Beyond stertor: A rare case report on nasopharyngeal mature teratoma in an infant

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SUMMARY

Teratomas are the most frequent congenital tumours in children, originating from two or three germ layers. Since the sacrococcygeal region is the most prevalent site for this tumour, the nasopharyngeal region is uncommon in its localisation. Teratomas are generally benign and have a well-recognised clinical and histopathological entity. This case report describes a 4-month-old boy who had stertor since day 10 of life and presented to the emergency department with worsening noisy breathing and rapid breathing for two weeks prior to the admission. During the admission, he experienced many desaturation episodes necessitating non-invasive ventilation. Imaging supported the endoscopy's discovery of a solitary cystic lesion obstructing the left posterior choanae. The patient underwent a successful surgical excision of the mass, and a histological examination revealed a mature teratoma free of any indications of cancer. He had a smooth recovery following surgery, and regular check-ups revealed no signs of a tumour recurrence. This report addresses the need for a thorough assessment and evaluation of the stertor to rule out additional serious upper airway issues, such as the one in this instance. As a result, we can prevent severe morbidity and mortality.

INTRODUCTION

Teratomas represent one of the most common extragonadal germ cell tumours of childhood. It is a benign tumour composed of cells from ectodermal, mesodermal and endodermal layers.[1] They can be found in any body site, but most are in a midline or paraxial location from the brain to the sacral area. The most common locations for teratomas are the sacrococcygeal site, anterior mediastinum, ovaries, testicles and retroperitoneal area. The head and neck are rarely involved, 2 to 9% of all teratomas, and the incidence is only one in every 40,000 births.[1,2] Although nasopharyngeal teratomas are rare, they can lead to severe airway obstruction. The presentation will be stertor, respiratory distress, feeding problems and cosmetic deformity. Neonates who present with stertor, a low-pitched breathing sound, should be alerted to the possibility of obstruction along the nasal cavity, nasopharynx or oral cavity. Congenital mass obstruction in those mentioned areas should be considered. Stertor in neonates signifies a critical airway blockage demanding urgent attention from clinicians familiar with the intricacies of neonatal physiology. Managing this condition requires a multidisciplinary approach, emphasising effective communication among healthcare providers and caregivers. This collaborative effort is paramount for ensuring favourable outcomes for these vulnerable infants.

CASE PRESENTATION

A 4-month-old boy was brought to the emergency department (ED) with worsening noisy breathing and rapid breathing. The noisy breathing had been presented since he was 10 days old but was not associated with rapid breathing or issues with feeding and sleep. However, over the past 2 weeks, his breathing has worsened, becoming more rapid and disturbing his sleep. He has no fever, cough, runny nose, bluish discolouration or seizure. He also has no previous history of hospital admission. Antenatal history was uneventful. He was born full-term with a birth weight of 3 kg and good Apgar scores. During the neonatal period, he has no other issues and has been followed up regularly at the nearest health clinic. His growth and development have been appropriate for his age, and his immunisation is up to age.

Upon examination on the day of admission, he had a low-pitched inspiratory stertor and was in respiratory distress, as evidenced by the tachypnoeic and subcostal recession. Lung auscultation was equal, and air entry was clear. Other systemic examinations were unremarkable.

He was referred to the paediatrics team and admitted to the intensive care unit, where he required continuous positive airway pressure (CPAP) with positive end-expiratory pressure (PEEP) of six due to three episodes of oxygen desaturation down to 85% while on room air. Despite this intervention, he continued to experience multiple desaturation episodes, with the oxygen level dropping to 85% and associated with tachycardia while on CPAP. Consequently, he was referred to the otorhinolaryngology (ORL) team for an upper airway assessment.

Surprisingly, flexible nasopharynx-laryngoscopy showed a single cystic lesion at the posterior nasal space, completely obstructing the left choanae and likely originating from the left side and crossing midline toward the right posterior nasal

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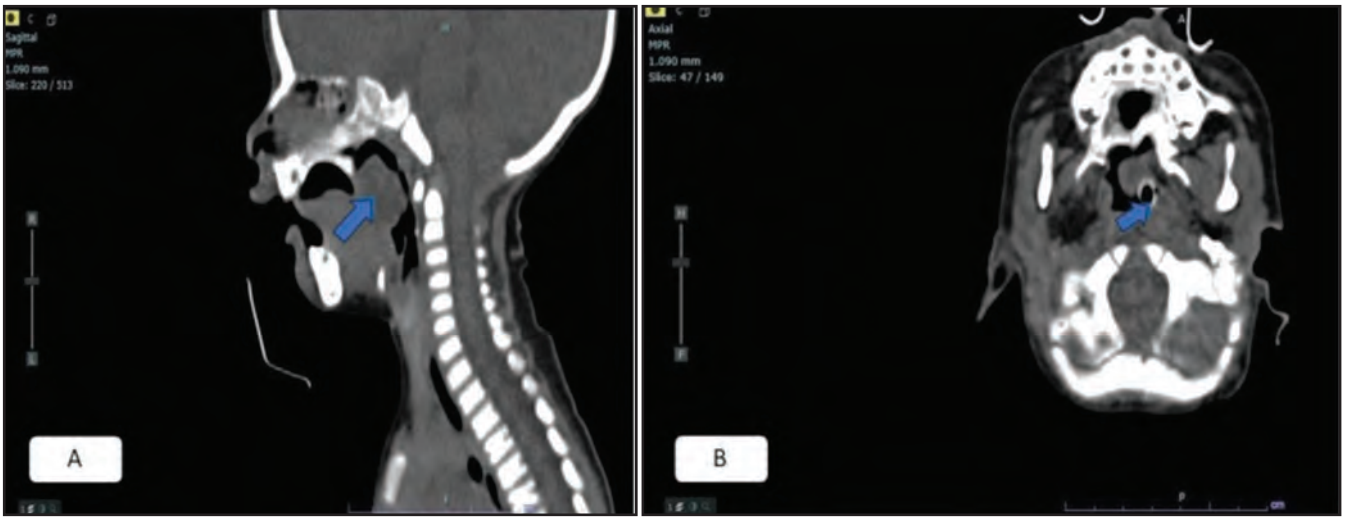


Fig. 1: A&B. Blue arrow; CECT brain showing a round non-enhancing cystic lesion likely arising from the left posterior nasal space protruding onto the nasopharynx measuring approximately 1.7 × 1.7 × 2.2 mm (AP × W × CC). There is thin enhancing septation at the inferior part of the lesion. No calcification, solid or fatty component within. There is no abnormal enhancement to suggest infiltration—the features of benign nasopharyngeal cysts.



Fig. 2: Intact cyst measuring 13 × 10 × 8 mm. The cyst is surrounded by fatty tissue measuring 8 × 3 × 3 mm. The cyst wall is 2 to 3 mm in thickness.

space. There were no features to suggest laryngomalacia. An urgent contrast-enhance computed tomography (CECT) scan of the neck and base of the skull was arranged. The CECT result indicated a benign nasopharyngeal cyst from the left posterior nasal space protruding onto the nasopharynx, measuring approximately 1.7 × 1.7 × 2.2 mm (AP × W × CC) (Figure 1).

After 6 weeks of admission and undergoing the CECT scan, he was discharged home with CPAP for use during sleep and was well at home. There was no respiratory distress, sleep or feeding issues. He was then re-admitted again 2 weeks after the discharge for excision and biopsy surgery under elective general anaesthesia (GA). During the surgery, a nasopharyngeal cyst and base (Figure 2) were removed and sent for biopsy. The surgery was successful without any complications, and he was able to wean off the CPAP postoperatively. A few days after the surgery, he was discharged on room air. Histopathological findings were consistent with features of mature cystic teratoma (Figure 3). Subsequent follow-ups at the outpatient ORL clinic showed no further noise or fast breathing, and he was clinically well. The ORL team will continue the surveillance by monitoring the symptoms like stertor and regular flexible scope during scheduled follow-ups. Paediatrics teams closely follow up with him to observe for any recurrence of teratomas elsewhere. At the same time, he continues the follow-up at the nearest health clinic for developmental and vaccinations.

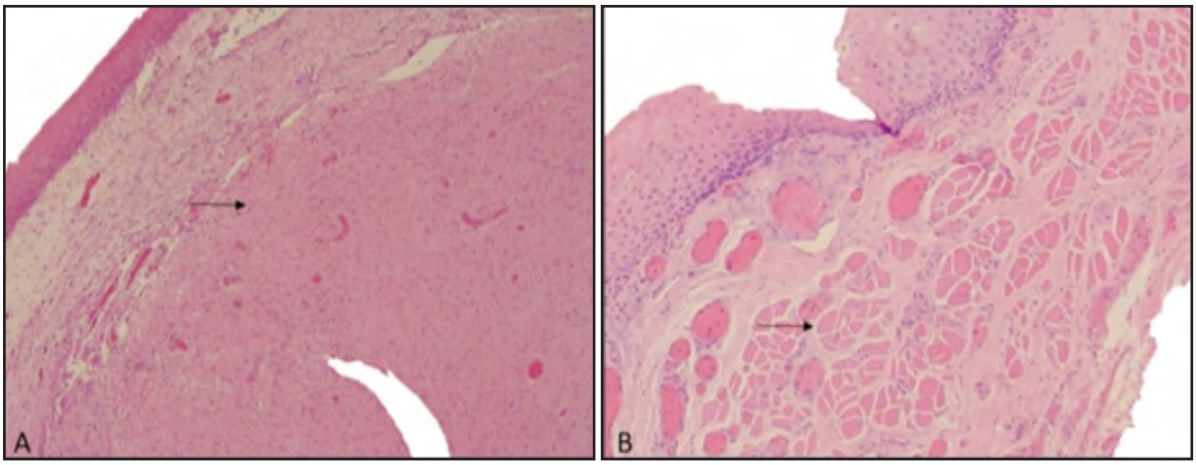


Fig. 3: A. Histopathology showing glial tissue within the subepithelial layer (arrow) (H and E stain, x 100). B. Histopathology showing smooth muscle bundles (arrow) with overlying pseudostratified squamous epithelium and congested vascular channels (H and E stain, x 200).

DISCUSSION

Noisy breathing can be described by its quality, location of obstruction, and underlying pathology. Stertor is a low-pitch sound from the base of the tongue and hypopharynx, often heard during sleep due to decreased pharyngeal muscle tone and gravity during inspiration. Stridor is a high-pitched sound resulting larynx and trachea obstruction, heard during inspiration, expiration or both.[3]

Neonates are obligate nasal breathers, meaning they exclusively breathe through their nose until six months, allowing them to breathe and eat simultaneously.[3] Obstruction from the nasal cavity downward is crucial for them. Most literature reports respiratory distress and feeding problems appearing in the neonatal period.[1,2,4,5] However, in this case, the patient had stertor since day ten of life, but he had no alarming issues like feeding, poor weight gain or respiratory distress until at four months old. Subtle symptoms led to missed recognition by the physician until signs and symptoms of upper airway obstruction developed.

That is why an early diagnosis of potential airway obstruction in neonates will improve outcomes. This can be obtained with a comprehensive history and physical examination, encompassing familial genetic factors, prenatal diagnoses, and birth-related details such as prematurity, labour duration, presentation, and delivery trauma. Recognising signs suggestive of nasal obstruction, including tachypnoea, nasal flaring, cyclical cyanosis, epistaxis, sternal retraction, and episodic apnoea, is crucial.

In the primary health clinic, airway obstruction can be assessed using a cold spatula test for nasal/nasopharynx patency or Ryle's tube insertion test. Consulting the ORL team is recommended before proceeding with Ryle's tube test to avoid complications. Referral to the ORL team for an endoscopic examination is essential to identify any possible obstruction from the nasal cavity, posterior choana and nasopharynx. This holistic approach at the primary care level significantly influencing outcomes, even though the most common causes of neonatal stertor do not cause immediate harm to the neonate.

Nasopharyngeal teratoma is a rare benign entity. During antenatal, ultrasonography can help detect pharyngeal teratoma early, often polyhydramnios and increase alpha-fetoprotein levels.[6] Differential diagnoses include other oropharyngeal masses like encephalocele, meningocele, nasal glioma, congenital rhabdomyosarcoma, haemangiomas, neurofibromatosis, congenital epulis, congenital epignathus, and lymphatic malformation. Nasopharyngeal causes of airway obstruction to consider are choanal atresia, choanal stenosis, mid-nasal stenosis, piriform aperture stenosis and craniofacial abnormalities.[2]

A CT scan was chosen for our case due to its immediate availability and the no need for general anaesthesia (GA). It effectively provided information on the mass's vascularity and extension. While magnetic resonance imaging (MRI) is ideal for evaluating soft tissue mass, it has a long waiting time and requires GA. The treatment for this case is complete surgical removal, and the timing is as early as possible. In our case, the surgery occurred eight weeks after admission and alpha-fetoprotein was not measured as it is not a sensitive marker.[2]

The prognosis depends on the patient's age, tumour resectability, and metastasis. Early resection, pathological evaluation for premalignant or malignant changes, and long-term follow-up are critical for positive outcomes.[7] The case series reveal that the survival and morbidity of head and neck teratoma is excellent following surgery, with rare recurrence.[2] Therefore, the primary care team play a crucial role in coordinating follow-up care, monitoring for recurrence or complications, and supporting appropriate growth and development.

CONCLUSION

Mature nasopharyngeal teratoma is a rare disease in neonates. However, proper assessment is warranted when a newborn presents with marked noisy breathing because it can lead to a life-threatening event that is high in morbidity and mortality. Even though nasopharyngeal teratoma in otorhinolaryngology is urgent in localisation, other

disciplinary teams, such as primary physicians, obstetricians, and paediatricians, should collaborate closely for a successful early diagnosis and treatment.

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DECLARATIONS

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