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Case report

Perinatal airway management of neonatal cervical teratomas

Kiarash Taghavi [a], Robert G. Berkowitz [b,c,d,*], A. Michelle Fink [e,f,g], Ross D. Farhadieh [h], Anthony J. Penington [i,j,k]

[a] Royal Children's Hospital, Flemington Road, Parkville 3052, Melbourne, Australia
[b] Department of Otolaryngology, Royal Children's Hospital, Melbourne, Australia
[c] Murdoch Children's Research Institute, University of Melbourne, Melbourne, Australia
[d] Australian School of Advanced Medicine, Macquarie University, Sydney, Australia
[e] Department of Medical Imaging, The Royal Children's Hospital, Melbourne, Australia
[f] Fetal Medicine Unit, The Royal Women's Hospital, Melbourne, Australia
[g] University of Melbourne, Department of Radiology, Melbourne, Australia
[h] Royal Melbourne Hospital, Grattan Street, Parkville 3050, Melbourne, Australia
[i] Department of Plastic and Maxillofacial Surgery, Royal Children's Hospital, Flemington Road, Parkville 3052, Melbourne, Australia
[j] Department of Surgery, St. Vincent's Hospital, Melbourne, Australia
[k] University of Melbourne, Department of Surgery, Melbourne, Australia

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ABSTRACT

Cervical teratomas are rare but life-threatening neonatal tumors and management of the fetus with a cervical teratoma that threatens the airway remains a clinical challenge. This has been revolutionized by advances in fetal imaging and management of the airway at delivery including the use of Ex-utero Intrapartum Treatments (EXIT procedures). We present a retrospective case series of three neonates managed over a 12-month period. Following pre-natal fetal MRI and a multi-disciplinary management approach, two newborns were managed by prompt post-natal endotracheal intubation while an EXIT procedure was required in one. All three underwent surgical resection in the first few days of life. A decision regarding the best means by which to manage the airway in fetal cervical teratoma requires fetal MRI and a multi-disciplinary team approach to determine whether EXIT, or a safer approach from a maternal perspective can be employed. We also recommend routine endotracheal intubation at birth, due to the risk of spontaneous intra-tumoral hemorrhage. The need for surgery should be planned early, as rapid growth of the tumor can threaten the viability of the overlying skin and surrounding structures.

1. Introduction

Teratomas, etymologically 'monstrous tumors', are rare congenital tumors derived from pluripotent cells from all three primordial germ-cell layers. As such, they involve at least one-type of tissue originating from these three embryonic layers, each with varying degrees of differentiation. They also can include non-germ totipotential embryonic cells [1]. They are usually benign; however there have been very rare cases of malignancy or malignant transformation [2–4]. These can be from areas of germ cell or non-germinal malignant tumor patterns [5].

Teratomas can occur in almost any part of the body [6]. However, they are most commonly located in the sacrococcygeal region, followed by the ovaries, testis, anterior mediastinum, retroperitoneum, and finally the head and neck which account for less than 5% of the total [7]. Cervical teratomas have a nearly 100% mortality due to airway complications if not pro-actively managed, due in part to enlargement of the lesion in the first few days of life [8–10]. Poor prognostic indicators on pre-natal sonography include: large size (>5 cm), polyhydramnios and hydrops fetalis [6,9,11,12]. Management starts at the time of fetal diagnosis and involves a multi-disciplinary team including: obstetricians, sonologists, pediatric radiologists, neonatologists and pediatric surgery and otolaryngology teams. Ex-utero Intrapartum Treatments (EXIT procedures) are employed in the delivery of fetuses with large head and neck masses to obtain a stable airway while the newborn is maintained on utero-placental circulation [13]. The availability of fetal MRI allows for a more accurate assessment of the airway prior to birth, and therefore more fastidious use of intrapartum treatments.

In light of our recent experience with establishment of a multi-disciplinary fetal airway management team, we performed a retrospective review of newborns with cervical teratomas at our institution.

* Corresponding author at:.
E-mail address: Robert.Berkowitz@rch.org.au (R.G. Berkowitz).

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2. Methods

After gaining ethical approval from The Royal Children's Hospital Human Research Ethics Committee a retrospective analysis was performed by searching the theatre audit database for keyword "teratoma". We present the three cases of cervical teratoma that were identified over the preceding twelve-month period.

3. Case presentation

3.1. Case 1

A baby girl from a 22-year-old primigravida mother who was heterozygous for hemochromatosis (carrier phenotype) was found to have a complex cervical mass that was initially detected at the 20-week antenatal ultrasound scan, measuring 45 mm × 30 mm × 34 mm. At 25 weeks the mass measured 52 mm × 43 mm × 45 mm with no evidence of obstructive polyhydramnios. Fetal MRI confirmed a displaced but non-obstructed airway. At 36 weeks the child was delivered via elective cesarean section with neonatal, anesthetic and otolaryngology teams in attendance. Apgar scores were 8^{1} and 8^{5}. At approximately 5 min the neonate underwent semi-elective endotracheal intubation due to desaturations. The vocal folds were easily viewed and flexible bronchoscopy revealed normal tracheal anatomy.

A gradual increase in the size of the lesion was noted with compromise of overlying skin (Fig. 1). Resection was performed on day two post-delivery through a laterally placed elliptical incision. This allowed the mass to be easily shelled out from under the angle of the mandible. The cystic stalk was followed medially to the laryngeal cartilages. A large feeding vessel from the external carotid was divided and the ansa cervicalis was sacrificed. The hypoglossal nerve was identified and preserved.

Histological examination showed a benign teratoma measuring 110 mm × 65 mm × 65 mm. On day six after birth the neonate was successfully extubated. At three weeks of age the baby was still not taking oral feeds well. Video fluoroscopy showed an uncoordinated suck and swallow. On discharge, the baby remained on full nasogastric feeds that were weaned under speech therapy supervision over the succeeding months. It was felt that considerable nerve stretching had caused a transient neuropraxia.

3.2. Case 2

A baby girl was born from a 42 year-old primigravida mother with an unremarkable antenatal history. A cervical mass was initially visualized on ultrasound at 32 weeks causing the neck to be held in constant hyperextension. There was evidence of marked

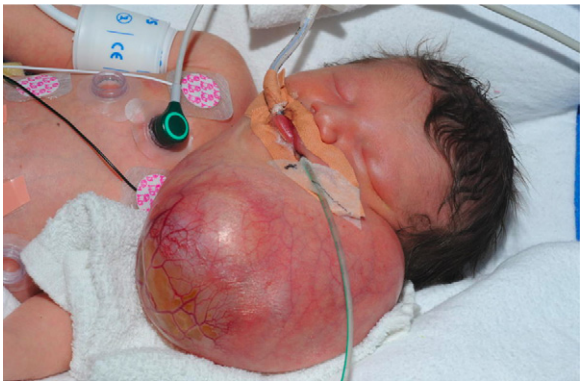

Fig. 1. Case 1: Pre-operative clinical photograph.

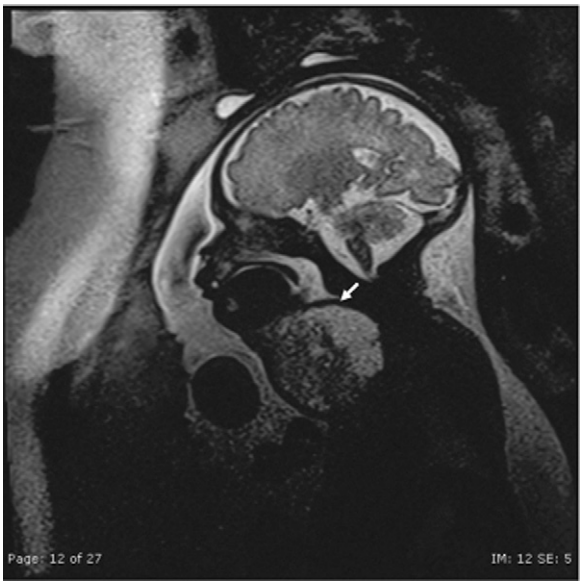

Fig. 2. Case 2: Fetal MRI at 37 weeks; sagittal T2 weighted image demonstrates the ballooning of the amniotic fluid filled pharynx, and its distal obstruction by the mass (arrow).

polyhydramnios, suggestive of esophageal compression. Fetal MRI at 36 weeks showed an enlarging mixed solid and cystic mass measuring 61 mm × 55 mm × 68 mm in keeping with a cervical teratoma. The oropharynx and nasopharynx were patent with ballooning of the laryngopharynx (Fig. 2). The esophagus and airway were extrinsically compressed at the level of the glottis. No fluid could be demonstrated below this level. A repeat fetal MRI scan at 37 weeks was performed for further delineation of the distal airway. High-resolution sagittal imaging showed occlusion of the subglottic trachea over a length of 2 cm, with a patent distal trachea. Fetal lung volumes were normal. The decision to deliver by EXIT was based on this scan. At 39 weeks the child was delivered and an airway was secured on placental support. Apgar scores were 3^{1}, 3^{5} and 7^{10}.

Two days after birth, following closely observed enlargement of the lesion, the teratoma was enucleated via a transverse neck incision. It was identified deep to the infra-hyoid strap muscles, with the trachea and esophagus forced to the contralateral side and carotid sheath displaced laterally. The vagus nerve was identified and preserved.

Histological examination was consistent with a cervical teratoma measuring 70 mm × 70 mm × 40 mm. At 12 days of age the neonate was returned to theatre for elective extubation and bronchoscopy. Edema at the level of the anterior commissure was identified, but this resolved and no further respiratory complications developed. Full enteral feeds were achieved after 12 days post-operatively and the neonate was discharged at 17 days of age with no post-operative problems.

3.3. Case 3

A 39 year-old primigravida mother with an IVF pregnancy had shown a normal female fetus on six previous antenatal ultrasounds. A 36-week prenatal ultrasound showed a heterogeneous neck mass with polyhydramnios present. Fetal MRI at 37 weeks showed extrinsic compression of the glottis and subglottic airway that was deviated and significantly narrowed with a short segment of probable total occlusion (Fig. 3). The tumor extended anteriorly in the neck to overlie the trachea right down to the thoracic inlet. After extensive counseling the mother refused an EXIT procedure on the basis of maternal risks.