DOI: 10.7860/NJLM/2024/73169.2886

Case Report

Pathology Section

Beyond the Crib: Unraveling the Enigma of Neonatal Nasopharyngeal Mass

M SONALI SHANMATHI[1], K CHANDRAMOULEESWARI[2], M DOUGUL REGIS[3], A USHA NANDHINI[4], M SIDHU[5]

ABSTRACT

Germ cell tumours that develop at birth are rare, with teratomas most frequently occurring in the sacrococcygeal area. Nasopharyngeal teratomas are also rare, occurring in less than 10% of head and neck teratomas. Immature Nasopharyngeal Teratoma (INT) in neonates presents a rare yet challenging clinical scenario. It can manifest with significant respiratory distress and airway compromise, necessitating prompt recognition and intervention. Diagnostic modalities such as imaging studies and biopsy aid in accurate diagnosis and treatment planning. Surgical resection remains the cornerstone of management, with complete excision being essential to optimise outcomes. The present case report highlights the importance of multidisciplinary collaboration and continued research efforts to refine therapeutic approaches and improve patient outcomes. Here, the authors present a rare case of INT in a 51-day-old male child. A 51-day-old male child, normal vaginal delivery and cried immediately after birth with complaints of respiratory distress from 8th day of life, for which he was treated with antibiotics and started on mechanical ventilation. In view of failure of extubation, bronchoscopy was done which revealed a nasopharyngeal mass. Magnetic Resonance Imaging (MRI) showed a heterogeneous mass measuring 2.3×3.9×3.1 cm (anteroposterior×transverse×craniocaudal) involving the parapharyngeal space and carotid space, extending into the retropharyngeal space, with no evidence of intracranial extension. Histopathology showed features of an immature solid teratoma. Nasopharyngeal teratomas are extremely rare congenital tumours, with only a few cases reported in the medical literature. INT in neonates represents a rare yet clinically significant entity that requires prompt recognition and intervention. Multidisciplinary collaboration and comprehensive management strategies are essential to optimise outcomes for affected neonates.

Keywords: Adjuvant therapy, Immature nasopharyngeal teratoma, Multidisciplinary approach, Neonate

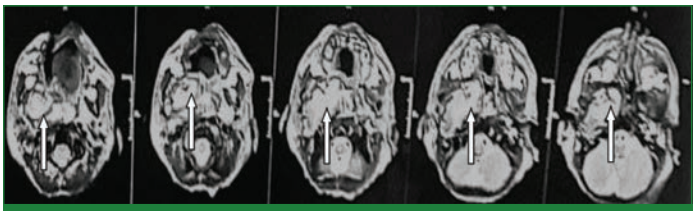
CASE REPORT

A 51-day-old male infant, delivered vaginally and who cried immediately after birth, had a birth weight of 2.62 kg, subsequently developed respiratory distress on the 8th day of life. The infant was treated with antibiotics and required mechanical ventilation. Due to failure of extubation, a bronchoscopy was done which revealed a nasopharyngeal mass. Laboratory findings are listed in [Table/Fig-1]. Serological studies for Hepatitis B surface antigen and Hepatitis C virus antibody were negative. MRI showed a heterogeneous mass measuring 2.3×3.9×3.1 cm (anteroposterior× transverse×craniocaudal) involving the parapharyngeal space and carotid space, extending into the retropharyngeal space, with no evidence of intracranial extension, as shown in [Table/Fig-2]. Based on the clinical presentation, the differential diagnosis included teratoma, olfactory neuroblastoma, nasal glioma and nasal polyp. A transpalatal approach for debulking of the mass and tracheostomy was done. Gross examination showed a single grey-brown soft-tissue fragment, and many teeth were noted. The grey-brown soft-tissue fragment measured 2.5×2×0.5 cm, as shown in [Table/Fig-3]. The cut surface exhibited mild congestion.

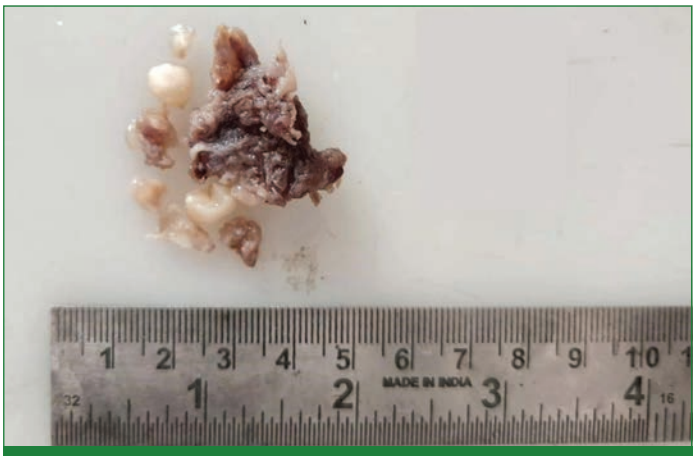
Haematological parameter	Units	Patient values	Normal values
Haemoglobin	g/dL	13.8	9.4-13.0
Platelet	×10^3/µL	5.58 lakh	150-450
Total white blood cell counts	×10^3/µL	17,600	9.0-37.0
Bleeding time	Minutes	1 minute 35 seconds	2-6 minutes
Clotting time	Minutes	5 minutes 45 seconds	2-8 minutes

[Table/Fig-1]: Summary of laboratory data [1].

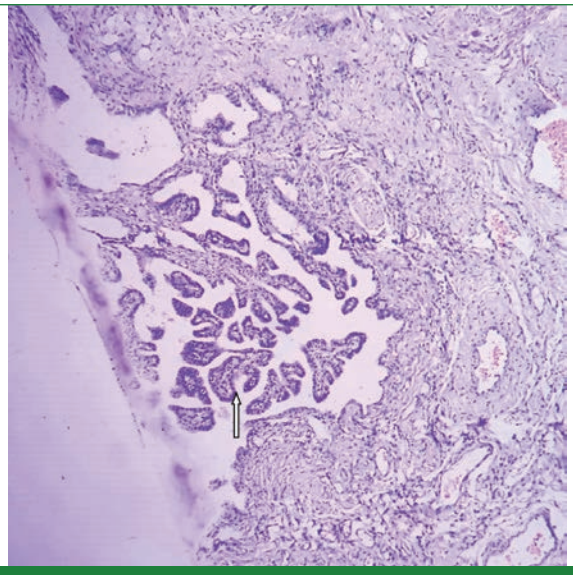
Microscopic examination revealed fibrous stroma with spindle-shaped cells many congested blood vessels. Foci showing choroid plexus, mature glial tissue, and a neoplasm composed of small round blue cells arranged in sheets and lobules with hyperchromatic pleomorphic nuclei and scant cytoplasm, admixed with round to oval cells with pleomorphic nuclei, some showing prominent nucleoli and moderate eosinophilic cytoplasm [Table/Fig-4a-c]. Based on the histomorphology, the mass was diagnosed to be immature teratoma.



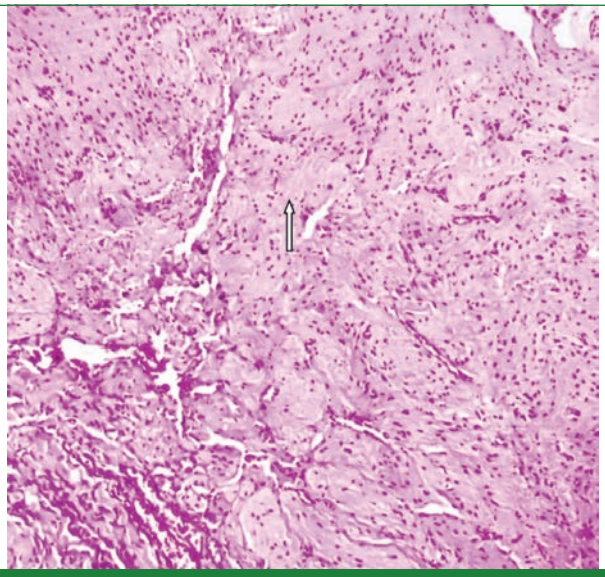
[Table/Fig-2]: MRI T1 Weighted (T1W) showing (White arrow) iso to mildly hyperintense collection within right side of neck involving pharyngeal and carotid space measuring 2.3×3.9×3.1 cm (anteroposterior×transverse×craniocaudal) with no evidence of intracranial extension.



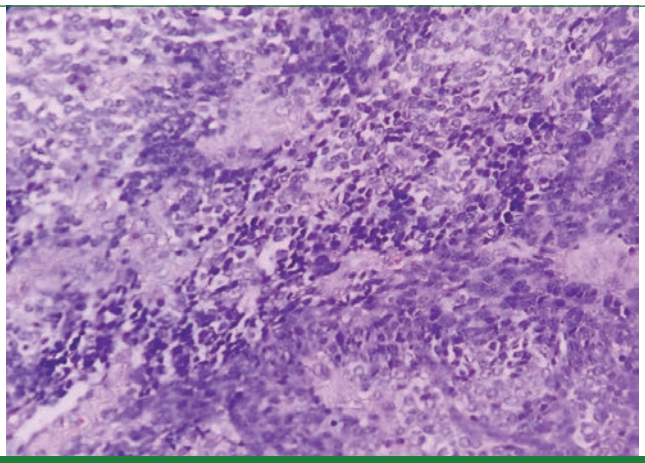
[Table/Fig-3]: Gross image showing single grey brown soft-tissue fragment and many teeth were noted.



[Table/Fig-4a]: Choroid plexus epithelium (white arrow) (H&E, 40x).

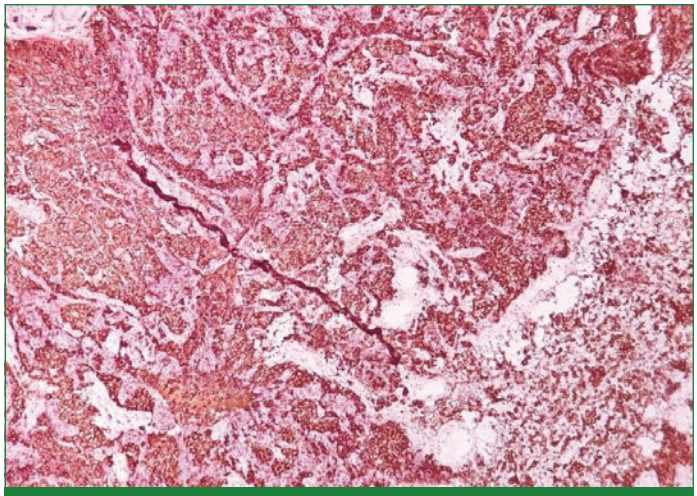


[Table/Fig-4b]: Glial cells with hyperchromatic round nuclei (white arrow) (H&E, 40x).

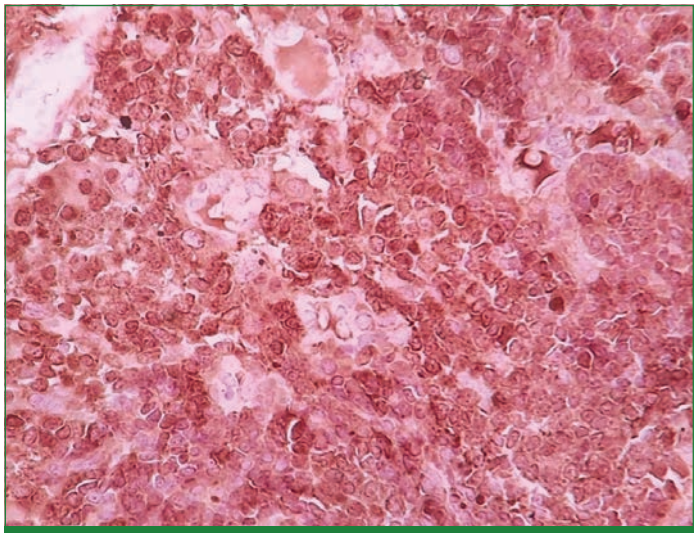


[Table/Fig-4c]: Sheets of small round blue cells (H&E, 40x).

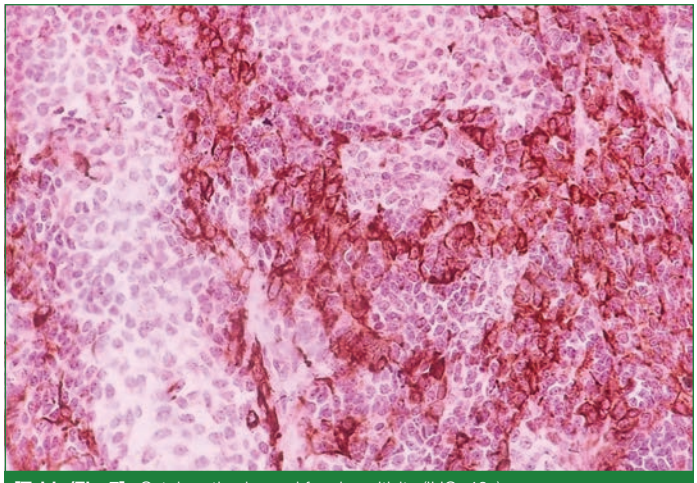
To confirm the diagnosis, a panel of immunohistochemical markers, including synaptophysin, chromogranin, Neuron-specific Enolase (NSE), desmin, cytokeratin, cluster differentiation 99 (CD99) and human melanoma black 45 (HMB45), were done. The following immunohistochemical findings were observed: synaptophysin showing diffuse strong cytoplasmic positivity in 70-80% of tumour cells [Table/Fig-5], while NSE demonstrated diffuse strong cytoplasmic positivity in 90-100% of tumour cells [Table/Fig-6]. Cytokeratin showing focal positivity [Table/Fig-7]. The child was discharged against medical advice and expired shortly after discharge.



[Table/Fig-5]: Synatophysin showed diffuse strong cytoplasmic positivity in 70-80% of tumour cells (IHC, 40x).



[Table/Fig-6]: Neuron-specific Enolase (NSE) showed diffuse strong cytoplasmic positivity in 90-100% of tumour cells (IHC, 40x).



[Table/Fig-7]: Cytokeratin showed focal positivity (IHC, 40x).

DISCUSSION

Teratomas are most commonly found in the sacrococcygeal region, occurring in approximately one in every 4,000 live births. The head and neck region represents the second most prevalent site, comprising 6-10% of all teratomas [2,3]. Sacrococcygeal and head and neck teratomas typically manifest within the first two months of life, whereas nasopharyngeal teratomas may present with symptoms at birth, as evidenced in the present case [4].

Nasopharyngeal teratomas are uncommon in newborns and are more prevalent in females. They encompass tissues originating from all three embryonic germ layers: ectoderm, mesoderm and

Authors	Month and year of publication	Gender/Age	Clinical features	Radiological findings	Treatment	Outcome
Isken T et al., [5]	September 2007	Female, newborn	Mass, blocked airway, bilateral cleft palate/lip, columnar sinus	Soft mass in nasopharyngeal area	Surgical removal at 94th day	Under follow-up
Mirshemirani A et al., [10]	June 2011	Female, 20 days	Mass, blocked airway, cleft palate	Soft-tissue tumour in nasopharynx with a focus of calcification of a tooth	Surgical removal of the mass, cleft palate corrected after 2 years	100% survival
Ramadas MM et al., [13]	April 2024	Female, newborn	Cystic and partially solid mass protruding through mouth and right nostril, with severe birth asphyxia	Poorly marginated soft-tissue density mass arising from the right ethmoidal air cell region, with extension into the nasopharynx and oral cavity, but no intracranial extension	Surgical removal of the mass	Death
Present case	2024	Male, 51 days	Respiratory distress	A heterogeneous mass involving parapharyngeal space, and carotid space extending in to theretropharyngeal space with no evidence of intracranial extension	Surgical removal	Death

[Table/Fig-8]: Comparison of published literature with the present case [5,10,13].

endoderm. These teratomas can be associated with additional anomalies such as cleft palate, microcephaly, atresia of the left common carotid artery and cardiac abnormalities [5].

Diagnosis of nasopharyngeal teratomas typically starts during pregnancy. Maternal indicators may include polyhydramnios caused by difficulty in swallowing and increased levels of alpha-fetoprotein. Airway blockage and breathing difficulties due to the size and position of these teratomas are the primary reasons for illness and death. For patients diagnosed prenatally with large cervical teratomas, the preferred delivery approach involves managing the condition outside the uterus during childbirth to optimise outcomes [6].

At birth, there is a potential for critical airway blockage that may necessitate immediate resuscitation, including interventions such as intubation aided by bronchoscopy or performing a tracheostomy [7,8]. After birth, imaging methods such as Computed Tomography (CT) scans or MRI scans are used to confirm the diagnosis and assess the extent of involvement of nearby blood vessels, bones, organs, or any extension into the skull [6].

In the present case, a nasogastric tube was successfully passed through the right nasal cavity. However, the nasopharyngeal teratoma was not observed during laryngoscopy, intubation and bronchoscopy, making it difficult to confirm the diagnosis. This was likely because the pedunculated mass slipped back into the upper nasopharynx during these procedures [9]. The prenatal identification of cystic hygroma might have redirected focus away from other potential causes of breathing difficulties in the neonatal period. Among cases diagnosed before birth, cystic hygroma is most commonly confused with cervical teratoma. The similarities in size, location, clinical features, and ultrasound findings can complicate distinguishing between these conditions [5].

Bronchoscopy should involve a thorough assessment of both the upper and lower airways. CT or MRI scans are crucial for distinguishing nasopharyngeal teratomas from other causes of neonatal neck masses, thereby improving management strategies, which, in this instance, involved surgical removal [10].

There are two types of teratomas: mature and immature. Mature teratomas are non cancerous and do not invade or spread to surrounding tissues. They can contain a variety of tissues such as hair, sweat glands, fat glands, teeth, nails, nerves, muscles, cartilage and bones. These teratomas may also include adipose tissue originating from all three embryonic germ layers, resembling organs like exocrine glands, liver, pancreas, thyroid, respiratory tract and gastrointestinal tract epithelium [11,12].

Teratomas are believed to originate from the development of embryonic cells, specifically germ cells in some cases. Alternatively, there is a hypothesis suggesting that teratomas can also arise from embryonic cells other than germ cells, which possess the potential to develop into various tissue types due to their complete genetic information [12].

Comparison of published literatute with the present case report has been presented in [Table/Fig-8] [5,10,13]. Although nasopharyngeal teratomas are benign tumours, they pose significant risks of morbidity and mortality. Hence, careful planning should include preparedness for potential airway emergencies when managing pregnant patients suspected of having a teratoma. Early surgical intervention is recommended for these infants. Studies have shown that early surgery reduces the duration of intubation, hospital stay, and intensive care unit stay [2].

The incidence of nasal teratoma is one in 4,000 births, with a female predominance of 6:1. The following conditions are also potential differential diagnoses for infantile nasopharyngeal masses: meningoencephalocele, congenital rhabdomyosarcoma, haemangiomas, neurofibromatosis, congenital epulis, congenital epignathus (which can occur in several instances) and lymphatic malformation.

CONCLUSION(S)

In conclusion, nasopharyngeal teratoma in neonates represents a rare yet significant clinical entity. Despite its infrequency, a thorough understanding and prompt management are imperative due to its potential for significant morbidity and mortality. The cornerstone of treatment remains early diagnosis followed by complete surgical resection of the tumour. The histological identification of the primitive neuroepithelial component is of paramount importance, and coexisting somatic malignancies has to be excluded. As immature teratoma in the nasopharyngeal location is a rarity, continued research and collaborative efforts are warranted to further elucidate optimal management strategies and improve patient outcomes in this challenging condition.

Acknowledgement

The authors would like to acknowledge Dr. Kanchan Murhekar, Additional Professor, Department of Oncopathology, Cancer Institute, Adyar, Chennai, Tamil Nadu, India, for her contribution to the present case report.

REFERENCES

[1] Bain BJ, Bates I, Laffan MA. Dacie and Lewis Practical Haematology. 2017, 12th ed. Elsevier.
[2] Wong BY, Ng RW, Yuen AP, Chan PH, Ho WK, Wei WI. Early resection and reconstruction of head and neck masses in infants with upper airway obstruction. Int J Pediatr Otorhinolaryngol. 2010;74(3):287-91.
[3] Tonni G, De Felice C, Centini G, Ginanneschi C. Cervical and oral teratoma in the fetus: A systematic review of etiology, pathology, diagnosis, treatment and prognosis. Arch Gynecol Obstet. 2010;282(4):355-61.
[4] Hossein A, Mohammad A. Huge teratoma of the nasopharynx. Am J Otolaryngol. 2007;28(3):177-79.
[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(5):1083-85.
[6] Elluru RG, Wootten CT. Congenital malformations of the nose. Cummings Otolaryngology. Head and Neck Surgery. 5th ed. Philadelphia: Mosby. 2010:2686-96.

[7] Azizkhan RG, Haase GM, Applebaum H, Dillon PW, Coran AG, King PA, et al. Diagnosis, management, and outcome of cervico facial teratomas in neonates: A Childrens Cancer Group study. J Pediatr Surg. 1995;30(2):312-16.
[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(3):277-78.
[9] Tiwari L, Baijal N, Puliyel JM. Nasopharyngeal teratoma as a cause of neonatal stridor. Indian Pediatr. 2009;46(12):1097-98.
[10] Mirshemirani A, Khaleghnejad A, Mohajerzadeh L, Samsami M, Hasas Yeganeh S. Congenital nasopharyngeal teratoma in a neonate. Iran J Pediatr. 2011;21(2):249-52. www.njlm.net
[11] Smirniotopoulos JG, Chiechi MV. Teratomas, dermoids, and epidermoids of the head and neck. Radiographics. 1995;15(6):1437-55.
[12] Varras M, Akrivis Ch, Plis Ch, Tsoukalos G. Antenatal sonographic diagnosis of pharyngeal teratoma: Our experience of a rare case with review of the literature. Obstet Gynecol Int. 2009;2009:180643.
[13] Ramadas MM, Das KH, Shaniba MK, Dilshath RK. A rare case of large nasopharyngeal teratoma. Kerala Medical Journal. 2024;17(1):38-40. Available from: https://doi.org/10.52314/kmj.2024.v17i1.629.

PARTICULARS OF CONTRIBUTORS:
1. Postgraduate, Department of Pathology, Chettinad Hospital and Research Institute, Chennai, Tamil Nadu, India.
2. Professor and Head, Department of Pathology, Institute of Child Health and Hospital for Children, Madras Medical College, Chennai, Tamil Nadu, India.
3. Assistant Professor, Department of Pathology, Institute of Child Health and Hospital for Children, Madras Medical College, Chennai, Tamil Nadu, India.
4. Postgraduate, Department of Pathology, Chettinad Hospital and Research Institute, Chennai, Tamil Nadu, India.
5. Postgraduate, Department of Pathology, Chettinad Hospital and Research Institute, Chennai, Tamil Nadu, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. M Dougul Regis,
Assistant Professor, Department of Pathology, Institute of Child Health and Hospital for Children, Madras Medical College, Tamil Salai, Egmore,
Chennai-600008, Tamil Nadu, India.
E-mail: dougulregis@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Jun 03, 2024
- Manual Googling: Aug 01, 2024
- iThenticate Software: Aug 09, 2024 (6%)

ETYMOLOGY: Author Origin

EMENDATIONS: 7

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **May 30, 2024**
Date of Peer Review: **Jun 20, 2024**
Date of Acceptance: **Aug 10, 2024**
Date of Publishing: **Oct 01, 2024**