CASE REPORT

Intraoral teratoma in a newborn presenting as severe respiratory distress

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SUMMARY

A new born female presented to us immediately after birth with mass protruding from the mouth, with severe respiratory distress and was immediately intubated. Antenatal ultrasonography did not reveal any abnormality. On examination, a non-pulsatile, multilobulated, 4 cm×5 cm sized mass of variable consistency was seen originating from the palate with no other associated congenital anomaly. CT showed a large heterogeneous mass arising from the hard palate with few hyperdense calcific areas with no intracranial extension. Mass was excised completely. Histopathological examination showed features of mature teratoma.

BACKGROUND

Neonatal oral teratomas are benign lesions with very high chances of respiratory tract obstruction with risk of asphyxiation and death without life-saving endotracheal intubation or tracheostomy or an EXIT (ex utero intrapartum technique) procedure.

With a planned multidisciplinary approach, a potentially fatal emergent airway obstruction can be converted into controlled emergent situation and treated successfully even in less resourceful developing countries.

CASE PRESENTATION

A full-term vaginally delivered 2 kg female baby presented immediately after birth with mass protruding from the oral cavity. The baby had severe respiratory distress. The patient was immediately intubated and a tracheostomy was not needed (figure 1). Baby was born out of non-consanguineous marriage with no history of any congenital malformation in the family. Antenatal ultrasonography did not detect any abnormality. On intraoral examination, mass was originating from upper palate, 4×5 cm in size, multilobulated, non-pulsatile, variable in consistency with firm and cystic areas on palpation (figure 2). No other congenital abnormality was detected. The mass was excised surgically with intraoral endotracheal anaesthesia. Postoperative period was uneventful. On gross examination mass showed multiloculated cysts with serous fluid and solid areas. The histopathological examination showed features of benign teratoma, tissue from all the three germ cell layers was seen, stratified keratinising squamous epithelium with dermal appendages, adipose tissue, cartilage and skeletal muscle was seen. No immature component was seen (figures 3–5).

Figure 1 Intraoral teratoma in neonate with severe respiratory distress requiring immediate endotracheal intubation.

INVESTIGATIONS

Haematological and biochemical investigations were normal. The serum α -fetoprotein level was 1400 IU. CT revealed a large heterogeneous mass arising from hard palate. The mass had few hyperdense calcific areas and fluid-dense cystic areas, with no intracranial extension. The chest x-ray was normal.

Figure 2 Resected gross specimen.

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Figure 3 Microscopic picture showing skin appendages in the resected specimen.

TREATMENT

Management of oropharyngeal teratomas requires careful planning and team effort by highly dedicated obstetrician, neonatologist and pediatric anaesthesiologist.¹⁻³ There is little hope of survival without multidisciplinary approach if the teratoma is compromising the airway. In such cases immediate tracheostomy or intubation can be life saving measure if proper antenatal diagnosis is made.⁴ In advanced centers, techniques such as EXIT procedure is employed. This technique consists of securing the airway of the fetus by endotracheal intubation, partially delivered, that is, head and thorax delivered out and placenta still connected to the fetus.⁵ Fetal airway is well secured by this manouver maintaining the oxygen saturation.⁶ Once the airway is maintained by endotracheal intubation or tracheostomy, the cord is clamped. The mainstay of treatment after securing the airway is complete surgical removal followed by adjuvant therapy. If the mass is attached by thin stalk, it can be excised under local anaesthesia.

Figure 4 Microscopic picture showing glial tissue in the resected specimen.

Figure 5 Microscopic picture showing glandular epithelium in the resected specimen.

OUTCOME AND FOLLOW-UP

The histopathological findings in our patient were in favour of benign teratoma. The patient had a regular follow-ups with serum levels of α fetoprotein and human chorionic gonadotropin within normal limits. No recurrence was detected after 12 months.

DISCUSSION

The term 'teratoma' was first coined by Virchow in 1869 for monstrous lesions which contained a foreign tissue to the part in which they arise.⁷ The exact cause is not known but teratomas are thought to arise from pluripotent cells and contain poorly organised tissue derived from all the three embryonic germ cell layers. These pluripotent cells sometimes miss the normal path of migration from allantois to genital ridge during fourth and fifth weeks of gestation and give rise to teratoma. Some researchers consider it to be a remnant of the primitive streak.

The incidence of teratomas is 1 in 4000 live-births with predominance in females.⁸ The most common site of teratoma is sacrococcygeal region, followed by gonads, retroperitoneum.⁹ Head and neck teratomas constitutes 1-9% of all teratomas.¹⁰ Oropharyngeal teratomas are extremely rare tumours and represent 2% of all teratomas. These tumours commonly arise from palate followed by pharyngeal wall. Cheek is the least common site of teratomas.¹¹ Teratomas arising from soft or hard palate, cheek, buccal mucosa, tongue and protruding from mouth are called epignathus. These teratomas are mostly benign but have a high death rate due to respiratory tract obstruction and feeding difficulties. Large teratomas can interfere with fetal swallowing and produce polyhydramnios, cause severe respiratory distress at birth, and may lead to stillbirth.¹² The clinical presentation includes dyspnoea, suffocation and difficulty in swallowing. Oropharyngeal teratomas have a higher incidence of preterm birth and require emergent airway management after birth.¹³

Oropharyngeal teratomas are also associated with midline anomalies like tongue hamartoma, choristoma and encephalocele.¹⁴ Contrast-enhanced CT is useful in defining the origin of the tumour and determining the exact anatomic extent of the tumour. Calcifications along with the heterogeneous nature of the mass are typical of teratomas. Differential diagnosis includes haemangioma, rhabdomyosarcoma, encephalocele, gliomas and von Recklinghausen's disease.

Learning points

- ▶ Normal antenatal ultrasonography does not rule out oropharyngeal teratomas in many instances.
- ▶ Most of oropharyngeal teratomas are benign.
- ▶ There are very high chances of respiratory tract obstruction with risk of asphyxiation in oropharyngeal teratomas.
- ▶ With a planned multidisciplinary approach, a potentially emergent airway obstruction can be converted into controlled emergent situation.
- ▶ Even in developing countries without facilities of advanced techniques like EXIT (ex utero intrapartum technique) procedure, such cases can be successfully managed with a multidisciplinary approach.

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