Non-operating room anaesthesia for residual neonatal epignathus: *Small pill, big thrill...*

Sir,

Epignathus is a rare congenital oropharyngeal teratoma with incidence of 1:35000-2,00,000 live births (female >male babies).[1] It is most commonly thought to arise from the pluripotent cells of Rathke's pouch.[2] Delineating the size, origin and extent of tumour during antenatal scanning aids in formulating a definitive airway plan and circumventing fatal airway compromise at birth.

A 17-day-old male neonate, antenatally diagnosed with nasopalatine cyst at 20 weeks of gestation was planned for contrast enhanced computed tomography (CECT) of neck. The baby was born by caesarean section at term, weighed 3.1 kg and had no facial dysmorphism. Immediately after birth, the neonate underwent coblation assisted partial palatal cyst excision under general anaesthesia, histopathology of which revealed mature cystic teratoma. After pre-oxygenation with 100% oxygen using #3 facemask at 10 L/min, inhalational induction with incremental sevoflurane (2-6%) using nasally inserted 2.5 mm endotracheal tube was done while maintaining spontaneous ventilation. On finding the tongue to be free on left side, direct laryngoscopy using Miller blade was attempted using left paraglossal approach and trachea intubated with 3 mm uncuffed tube. Muscle relaxation was instituted with atracurium after confirming capnographic waveform. Anaesthesia was maintained using 3% sevoflurane in air-oxygen (50:50) and the baby was extubated after six days of mechanical ventilation.

On presentation to us on day-17 of life for CECT neck [Figure 1a and b], examination of oral cavity revealed a residual cystic lesion (approximately 5 × 4 × 5 cm) pushing the tongue backwards along with cleft palate with no clinical signs of upper airway obstruction. After obtaining written informed parental consent, child was premedicated with glycopyrrolate (5 μg/kg IV) and ketamine (1 mg/kg IV). Standard American Society of Anesthesiologists (ASA) monitors were attached and right nostril prepared with one drop 0.05% xylometazoline. 2.5 mm uncuffed endotracheal tube was inserted nasally and used for maintaining airway patency [Figure 2]. Inhalational induction using incremental sevoflurane (2-6%) with air-oxygen (50:50) was performed. Anaesthesia was maintained with 2% sevoflurane in air-oxygen (50:50) on spontaneous ventilation with Jackson Rees circuit attached to Penlon Prima SP2 anaesthesia system. C-MAC® videolaryngoscope (Karl Storz, Tuttlingen, Germany) and 8Fr/35 cm Frova intubating introducer were kept standby for securing airway as backup measure. No peri-procedure respiratory adverse event (stridor, laryngospasm, bronchospasm) occurred during the scan and the baby was shifted to neonatal intensive care unit.

Cleft palate is the most commonly associated malformation with epignathus as the mass obstructs palatal closure in foetal life.[2] Other associated anomalies include bifid tongue and/or nose, glossoptosis and resulting functional mandibular micro-retrognathia due to impaired foetal jaw growth. Infrequently, epignathus may have an intracranial extension which portends a poor prognosis.[2] Polyhydramnios is characteristically seen in nasopharyngeal, oropharyngeal and large cervical teratomas, which hinder foetal swallowing. Nasopharyngeal and large cervical teratomas are particularly ominous due to their propensity to cause fatal airway compromise. Pure oral teratomas on the other hand, although frequently missed on prenatal scans are extremely rare, with tongue being most commonly involved and usually present with feeding problems.[3] Generally devoid of critical respiratory presentations, they tend to grow outwards away from the oral cavity, which explains the success of paraglossal direct laryngoscopic approach for intubation during the initial tumour resection. However, possibility of fatal airway

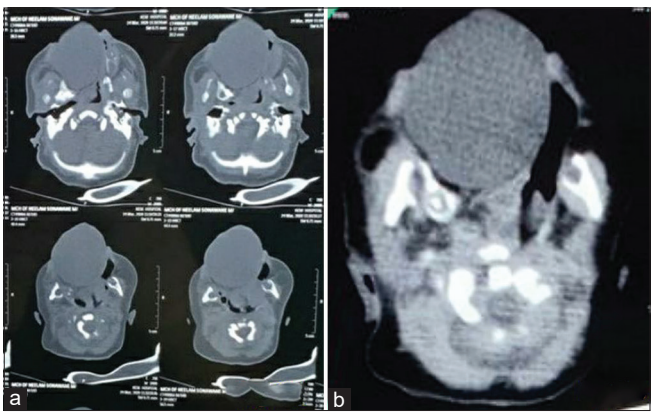

Figure 1: (a) Epignathus post partial excision (b) Nasally inserted ETT delivering the anaesthetic mixture

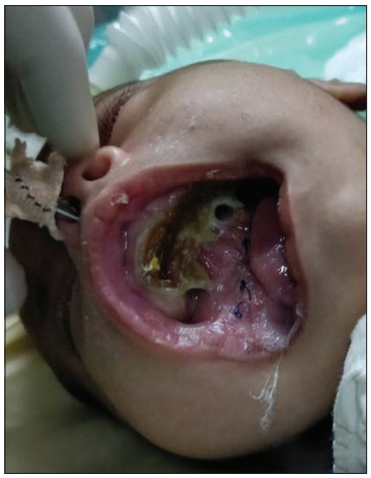

Figure 2: Well-defined hypodense cystic lesion arising from the palate and pushing the tongue postero-inferiorly with airway compromise

compromise at birth still exists and remains a formidable anaesthetic challenge particularly with nasopharyngeal and large cervical teratomas. In life threatening cases, Ex utero intrapartum treatment (EXIT) procedure allows securing the airway with a tracheostomy or partial cyst excision followed by oral or nasal endotracheal intubation in the presence of intact foetoplacental circulation without foetal oxygenation compromise.[4]

In our case, the child had a residual lesion along with cleft palate and was posted for non-operating room anaesthetic procedure, all of which added to the complexity of the case. We successfully maintained spontaneous ventilation with endotracheal tube modified as a nasopharyngeal airway. The use of nasopharyngeal airway in the setting of paediatric difficult airway remains a reliable and trusted technique of ensuring airway patency, oxygenation and delivery of anaesthetic agent.[5]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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