Int. J. Oral Maxillofac. Surg. 2014; 43: 433–436
http://dx.doi.org/10.1016/j.ijom.2013.09.004, available online at http://www.sciencedirect.com

International Journal of Oral & Maxillofacial Surgery

Case Report
Cleft and Craniofacial Disorders

Emergency management of a congenital teratoma of the oral cavity at birth and three-year follow-up

S. Dakpé[1,*], B. Demeer[2,3], C. Cordonnier[4], B. Devauchelle[1]
[1]Department of Maxillofacial Surgery and Stomatology, University Hospital Amiens, France; [2]Department of Prenatal Diagnosis, Gynaecology and Obstetrics, University Hospital Amiens, France; [3]Department of Medical Genetics, Rare Disease Centre, University Hospital Amiens, France; [4]Department of Anatomopathology, University Hospital Amiens, France

S. Dakpé, B. Demeer, C. Cordonnier, B. Devauchelle: Emergency management of a congenital teratoma of the oral cavity at birth and three-year follow-up. Int. J. Oral Maxillofac. Surg. 2014; 43: 433–436.

Abstract. Teratomas are congenital malformations that are rarely located in the head and neck region. We report a case of congenital teratoma of the oral cavity, which was causing an airway obstruction and was treated at the time of birth. This teratoma was discovered at 27 gestational weeks by ultrasonography. A multidisciplinary team was consulted for antenatal diagnosis; the options of therapeutic abortion or management of the birth with the prevention of respiratory distress were debated. However, preterm labour at 32 gestational weeks accelerated the parental and the medical decisions. The parents agreed to the birth. The various disciplines coordinated their work, and the predefined treatment plan for clearing the airway obstruction was applied to manage the birth. The reestablishment of patency of the airway was performed during delivery and removal of the tumour was performed immediately afterwards. The follow-up of this case over 3 years is also presented.

Keywords: teratoma; antenatal diagnosis; EXIT procedure; cleft palate; neonatal surgery; epignathus.

Accepted for publication 3 September 2013
Available online 24 January 2014

From the Greek root 'tera', meaning monstrous, teratomas are congenital malformations that occur in one out of every 4000 births; they are rarely located in the head or neck (2%). The discovery of an obstructing teratoma of the upper aerodigestive tract during the antenatal period is a challenging clinical scenario. As a direct consequence of the rarity of teratomas of the head and neck, most clinicians have very limited experience with the management of these tumours.[1] The case presented here demonstrates that severe respiratory obstructions can be managed and resection should be performed without wasting any time. Furthermore, successful management of severe respiratory obstructions caused by these tumours requires the timely coordination of various disciplines, including maxillofacial surgeons, neonatalogists, anaesthetists, and high-risk obstetrics specialists at the time of the birth.

We report the successful emergency management of a congenital teratoma of the oral cavity in a premature infant. The paediatric development of this patient has remained normal to date.

Case report

Following a second trimester morphological foetal ultrasound examination, a 28-year-old woman, who was gravida 3 para 2, was referred to our medical centre for a suspicion of an oral tumour, which was confirmed. A large mass of 35 mm × 20 mm was detected, with coexisting cystic and solid tissues. The oral cavity appeared to be blocked by this mass. Subsequently, a hydramnios developed (Fig. 1A and B).

Amniocentesis was performed and revealed a normal 46, XX foetus. Foetal

0901-5027/040433 + 04 $36.00/0

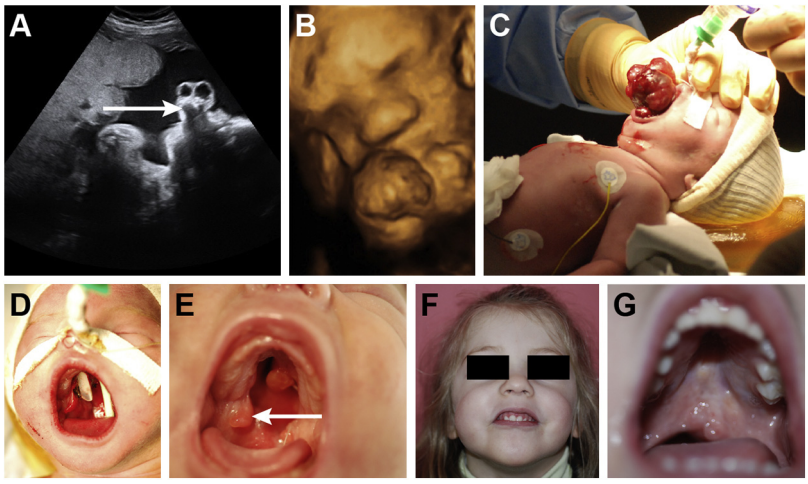

Fig. 1. (A) Ultrasound examination at gestational week 28 showing a large mass with coexisting cystic and solid tissues. The exteriorized mass measured 35 mm × 20 mm. Hydramnios was visualized (lateral view). (B) 3D ultrasound examination at gestational week 28 showing a large mass; the oral cavity appeared to be totally blocked by this mass. (C) Premature infant (32 gestational weeks) with a large teratoma blocking the oral cavity after nasofibroscopic intubation during the EXIT procedure. (D) Cleft palate and tumoural remnant located in the right oropharynx on day 1. Closure of the large cleft palate was performed at 19 months with a calvarial periosteal graft. (E) Cleft palate and tumoural remnant located in the right oropharynx on day 7. (F) Face of the patient at 36 months. (G) Endobuccal view of the patient at 36 months.

magnetic resonance imaging (MRI) was performed at 29 gestational weeks to attempt to accurately define the extent and nature of the tumour. The MRI showed a mass measuring 7 cm × 5 cm occupying the oropharyngeal area, although the limits of the mass with the hard palate could not be clearly visualized. The left maxillary bone was poorly defined, with a bulky aspect of soft tissue on the left hemiface. The epiglottis was well defined, whereas the upper aerodigestive tract above was not. The presence of a hydramnios suggested an upper airway obstruction of the upper aerodigestive tract.

Due to many uncertain issues arising from the difficulty to exactly define the extent of the mass, its nature, and the postnatal prognosis, several options were discussed with the antenatal multidisciplinary team consisting of obstetricians, paediatricians, anaesthetists, and maxillofacial surgeons. Finally the antenatal counselling team advised the parents of two possibilities: therapeutic abortion or continuing the pregnancy with close follow-up and an MRI before a planned delivery at 37 gestational weeks. For the latter possibility, the treatment procedures had to be discussed. Considering the diameter of the oral mass, a caesarean section would be scheduled to avoid the obstetric complication of pelvic dystocia. Owing to the presence of a suspected airway obstruction, a treatment plan for airway management would be put in place as follows. Firstly, on the day of the caesarean section, the multidisciplinary team, familiar with maternal high-risk caesarean sections, neonatal surgery and resuscitation, and foetal and maternal anaesthesia would be gathered in a designated operating theatre. Secondly, in the operating room, all the various equipments required needed to be prepared, keeping foetal instruments separate from maternal ones. Thirdly the caesarean section would be performed using an *ex utero* intrapartum (EXIT) procedure. After delivery but before the umbilical cord was clamped, the paediatric intensive care unit (ICU) planned to intubate the premature infant on placental support. If they failed, intubation with a fibre-optic flexible bronchoscope would be attempted. If this last option failed, a tracheotomy would be performed by the surgeons. If the tracheotomy became complicated, an attempt would be made to clear the airway of the obstruction by removing the mass. Afterwards, depending on the clinical birth data, the decision to whether or not remove the tumour immediately after birth in an adjoining operating room would be made.

Preterm labour ensued at 32 gestational weeks, which accelerated both the parental and the medical decision-making processes. The parents agreed to the birth of the infant as previously described. The different disciplines coordinated themselves and followed the defined procedure. The mother was transferred from the obstetric emergency department to a paediatric surgery theatre.

The caesarean section was initiated, followed by an EXIT procedure, and the treatment plan for the neonate was applied. During the hysterotomy, a 3 l hydramnios was evacuated. Once the foetus was extracted, a nasofibroscopic intubation was attempted. The intubation failed through the right nostril, but was successful through the left one (Fig. 1C). Indeed, during removal of the tumour, we discovered that it demonstrated a pedicle on the right side of the palate.

Once the airway was secured, the umbilical cord was cut. While the obstetricians performed the caesarean closure, the premature infant was entrusted to the neonatalogist for further examinations. It was decided to remove the protruding tumour immediately. The tumour was attached in the right nasopharynx area and had migrated into the cleft. The premature girl (2500 g, 43 cm) had macrostomia and a large cleft palate.

After removal of the tumour, the neonate was transferred to the neonatal ICU. A clinical examination of the oral cavity showed a residual ovoid mass of 1 cm × 0.5 cm remaining in the right oropharynx, in the area where the mass had been attached (Fig. 1D and E). She was extubated on day 2. At 7 days, bottle feeding was started. At 10 days, she was discharged from the neonatal ICU.

Histopathological analysis confirmed that the tumour was a mature teratoma. The mass weighed 61 g and measured 8 cm × 5 cm × 3.8 cm. The glial component was evaluated at 10%, and the immature component was evaluated at 1%. Different tissues were clearly identified as cutaneous tissue, tooth sketching, bronchial wall tissue, and bone tissue (Fig. 2A–F).

At 7 months, the infant underwent a further operation to remove the residual ovoid tumour mass. At 19 months, the large cleft palate was closed with a periosteal graft in a calvaria procedure.

To date, at 36 months old, this child has exhibited normal development (Fig. 1F and G). Her monitoring consists of regular paediatric and oral examinations and standard blood analyses for levels of alpha-fetoprotein (AFP).

Discussion

Foetus-in-foetu should be discussed as part of the differential diagnosis.[2,3] This condition was first described as a mass