Case Report

BIOMEDICAL Journal of Scientific & Technical Research

ISSN: 2574 -1241

DOI: 10.26717/BJSTR.2024.59.009300

Cranio Cervico Facial Oral Teratoma in a 32-Day-Old Infant with Intrauterine Diagnosis

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ARTICLE INFO

Received: October 31, 2024

Published: November 12, 2024

Citation: Maria Eugênia Carvalho, Luiz Eduardo Imbelloni, Anna Lúcia Calaça Rivoli, Sylvio Valença de Lemos Neto, Ana Cristina Pinho, Sara Pereira Lima Soares de Sá and Isabela Moreira Silva. Cranio Cervico Facial Oral Teratoma in a 32-Day-Old Infant with Intrauterine Diagnosis. Biomed J Sci & Tech Res 59(3)-2024. BJSTR. MS.ID.009300.

ABSTRACT

Background: Congenital teratomas of the oropharyngeal cavity are extremely rare and are associated with a high neonatal mortality rate due to severe airway obstruction. Treatment has improved with progress in diagnosis during pregnancy. The authors describes a case of an infant who underwent surgery at 32 days of age.

Case Report: Intrauterine diagnosis was made at 36 weeks of gestation, with a right-sided cranio cervical facial-oral tumor. The Cesarean section was indicated, performed at 37 weeks, and tracheostomy after birth. At 18 days, a biopsy revealed the diagnosis. The baby was prepared with red blood cell replacement and operated on at 32 days of extrauterine life. General anesthesia and surgery lasted 6 hours, and the baby was transferred to the ICU, being extubated on the 2nd postoperative day. The final diagnosis was mature teratoma.

Conclusion: Surgery for head and neck tumors in newborns, most of which are benign, is very challenging and requires a multidisciplinary team. Perinatal planning allows for appropriate management at birth, reducing the risk of airway obstruction. Surgery remains the mainstay of teratoma treatment, with excellent aesthetic results. The final diagnosis of mature teratoma was made on histopathological examination.

Keywords: Congenital Disease; Cervical Tumor; Oral Teratoma; Pediatric Head; Neck Surgery; Mature Teratoma; Multidisciplinary Team

Introduction

Teratomas are germ cell tumors and are commonly composed of multiple cells derived from one or more of the three germ layers. Immature teratomas represent 10% of germ cell tumors (GCT) and 42% of malignant germ cell tumors [1]. In Brazil, in 2020, there were 2,289 deaths from childhood cancer, corresponding to 38.20/million [2]. The number of new cases of childhood cancer estimated for Brazil, for the three-year period from 2023 to 2025, is 7,930 cases, which corresponds to an estimated risk of 134.81/million children and adolescents [2]. When the diagnosis of teratoma is made in utero, close monitoring of affected fetuses should be considered, as fatal complications can be associated with any teratomas, as they grow rapidly and are highly vascular; these include hydrops fetalis, heart failure, and polyhydramnios [3]. This monitoring is achieved with serial ul-

trasound (US) to determine the relationship between tumor volume and fetal weight, a predictive tool for fetal outcomes [4]. In fetuses with a gestational age greater than 28 weeks, elective delivery may be offered. In contrast, in fetuses of younger age who are at high risk of intrauterine hemorrhage or vascular steel, surgical procedures are necessary for fetal survival [5]. Teratomas most commonly occur in the sacrococcygeal region but can arise anywhere along the midline of the body. Head and neck teratomas account for 1% to 9% of all teratomas [6]. When they occur in the head and neck region, the cervical and nasopharynx are the most common sites of occurrence, and they have the potential to cause airway obstruction.

Due to their infrequent occurrence, there is little evidence on the diagnosis and treatment of immature teratomas diagnosed during pregnancy. We present a case of an immature teratoma diagnosed at 36 weeks of gestational age, through US and subsequent fetal magnetic resonance imaging, which showed an expansive solid-cystic formation (58x21x55 mm), with partially defined limits, which obliterated the oral cavity and oropharynx and extended superiorly to the right middle cranial fossa, pushing the temporal lobe superiorly, which allowed for a scheduled cesarean section with a multidisciplinary team.

Case Report

A protocol was recently created to study pediatric patients undergoing solid tumor resections, registered on the Plataforma Brasil (CAAE: 74205517.3.0000.5274) and approved by the Ethics and Research Committee of the National Cancer Institute (INCA) under number 6,706,870. Fetal ultrasound and MRI performed at 36 weeks, showing a mixed cyst and solid image in the right temporal region, cervicosternal region, obliterating the oral cavity and oropharynx, and extending superiorly to the right middle cranial fossa, superiorly pushing back the temporal lobe, measuring approximately 58x21x-55mm. The preparation of this case followed the guidelines of the Department of Anesthesia and Pediatric Oncology Surgery of the National Cancer Institute. Male infant, born after 37 weeks of gestation by cesarean section, weighing 3,500 g, with intrauterine diagnosis of a right-sided cranio cervical facial-oral tumor, having been tracheostomy performed at birth. At 18 days of life, a tumor biopsy was performed showing glial heterotopia, with subsequent confirmation of mature teratoma, and surgery was indicated 32 days after birth. Preoperatively, he presented episodes of SpO2 drop due to obstruction of the tracheostomy tube and hemodynamic instability, requiring infusion of adrenaline 0.1 μg/kg/min, with satisfactory response. The infant received packed red blood cells 11 days and 3 days before the surgical procedure and on the day of surgery. The infant was referred to the surgical center without preanesthetic medication.

After checking the basilic vein, monitoring with continuous ECG, noninvasive blood pressure, invasive blood pressure, pulse oximetry, capnography, gas analysis and BIS, he was induced under a mask through the tracheostomy. Balanced general anesthesia with continuous infusion of dexmedetomidine 0.5 μg/kg/h, fentanyl 1 μg/kg/h, rocuronium 0.5 mg/kg/h, ketamine 0.2 mg/kg/h, the metal cannula was replaced by a tracheal tube no. 3 with cuff through the tracheostoma. After the replacement, he had an episode of bronchospasm with a drop in oxygen saturation, reversed with the administration of inhaled salbutamol 200 μg, adrenaline 2 μg and hydrocortisone 35 mg intravenously. Puncture of the right femoral artery and venous dissection of the left internal jugular vein were performed (Figure 1). The surgery performed was cervical and middle fossa tumor resection, associated with right lateral pharyngotomy lasting six hours, and at the end of the surgery he was transferred to the pediatric ICU, sedated, on mechanical ventilation, with blood gas parameters showing compensated respiratory acidosis and lactate of 2.2 mmol/l (Figure 2). Postoperatively, the patient progressed well, being weaned from amines on the 2nd day and from oxygen therapy on the 29th day. He was receiving respiratory physiotherapy and was monitored by pediatric surgery and speech therapy. Three and 13 days after the procedure, he received red blood cell concentrates, and the examination performed 29 days after surgery revealed: Hb=9.3 g/l, Ht=26.6%, platelets=524,000 mm^3.

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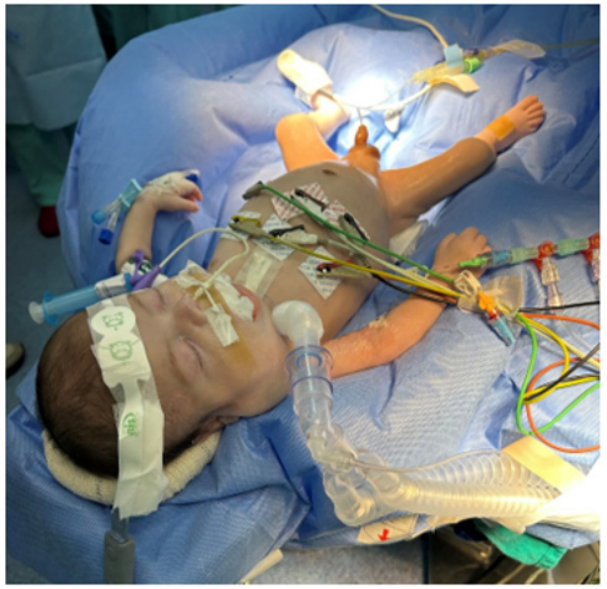

Figure 1: Anesthetized infant ready to begin surgery.

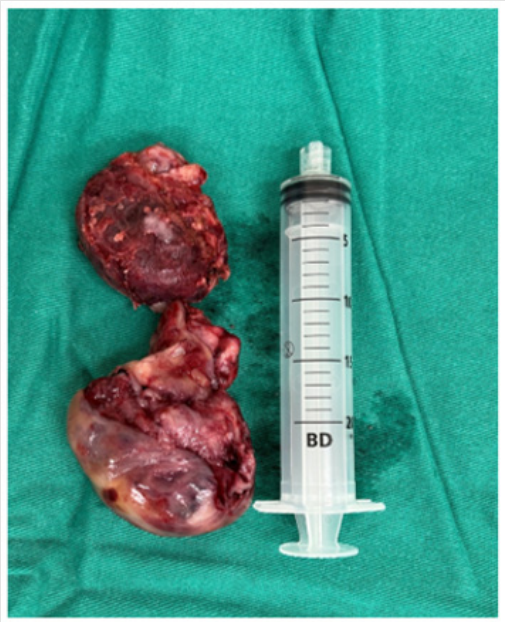

Figure 2: Size of the removed part.

After extubation, the infant was awake, calm, following the examination with his eyes and smiling, with normotensive anterior fontanelle, facial asymmetry, depression in the mandibular region on the right, slight increase in the cheek, and the fundus with a good appearance, hydrated, pale, acyanotic and anicteric. The parameters were HR=129 bpm, RR=38 irpm, SpO2=96% in room air, Tax=36.5º C and BP=88 x 53 (66) mmHg. The pathological anatomy showed macroscopically a tumor of two irregular segments measuring 9.0 x 7.0 x 3.5 cm together, partially covered by a smooth, shiny and transparent capsule. Microscopy revealed neoplasia composed of glial tissue, choroid plexus epithelium and retina, in addition to adipose, muscular, cartilaginous, bone, respiratory and glandular epithelium. Presence of immature neural tissue in a focal area (1% of the sample). The analyzed material was compatible with immature/malignant teratoma. Three months later, the report was reviewed and diagnosed as congenital craniofacial mature teratoma (epignatus). Magnetic resonance imaging performed two months after surgery showed subtotal craniofacial resection on the right, deformity of the remaining anatomical structures and a residual retro maxillary lesional component that projects into the oral cavity and is contiguous to the sphenoid.

Discussion

This infant's teratoma was initially diagnosed as immature and later confirmed as mature. One to 9% of all teratomas occur in the head and neck region, predominantly in the cervical region [6]. One of the main challenges in anesthesia of infants with large cranio cervical tumor masses is airway management. In this case, tracheostomy was performed immediately after birth and was assured at the time of surgery and anesthesia. At the beginning of anesthesia for the surgical procedure, the removal of the metal cannula to introduce a tube tracheal through the tracheostoma caused bronchospasm and significant desaturation, which were promptly reversed with usual treatment maneuvers. Teratomas in the head and neck region may present respiratory distress, and immediate excision is necessary, and surgery is curative, which is why tracheostomy was performed immediately after birth, given the size of the tumor. Several hypotheses have been proposed to explain the formation of teratomas. The most widely accepted hypothesis is that of abnormal proliferation of pluripotent cells sequestered during embryogenesis, which can form disorganized structures comprising various types of tissues foreign to the region [7]. Teratomas are complex tumors composed of various lineages of embryonic cells: ectoblastic, endoblastic, neuroblastic or mesenchymal. Prenatal diagnosis allows for optimal treatment planning. Neonatal teratomas of the head and neck are often present in the form of neonatal airway obstruction (35%) with a high mortality rate [8].

Intrauterine diagnosis was performed at 36 weeks but allowed the newborn to be treated immediately to avoid airway obstruction and to schedule surgery under better conditions at 32 days of life. Intraoperative hemodynamic management used restricted volume replacement to avoid hypervolemia and pulmonary congestion, based on a recently published guideline [9]. Adrenaline infusion and the need for packed red blood cell transfusion during the procedure were guided by macro hemodynamic parameters such as heart rate and AIP, and micro hemodynamic parameters such as central venous saturation, CO_2 gap and lactate clearance, with good results and no complications during the procedure. Patients undergoing early resection surgery, with an average duration of 4 days, presented minimal perioperative complications. In a scoping review, one hundred and eight studies totaling 137 patients were identified, showing that the median gestational age at birth was 37 weeks, respiratory distress, which led to emergency tracheal intubation or tracheostomy, was present in the majority (58%) of patients [10]. Data confirm the report of this case, born at 37 weeks of gestation, but it was only possible to perform the surgery at 32 days of extrauterine life. The cesarean section is recommended for newborns with very large cervical tumors to avoid dystocia during labor [11]. The presence of a multidisciplinary team in the delivery room is recommended, even when prenatal evaluation shows no signs of airway obstruction. These recommendations were practiced at the birth of the newborn, because the tumor showed solid-cystic expansile formation, and tracheostomy was immediately performed.

Conclusion

Early surgical approach for congenital cervical teratomas allows the best results, with low complication and recurrence rates, and usually with a good aesthetic result. The indication of cesarean section for birth is essential if the tumor is very large and a multidisciplinary team and monitoring are recommended for early diagnosis and surgical excision. Neonatal teratomas of the head and neck are rare and are usually mature benign tumors. Early surgical intervention is still recommended, especially for teratomas that are causing airway obstruction, which was demonstrated in this case. Diagnosis is sometimes possible only after histopathological examination. In this case the preoperative diagnosis was epulis, but review of the histopathology revealed a congenital craniofacial mature teratoma (epignatus).

Authorization for the Use of Images

Authorization for the use of images for publication in TCC and in a medical journal was signed by the parents.

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ISSN: 2574-1241

DOI: 10.26717/BJSTR.2024.59.009300

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