Missed nasopharyngeal teratoma: A cause for recurrent respiratory distress in a neonate

INTRODUCTION

Teratomas are congenital tumours with incidence of 1:40,000 live births.[1] Nasopharyngeal teratomas (NPT) are the second most common among the head and neck teratomas. Symptoms of NPT are related to both size and location. Large ones cause airway obstruction and feeding difficulties, while small ones can cause intermittent symptoms resulting from the ball valve effect of the obstruction.[2] This case report stresses the importance of recognising NPT as an important

differential diagnosis for recurrent upper airway obstruction in a neonate. Excision of the lesion was the treatment of choice to relieve the airway obstruction.

CASE REPORT

Term female baby weighing 2.47 kg was born by a normal vaginal delivery to a primigravida. Left submandibular multi lobular cystic swelling suggestive of cystic hygroma was diagnosed in an antenatal ultrasound scan at 24 weeks. A repeat scan at 34 weeks showed a size of 3.5 cm × 2.5 cm × 2.4 cm of the same.

At birth, the baby had an Apgar score of 8/10 and was tachypneic with mild respiratory distress. Saturation was maintained with supplemental oxygen. No craniofacial or other congenital anomalies were seen. Baby was shifted to neonatal intensive care unit (NICU) for observation. She was intubated orally and mechanically ventilated within 6 h due to increasing respiratory distress. Mild trauma was suspected at the insertion of a nasogastric tube in the right nasal cavity, which otherwise was easily passed. After 40 h of mechanical ventilation, she was clinically stable and therefore extubated. Within 4 h, she deteriorated requiring reintubation and ventilation. An ultrasound scan showed a cystic hygroma of the left side of the face and neck with mild displacement of cervical vessels laterally, but no extraluminal compression on the trachea. Paediatric surgeons suggested conservative management at that stage, without emergency surgery for the cystic hygroma. Chest X-ray did not reveal any signs of aspiration or pneumonia.

Repeated trials of tracheal extubation failed, the search was now focussed on other airway abnormalities causing recurrent respiratory distress such as subglottic stenosis, laryngotracheomalacia, lymphangioma, haemangioma, cysts, or teratomas. A rigid bronchoscopy was performed in operating room (OR) which revealed no airway abnormality and reconfirmed no extraluminal compression of the trachea. The child was operated upon for cystic hygroma, after the bronchoscopy, and was transferred back to NICU for the ventilator support. In the post-operative period, during suctioning the oral cavity, small tuft of hair was retrieved and a closer laryngoscopic examination revealed a mass in the nasopharynx. A computed tomography (CT) scan obtained at this stage showed a pedunculated polypoidal mass (2.6 cm × 1.4 cm size) with foci of calcification, from posterior aspect of bony nasal septum and roof of nasopharynx extending into oropharynx obliterating the nasopharyngeal cavity on the right side suggesting NPT [Figures 1 and 2].

Surgical excision of the NPT was now planned by a team consisting of anaesthesiologists, neonatologists, paediatric, and ENT surgeons. The baby was transferred to OR, tube *in situ* and on continued ventilation, and anaesthesia was initiated with nitrous oxide – oxygen - isoflurane, fentanyl, and atracurium. The mass was successfully removed by nasoendoscopic and transoral resection. No intracranial extension or involvement of adjacent structures was noted.

The baby was managed in NICU and weaned off the mechanical ventilation in the next 2 days. Histopathological examination of the neck mass showed features of a cystic teratoma and that of

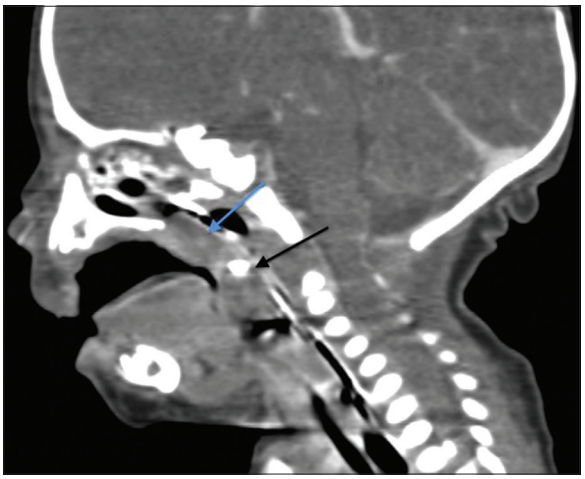

Figure 1: Sagittal reconstruction of a post-contrast computed tomography, which shows a lesion in the posterior wall of oropharynx (blue arrow) with focus of calcification (black arrow) causing significant compression of the airway

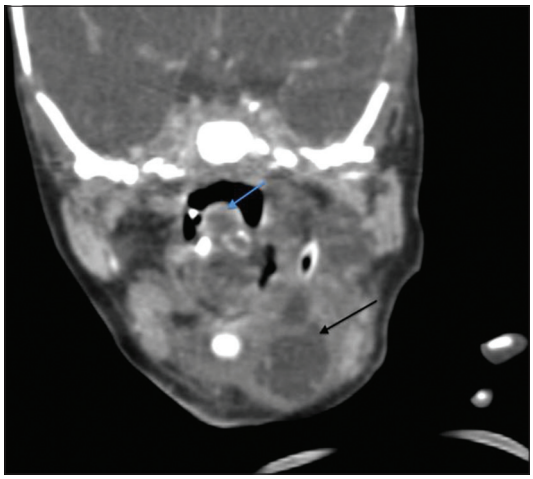

Figure 2: Coronal reformatted image of a post-contrast computed tomography showing mass in the oropharynx with calcification (blue arrow) and the residual cystic hygroma (black arrow) in the left side of the neck

excised polypoidal mass revealed a mature teratoma consisting of all three germ layers (skin, bone, cartilage, tooth buds, adipose tissue, neural, and vascular structures).

DISCUSSION

Head and neck teratomas account for 5% of all neonatal teratomas. These neoplasms may occur in brain, orbit, oropharynx, nasopharynx, or cervical region.[3] NPT is rare in neonates and has female predominance. It consists of tissues arising from all three embryonic germ layers - ectoderm, mesoderm, and endoderm.[4] It may be associated with other anomalies such as cleft palate, microcephaly, atresia of left common carotid, and cardiac abnormalities.[5] The differential diagnosis of the nasopharygeal mass includes lymphatic malformation, congenital rhabdomyosarcoma, haemangioma, intranasal glioma, meningomyelocoele, and encephalocoele.

Diagnosis of NPT begins in utero. Maternal clinical characteristics include polyhydramnios due to impaired swallowing and elevated alpha fetoprotein.[6] Airway obstruction and respiratory distress related to the size and location of these teratomas is the major cause of morbidity and mortality. Extrauterine intrapartum treatment procedure continues to be the optimal delivery strategy for patients with prenatally diagnosed giant cervical teratoma.[6] Life-threatening airway obstruction may occur at birth and require urgent resuscitation including bronchoscopy assisted intubation or tracheostomy.[7,8] Postnatally, imaging techniques such as CT/magnetic resonance imaging (MRI) are confirmatory and aid in diagnosis of the involvement of adjacent vascular, bony, visceral structures, or any intracranial extension.[6]

In the present case, a nasogastric tube could be passed in the right nasal cavity. NPT was not visualised during laryngoscopy, intubation and bronchoscopy precluding the diagnosis. This was probably due to the pedunculated mass slipping back into the upper nasopharynx in the above procedures.[1] The antenatal diagnosis of cystic hygroma may have diverted attention from other causes of respiratory distress in the neonatal period. Among cases detected prenatally, cystic hygroma is the most likely entity to be mistaken for cervical teratoma. Similarities in size, location, clinical characteristics and sonographic finding can make this distinction difficult.[3] Therefore, a high degree of suspicion is necessary during the evaluation of the cause of recurrent upper airway obstruction in the neonate and rare lesions such as teratoma should not be overlooked.[9] Bronchoscopy should be aimed at both upper and lower airway evaluation. CT/MRI play a key role in differentiating NPT from other causes of a neonatal neck mass thus optimising the management, which was surgical excision in this case.[10]

CONCLUSION

Nasopharyngeal teratomas form a rare clinical entity and may be associated with airway obstruction in the postnatal period. It may be missed as a cause for respiratory distress, when other associated craniofacial anomalies are also present. A thorough clinical examination, bronchoscopy and imaging (CT/MRI) aid in the diagnosis. Early resection is the most effective method to control the airway. Airway management of these cases require a multidisciplinary team comprising neonatologist, otolaryngologist, paediatric surgeon, and anaesthesiologist.

M Manjuladevi, Kshma A Kilpadi, Jiby Jose, Apoorwa Kothari

Department of Anesthesia and Critical Care, St. John's Medical College and Hospital, Bengaluru, Karnataka, India

Address for correspondence:
Dr. M Manjuladevi,
Department of Anesthesia and Critical Care,
St. John's Medical College and Hospital, John Nagar,
Bengaluru, Karnataka, India.
E-mail: drmanjula95@yahoo.com

REFERENCES

1. Tiwari L, Baijal N, Puliyel JM. Nasopharyngeal teratoma as a cause of neonatal stridor. Indian Pediatr 2009;46:1097-8.
2. Hermansen CL, Lorah KN. Respiratory distress in the newborn. Am Fam Physician 2007;76:987-94.
3. Azizkhan RG. Teratoma and other germ cell tumors. In: Grosfeld JL, editor. Pediatric Surgery. 6th ed. Philadelphia: Mosby Elsevier; 2006. p. 554-74.
4. Ulger Z, Egemen A, Karapinar B, Veral A, Apaydin F. A very rare cause of recurrent apnea: Congenital nasopharyngeal teratoma. Turk J Pediatr 2005;47:266-9.
5. Isken T, Alagöz MS, Günlemez A, Unal C, Sen C, Onyedi M, *et al.* A congenital true teratoma with cleft lip, palate, and columellar sinus. J Craniofac Surg 2007;18:1083-5.
6. Elluru RG, Wootten CT. Congenital malformation of nose. In: Flint PW, editor. Cummings Otolaryngology Head and Neck Surgery. 5th ed. Philadelphia: Mosby Elsevier; 2010. p. 2686-96.
7. Azizkhan RG, Haase GM, Applebaum H, Dillon PW, Coran AG, King PA, *et al.* Diagnosis, management, and outcome of cervicofacial teratomas in neonates: A Childrens Cancer Group study. J Pediatr Surg 1995;30:312-6.
8. Atasay B, Arsan S, Okulu E, Akin IM, Yagmurlu A, Turmen T. Hand intubation of a tiny neonate with a large obstructive mass in the oral cavity. Paediatr Anaesth 2009;19:277-8.
9. Rangachari V, Aggarwal R, Jain A, Kapoor MC. Neonatal

airway lesions: Our experience and a review of the literature. J Laryngol Otol 2013;127:80-3.
10. Mirshemirani A, Khaleghnejad A, Mohajerzadeh L, Samsami M, Hasas-Yeganeh S. Congenital nasopharyngeal teratoma in a neonate. Iran J Pediatr 2011;21:249-52.

Access this article online	
Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/0019-5049.135079