CASE REPORTS

Perinatal management of a neonate with airway obstruction caused by rhabdomyosarcoma of the tongue

V. A. Skelton and A. Goodwin

Department of Anaesthesia, King’s College Hospital, Denmark Hill, London SE5 9RS, UK

Intra-oral masses in neonates can seriously compromise the airway, potentially causing hypoxia and death if not recognized and managed appropriately. We report a case in which an intra-oral mass was diagnosed on antenatal ultrasound scan. Preparation for delivery involved a multidisciplinary team approach, with a strategy for management at delivery. The child was delivered by elective Caesarean section and had a patent airway. A tracheostomy was performed immediately after delivery. The infant underwent a debulking procedure 3 weeks after birth. A histological diagnosis of embryonal rhabdomyosarcoma was made and a course of chemotherapy commenced. The child had a partial response to treatment with considerable shrinkage of the tongue mass. We discuss the management options in neonates with intra-oral masses to provide an adequate airway and maintain fetal oxygenation. The differential diagnosis of fetal oral masses is reviewed.

Br J Anaesth 1999; 83: 951-5

Keywords: neonates; airway, obstruction; complications, rhabdomyosarcoma

Accepted for publication: June 25, 1999

Advances in prenatal ultrasound examination have led to a considerable increase in the number of diagnoses of congenital abnormalities before birth. Before this advance, unexpected airway problems relating to head or neck masses in the neonate led to potentially life-threatening airway obstruction and hypoxia. We describe a case in which an intra-oral mass was diagnosed on antenatal ultrasound scan. We discuss the subsequent management for delivery, and review the options available.

Rhabdomyosarcoma (RMS) have a relative predilection for the head and neck regions, but involvement of the tongue is particularly rare. In the large series of the Intergroup Rhabdomyosarcoma Studies (IRS) I, II and III, only seven cases of RMS of the tongue were documented, with none in the neonatal period.1

Case report

A 29-yr-old, gravida 3, para 2 woman presented to her local hospital at 38 weeks’ gestation with possible ruptured membranes. Her pregnancy had been uneventful until that time. She was discharged after a normal cardiotocographic trace and vaginal examination, but was readmitted 24 h later because of worsening symptoms. She was started on antibiotics because of a tentative diagnosis of pyelonephritis. An ultrasound scan was performed to confirm fetal well-being which showed a mass measuring approximately 60×49×51 mm on the fetal tongue (Fig. 1). The mother was transferred to the fetal assessment unit at our hospital where a further scan confirmed the presence of a large mass at the base of the tongue. This was pushing the tongue forward and was considered to be potentially obstructing the airway. Amniotic fluid volume was normal. Lung development and tracheal diameter were also considered normal, indicating that if airway obstruction was present, it was not complete.

A multidisciplinary team was convened comprising obstetricians, neonatologists, paediatric anaesthetists and paediatric ENT surgeons and an action plan formed before delivery of the baby by elective Caesarean section. As the prenatal ultrasound indicated that airway obstruction was unlikely to be complete, the decision was made to deliver the baby completely, with division of the umbilical cord, before inspection of the airway by a paediatric anaesthetist using direct laryngoscopy. (The alternative was to perform laryngoscopy or tracheostomy with the placental circulation intact.) The paediatric ENT surgeons were to be ready to operate with all the equipment necessary to perform immediate tracheostomy if the airway was compromised. The neonatologists were to assist in resuscitation of the baby and provision of vascular access.

In the event, an otherwise healthy male was delivered
and cried immediately, indicating a patent airway, before the umbilical cord was clamped. The cord was subsequently divided and the baby transferred to the neonatologists with Apgar scores of 8 and 10 at 1 and 5 min. His tongue was hugely enlarged with a non-compressible, non-pulsatile soft tissue mass protruding from his mouth (Fig. 2a, b). It was felt that he may encounter respiratory difficulties in the neonatal period, especially if his tongue enlarged further, and the decision was made to perform a tracheostomy immediately. Intravenous and intra-arterial access were secured via the umbilical vessels, atropine 100 μg was given and laryngoscopy performed. Intubation proved easy, with a grade I view at laryngoscopy. Anaesthesia was induced with an oxygen-nitrous oxide-isoflurane mixture and a bolus dose of fentanyl. A tracheostomy was performed and a 3.5-mm paediatric Shiley tracheostomy tube inserted.

Postoperative cranial and abdominal ultrasound scans were normal and an MRI scan confirmed a heterogeneous soft tissue mass in the mid and anterior part of the tongue. Surgery to debulk the tongue was performed without complications 3 weeks later. Subsequent histology reported a well differentiated embryonal rhabdomyosarcoma (RMS) of keratomuscular origin. Further investigation defined the tumour as stage I with no spread, and a course of chemotherapy was commenced. After 8 months of chemotherapy there was a partial response to treatment with considerable shrinkage of the tongue mass. Complete excision of the tumour will be considered after further chemotherapy.

Discussion

Congenital masses of the head and neck in the neonate can seriously compromise the airway. A review of neonatal airway obstruction from teratomas reported 20% mortality, with most cases occurring before the era of prenatal ultrasound diagnosis. Increasingly refined ultrasound imaging can alert clinicians to the potential problems, and may give an indication of the degree of obstruction. Airway obstruction is more likely if an ultrasound diagnosis of a head or neck mass is associated with polyhydramnios, probably because of the inability of the fetus to swallow because of oesophageal and tracheal compression. Extension of the fetal head in utero may also indicate airway compromise, the mass limiting head flexion. In complete airway obstruction, caused mainly by laryngeal stenosis, prenatal ultrasound demonstrates a dilated trachea, large echogenic lungs caused by filling with water, flattened or inverted
diaphragms and ascites. This has been termed the congenital high airway obstruction (CHAOS) syndrome.5 6

Our case emphasizes the importance of a multidisciplinary approach to the management of these patients with a clear action plan for each team. The options available to us were:

1. Partial (head and shoulders) delivery of the baby, maintaining the fetal-placental circulation with uterine relaxation to prevent separation of the placenta, and performing direct laryngoscopy or tracheostomy to secure the airway before delivery of the baby’s torso.
2. Complete delivery of the baby, but maintaining

Fig 2 The neonate after tracheostomy to secure the airway. The large tongue tumour protrudes from the oral cavity.
uterine relaxation to prevent separation of the placenta and inspection of the airway before division of the umbilical cord.

(3) Complete delivery of the baby with division of the umbilical cord and subsequent laryngoscopy or immediate tracheostomy if the airway was compromised.

The first option was considered as fetal oxygenation via the placenta can be maintained while the airway is secured. It was rejected on the basis that prenatal ultrasound did not indicate complete airway obstruction or obstruction of the trachea. The method is not without hazards to the mother and performance of laryngoscopy or tracheostomy within the confines of the mother’s abdominal incision can prove technically difficult. The technique of placental support should be considered however when the antenatal ultrasound indicates complete airway obstruction or lesions that will impede surgical access for rapid tracheostomy, such as neck masses.

The second option of complete delivery of the baby, but maintaining the maternal–fetal circulation by uterine relaxation was similarly rejected. There is greater risk of premature placental separation with complete rather than partial delivery, and transfusion of blood from the fetus to the placenta may occur, as may excessive maternal blood loss. The fetus should be maintained level with the placenta after this type of delivery, but positioning may be limited by the length of the umbilical cord.7

The third option was considered safest for both the mother and baby. It was technically easier to access the airway with the baby fully delivered, and with the support of ENT surgeons skilled in performing emergency tracheostomy.

We were fortunate in this case that the baby cried on delivery before division of the umbilical cord, allowing time to secure the airway with the reassurance that oxygenation was maintained. The majority of the mass was in the anterior part of the tongue pulling it forward and probably assisting airway patency. The decision to perform an awake laryngoscopy was made on the basis that inhalation induction, the safer option for a potentially obstructed airway, would prove technically difficult because of the inability to hold a mask over the protruding tongue. I.v. induction was rejected because of the possible difficulty of maintaining the airway and oxygenation if intubation proved impossible. We were fortunate that intubation proved surprisingly easy.

The ex utero intrapartum treatment (the EXIT procedure),8 also termed the OOPS procedure (operation on placental support)9 was first described in animals10 but has since been used successfully to secure the airway of neonates with head and neck masses.7 11–14 It allows up to 60 min of cord perfusion before utero–placental gas exchange begins to deteriorate.15 During progress from the second to the third stage of labour, delivery of the fetus stimulates uterine contractions initiating placental separation. With the neonate’s first breath, systemic vascular resistance and pulmonary blood flow increase markedly, the ductus arteriosus closes and neonatal circulation begins. With the EXIT or OOPS procedure, Caesarean section is performed under general anaesthesia using a halogenated inhalation agent to relax the uterus. The fetus is either fully or partially delivered, but the maternal–fetal circulation is maintained without division of the umbilical cord until the airway has been secured. The uterus can be relaxed pharmacologically with either glyceryl trinitrate or ritodrine to prevent premature placental separation and thus maintain fetoplacental flow and oxygen delivery. Skarsgard and colleagues give a clear account of the procedure.9

The differential diagnosis of head and neck masses in the neonate includes cystic hygromas, haemangiomas, congenital goitres, choristomas, epignathus, neuroblastomas, granular cell tumours, retinoblastomas, mucoceles, cervical teratoma, branchial cleft cysts, laryngoceles, rhabdomyomas and rhabdomyosarcomas (RMS).13 A rare multigenic disorder, the Beckwith–Wiedemann syndrome, associated with dysregulation of the expression of genes involving growth and the cell cycle, can result in macroglossia associated with exomphalos, gigantism and hypoglycaemia.16–18 In this syndrome, polyhydramnios with accelerated growth between 25 and 36 weeks’ gestation is seen commonly on antenatal ultrasound scans, but fetal tongue protrusion may not be identified.19

RMS of the tongue in the neonate appears to be particularly rare. The intergroup rhabdomyosarcoma studies (IRS) I, II and III reported just seven cases of RMS of the tongue (0.34% of cases), with an age range of 2 months to 18 yr at diagnosis and all with the tumour at the base of the tongue.1 A study in Johannesburg over 25 yr identified eight cases of oral RMS, but none was of the tongue and the mean age of patients was 14 (range 7–46) yr. A literature review found 113 cases of RMS involving the oral and para-oral regions of which 16 involved the tongue.20 RMS can be categorized histologically into alveolar, embryonal and pleomorphic subtypes, with approximately 70% being embryonal. Histology in our patient identified RMS of the embryonal subtype. The prognosis of embryonal RMS is good; 89 children with localized RMS of orofacial and laryngopharyngeal sites had a 3-yr actuarial survival of 83%.21 Use of multidisciplinary therapy involving surgery, radiotherapy and chemotherapy has significantly improved long-term survival with better cosmetic and functional results. However, chemotherapy and radiotherapy in children may interfere with the development of the dentition or the jaws, resulting in microodontia, hypoplastic enamel, underdeveloped roots, radiation caries and micrognathia.20

Prenatal magnetic resonance imaging (MRI) can be useful to provide information on the anatomy of a mass in relation to the airway and may have been useful in our patient.15 Excellent images can be obtained with fast scanning techniques which combat motion artefact from the fetus moving, and minimize the necessity of sedating the mother. For slower scans, which require a stationary patient, either
maternal sedation or direct injection of neuromuscular blocking agents to the fetus have been used safely.7 12 MRI scans also provide more accurate diagnoses of the aetiology of fetal masses than ultrasound.7 15 22 23 24

Our report emphasizes the importance of a multidisciplinary team approach to the delivery of a neonate with suspected airway obstruction, and careful consideration of the options available to maintain oxygenation while securing the airway.

References
11 Langer JC, Tabb T, Thompson P, Paes BA, Caco CC. Management of prenatally diagnosed tracheal obstruction: access to the airway in utero prior to delivery. Fetal Diagn Ther 1992; 7: 12–6