Successful in utero treatment of an oral teratoma via operative fetoscopy: case report and review of the literature

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Neonatal teratomas are rare tumors that arise from all 3 embryonic germ layers: endoderm, mesoderm, and ectoderm. The incidence of all teratomas is approximately 1/4000. Most tumors arise in the midline, the most common sites being the sacrococcygeal region, anterior mediastinum, testicles, and ovaries in that order. Teratomas of the head and neck region constitute 1-9% of all teratomas. The incidence of teratomas of the head and neck is between 1/35,000 and 1/200,000 live births.

Pure oral teratomas, or epignathi, occur in the sphenoid region on the palate or pharynx (Rathke’s pouch) and may rarely be found in the nasal cavity of the hard palate. Epignathi are thought to result from migration abnormalities of primordial cells settling in the mediastinum or hypothalamic regions and have a female to male ratio of 3:1.

Prenatal diagnosis of an oral teratoma can be made with ultrasound as early as 12 weeks and in the second trimester. However, most tumors are diagnosed in the third trimester, suggesting in utero growth or subsequent development. On ultrasound, the mass emerges from the fetal mouth and may produce hyperextension of the head. Oral teratomas are also associated with cleft palate and other oral malformations in 6-20% of patients.

The prenatal diagnosis of a nasopharyngeal teratoma carries a very grave prognosis. Although these tumors constitute only 9% of all teratomas, all previous cases diagnosed antenatally have been associated with either fetal demise or emergent surgery at birth. If the fetuses that survive to birth, delivery can be associated with airway obstruction and multiple postnatal surgeries. These complications could be averted if the tumor could be safely treated in utero. We hereby report the successful treatment of an oral teratoma via operative fetoscopy, with the birth of a healthy infant at term.

Key words: fetal surgery, nasopharyngeal tumor, operative fetoscopy, oral teratoma

The natural history of these tumors in utero include possible development of polyhydramnios, presumably secondary to pharyngeal obstruction, as well as unexpected fetal death. Perinatal management involves either intrapartum intubation (ex utero intrapartum treatment [EXIT procedure]) or resection of the tumor at the time of the cesarean section and prior to cutting the umbilical cord (operation on placental support [OOPS]). Despite all efforts, survival rates are typically in the 30-40% range. The purpose of this communication is to report successful in utero resection of a nasopharyngeal teratoma via operative fetoscopy.

Case Report

A 37 year old woman, G4 P1031, with an uncomplicated prenatal course was diagnosed at 20 weeks’ gestation using two-dimensional and three-dimensional ultrasound as having a fetus with a pedunculated 4.1 × 2.4 × 2.8 cm mass protruding from the fetal mouth. The tumor had cystic and solid components. Cleft lip or palate did not appear to be present, but they could not be entirely ruled out with ultrasound. A small feeding arterial vessel was identified at the stalk of the tumor using color and pulsed Doppler, although the actual anatomical site of origin of the mass could not be determined (Figure 1, A and B). Polyhydramnios, hydrops, or abnormal arterial or venous Dopplers were not present at the time of diagnosis. Fetal magnetic resonance imaging (MRI) confirmed an exophytic complex mass arising from the left paramedian aspect of the midface of the fetus. No obvious intracranial extension was apparent. Findings were suggestive of an oral teratoma.

Options discussed with the patient included expectant management, which could result in increased tumor size, polyhydramnios, hydrops, or fetal demise. If the fetus survived to an appropriate gestational age, delivery could require an EXIT procedure or an OOPS procedure. Termination of pregnancy was also offered. The patient was particularly concerned about the possible existence of oral clefting, which, if present, would make her consider the option of pregnancy termination more seriously. Diagnostic and possibly operative fetoscopy was also discussed as an alternative. The purpose of diagnostic fetoscopy was to assess the lips and palate and rule out clefting. In addition, if it were considered feasible, an attempt would be made to either devascularize or to resect the lesion altogether.

After serious consideration, the patient requested to undergo fetoscopy. The risk of pregnancy loss was quoted as 5%. The procedure was approved by the University of Miami and Jackson Memorial Hospital under the innovative therapy pathway. Amniocentesis revealed a normal 46, XX karyotype.

The patient was taken to the main operating room. She received Remifentanil intravenously at a rate of 0.1 μg/kg per
minute, increased by 0.025 μg/kg per minute increments up to a maximum dose of 0.15 μg/kg per minute. Remifentanil crosses the placenta readily and provides fetal analgesia. Under local anesthesia, a minimal skin incision was made. A 3.8 mm trocar (Richard Wolf Inc, Vernon Hills, IL) was inserted percutaneously under ultrasound guidance into the amniotic cavity. The fetal mouth was assessed externally and internally using a 25° diagnostic endoscope (Richard Wolf Inc.). There was no evidence of cleft lip or palate, findings that were relayed to the patient intraoperatively.

The teratoma was noted to have a distinct stalk, semirotated clockwise, stemming from the hard palate anteriorly and slightly off the midline to the left (Figure 2), confirming the ultrasound and MRI findings. Consideration was given at this point to proceed with excision of the mass. The tumor was resected at the base using a 600 micrometer contact YAG laser fiber on continuous mode with 10 W of energy. A minimal amount of bleeding from the base of the tumor was controlled with a noncontact YAG laser fiber. The tumor fell inside the amniotic cavity, where it was left. The procedure lasted 68 minutes. There were no maternal or fetal complications.

The postoperative course was uncomplicated. Serial weekly ultrasounds showed normal fetal mouth and profile, without recurrences. The amniotic fluid volume remained normal throughout the pregnancy. The patient went into spontaneous labor at 38 5/7 weeks. A female infant was born vaginally without complications, weighed 8 lb, and had Apgars of 9 and 9 at 1 and 5 minutes, respectively. Inspection of the lips and mouth at birth showed no evidence of clefting, injury, or residual mass (Figure 3). Surgical pathology showed necrotic tissue with cartilaginous differentiation, suggestive of a nasopharyngeal teratoma. Postnatal MRI at 1 month of life showed no evidence of recurrent mass in the brain or face.

**Comment**

To our knowledge, this is the first successful treatment of a fetal oral teratoma in utero. The experience in this case suggests that fetoscopy can be of use in the detailed assessment of the lesion as well as potentially allowing resection of the mass in utero in selected cases. In utero resection is aimed at avoiding the development of polyhydramnios, preterm labor/delivery, or potential fetal demise that could occur with expectant management. Furthermore, in utero resection allows for vaginal delivery and eliminates...
the need to perform an EXIT or OOPS procedure, with its attendant logistic and medical disadvantages. In utero treatment could potentially be associated with improved healing as well.

Oral teratomas have been classified into 4 subgroups: (1) dermoids or hairy polyps that are the most common and contain only epidermal and mesodermal elements; (2) teratoids that consist of ecto-, meso-, and endodermal elements but that are incompletely organized; (3) true teratomas that contain all 3 germ layers but with greater histological organization and recognizable early organ differentiation; and (4) epignathus that are highly differentiated tumors with well-formed organs and limbs, which are rare and generally incompatible with life.18-21

Nasopharyngeal teratomas, as in our case, are associated with an exceptionally high risk of neonatal mortality, particularly from airway obstruction. Polyhydramnios, hydrops, and cleft palate may also occur. Other tumors, such as pure oral teratomas, have also been reported. Of 15 reported cases of pure oral teratomas, fetal death occurred in 2 cases, 1 patient terminated the pregnancy, 2 other patients had cleft palate, and 1 neonate had nerve palsy.22-30 Prior to the year 2000, this type of tumor was considered essentially incompatible with life.5 In fact, a review article in 2008 stated that only 8 cases existed in which the teratoma had been successfully excised.31

Perinatal management of oral teratomas includes delivery by cesarean section and either resection (OOPS16) or an EXIT15 procedure with or without tracheostomy. Chiu et al15 reported 2 cases with prenatal diagnosis of upper airway obstruction. One case was an epignathus successfully treated with an EXIT procedure. Postnatal management of oral teratomas involves surgical resection, which is often curative, without recurrences if the initial surgery is successful.32

Our case suggests that in utero fetoscopic management of fetuses with oral teratomas may be feasible in selected cases. In utero endoscopic assessment or treatment may allow parents to make a better educated decision about their management of the pregnancy. If done early enough, as in our case, fetoscopic removal of the teratoma may avoid further growth of the mass, distortion of the facial structures, micrognathia, development of polyhydramnios, airway obstruction, the need for an EXIT or OOPS procedure, and the need for cesarean section delivery. Further experience with in utero fetoscopic treatment may be difficult because of the rarity of the condition but deserves consideration.

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REFERENCES


Image of infant at birth, showing no residual tumor, no clefting, and no evidence of injury from the antenatal fetoscopic surgery.