounds, so that such congenital defects can be diagnosed prenatally and their management can be planned accordingly.

Key words: Teratoma – Neck – Embryology – Fetus – Brachial cleft – Congenital anomaly

INTRODUCTION

Congenital cervical teratomas are extremely rare germ cell tumors of the neck, composed of tissues derived from at least two of the three embryonic germ layers, but foreign to the anatomic site of occurrence (Benhoummad et al., 2021). These tumors are benign but may lead to serious consequences. Congenital cervical teratomas of the neck region compress the structures in the neck region and make breathing difficult for the baby (Benhoummad et al., 2021). Teratoma of the head and neck are interesting because of their obscure origin, unpredictable behavior and often dramatic clinical presentation. Teratomas are embryological neoplasms that arise when totipotent germ cells give rise to more or less organoid masses in which tissues are derived from all the three blastomeric layers (ectoderm, endoderm and meso-

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derm) (Alharbi et al., 2017). Cervical teratomas are rare benign tumors consisting of 3% of all teratomas and occur one in every 20,000-40,000 live births (Mohammed et al., 2021). These neoplasms are asymmetrical with both cystic and solid areas.

Knowledge of basic embryology of the brachial apparatus helps to understand neck malformations and congenital abnormalities. During the third week of development, the flat trilaminar embryo undergoes a series of complex folds that result in the formation of a cylindrical embryo. During this time, the laterally placed clefts, known as branchial clefts appear. These clefts are due to flexion folds of the fetus within the amniotic cavity. The basic tissues of development within the head and neck (ectoderm, endoderm, mesoderm, neuroepithelium) become organized into the pharyngeal apparatus, also known as the branchial apparatus, which is the forerunner of the head and neck structures.

Cervical teratomas often occur on the anterolateral surface of the neck, extending midline from the thyroid gland, as far as 12 cm in their longest axis; hence, unsuspected obstructive fetal giant neck masses often prove fatal, because of an inability to secure the airway and ventilate the baby upon delivery, which leads to hypoxia and acidosis (Mohammed et al., 2021).

Current evidence suggests that most teratomas are due to abnormal differentiation of fetal germ cells that arise from the fetal yolk sac. Normal migration of these germ cells may cause gonadal tumors, while abnormal migration produces extra gonadal tumors. Chromosomal abnormalities that have been reported are trisomy 13, ring X chromosome mosaicism with inactive ring X

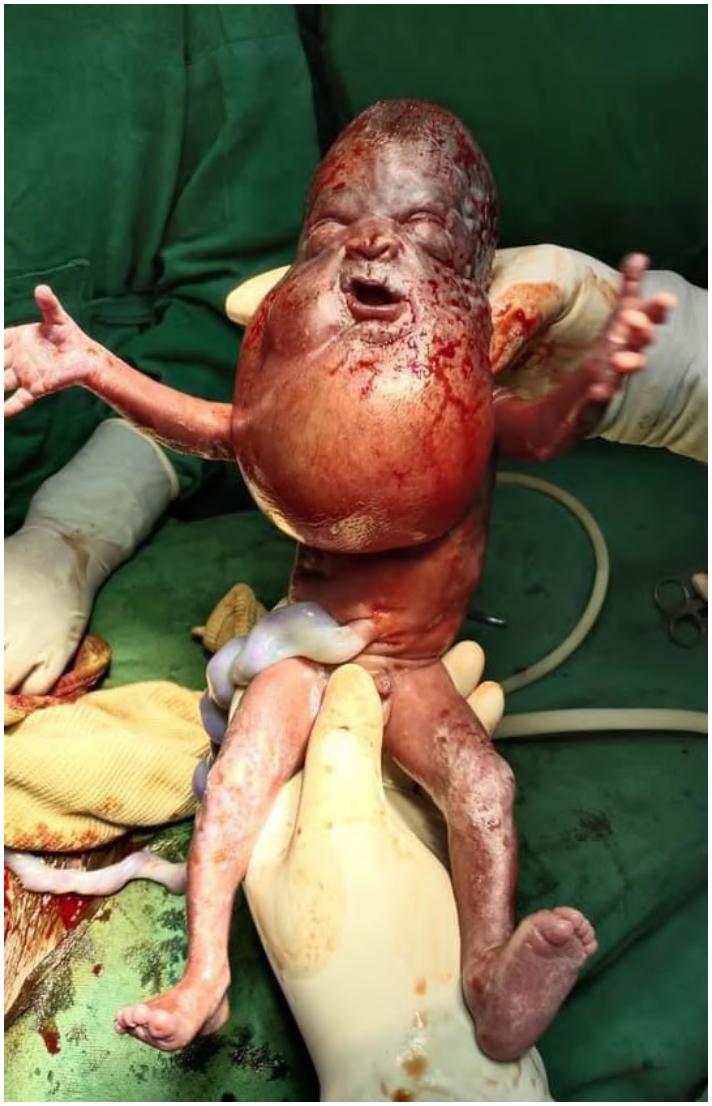

Fig. 1.- A male baby with congenital cervical teratoma.

chromosome, gene mutation or abnormalities in early embryonic development (Mohammed et al., 2021). The etiology can be also aberrant fertilization, abnormal meiotic division, or even asexual development of an unfertilized ovum (Azam et al., 2021). Giant neck masses can obstruct the airway and cause serious ill effects, which are not compatible with life after delivery.

CASE REPORT

A pregnant woman of gravida 2, aged 28 years, presented with severe pain abdomen in the 37th week of gestation. On examination, the abdomen was over-distended, with fundal height of 40 weeks. As she came from a rural background, there was no history of any previous ultrasounds. There was history of a previous abortion. An ultrasound examination was done immediately. Her ultrasound report revealed a single, viable, cephalic fetus with a cystic and solid mass in the neck region. The length and breadth of the neck mass were 9.2 cm and 6.9 cm respectively. Both solid and cystic portions containing blood vessels were present. Moderate polyhydramnios was detected. No other congenital abnormality was detected. The parents were counseled about the outcomes, their consent was received and a decision was taken to proceed further with the EXIT (Extra Uterine Intrapartum Treatment) procedure.

A multidisciplinary team was formed which included anaesthetic, obstetricians, pediatric surgeon and a radiologist. A male, living baby weighing 1.8 Kg was delivered by caesarean section. A well-defined, predominantly solid lesion was present in the anterior neck region (Fig. 1). The baby was apneic, centrally cyanosed with a heart rate (HR) of 50 beats per minute. Apgar scores were 1 and 2 at 1 and 2 minutes. A diagnosis of giant congenital cervical teratoma with airway compromise was made. The baby was shifted to NICU (Neonatal Intensive Care Unit) and intubated immediately with much difficulty. The mass was irregularly-shaped with visible superficial vessels and spread to the lower half of face, including the mandible and both temporomandibular joints. A Cervicofacial Computerised Tomography (CT) scan was planned to enquire about any infiltrations, but unfortunately, he only survived for a few hours and then died. The parents were referred for Genetic Counseling.

DISCUSSION

Perinatal mortality is very high in cases of cervical teratoma. A large cervical teratoma usually causes airway obstruction, and any delay in treatment can cause 80% to 100% mortality (Azam et al., 2021). A congenital cervical teratoma is difficult to diagnose in utero, because of the similarities with other fetal neck masses such as cystic hygroma., branchial cyst, hemangioma or lymphangioma (Azam et al., 2021). To prevent such misdiagnosis, thorough knowledge of anatomy and embryology is crucial. Embryologically, brachial cleft cysts are developed by incomplete involution of brachial cleft structures; hence, they are present more posterior and lateral in position. Similarly, cystic hygroma, lymphangioma and hemangioma are also present laterally. On the other hand, cervical teratomas are usually found in mid-line structures such as anterior neck region, pineal body and supra-sellar space (Uchiyama et al., 1995).

CONCLUSION

The present case study is a compelling amalgamation of knowledge of embryology with radiology, obstetrics and pediatrics. Although rare, cervical teratomas can be diagnosed in utero. This particular case study highlights the importance of antenatal checkups along with anatomical anomaly scan. The importance of a skilled multidisciplinary team with complete understanding of the anatomical relations of the neoplasm is also emphasized. To ensure a successful management, careful planning and coordination is the key. The majority of these tumors are benign and cured by complete resection of the mass during neonatal period (Peiro et al., 2016). The author recommends a comprehensive prenatal evaluation including conventional ultrasounds, doppler, and echocardiography along with anatomical anomaly scan to be made compulsory before 20 weeks of gestation. Antenatal counseling helps the parents to better understand the natural history, fetal intervention and perinatal management[6]. Late diagnosis, lack of pre-plan-

ning and ignorance can lead to both physical and mental turmoil. Counseling, serial imaging, and well-planned deliveries with EXIT procedure are the mainstay of management (Mohammed et al., 2021; Azam et al., 2021; Uchiyama et al., 1995). Favorable prognosis depends on prenatal diagnosis by radiological examinations and proper management.

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