

Case Report

Challenges in removal of congenital nasopharyngeal teratoma in a 5 day old neonate

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ABSTRACT

Neonatal stridor is a serious condition which poses a challenge in diagnosis and requires immediate evaluation of the underlying etiology. Tumors of the head and neck region are among the more unusual causes of airway obstruction in the neonatal period. Amongst them, nasopharyngeal teratomas are rare entities accounting for less than 10% of all tumours. We present the rare case of a nasopharyngeal teratoma causing inspiratory stridor and critical airway obstruction in a newborn at birth. This tumour was removed successfully using novel coblation technique in a 5 day old neonate managed without even a tracheostomy. We present the challenges faced in diagnosis and surgical management of teratoma at a site with difficult access like nasopharynx in a 5 day old newborn which could be safely and successfully removed using coblation technology.

Keywords: Tumors, Coblation, Neonatal stridor, Teratomas

INTRODUCTION

Neonatal stridor is a serious condition which implies airway obstruction in a newborn. This condition poses a definite challenge and necessitates immediate evaluation of the underlying etiology. In the newborn period, laryngomalacia, choanal atresia are the more common causes of inspiratory stridor. Tumors of the head and neck region are among the more unusual causes of airway obstruction in the neonatal period.¹

Teratomas are the common tumors encountered in the newborn period. The head and neck region is seldom involved; oropharyngeal and nasopharyngeal teratomas account for less than 10% of all neonatal germ cell tumors.¹

We present the rare case of a nasopharyngeal teratoma which was not identified in utero and presented with inspiratory stridor and critical airway obstruction in a

newborn at birth which was successfully removed by novel coblation technique.

CASE REPORT

Our patient was a male baby who presented with respiratory distress at birth. He was born to a 32 year old female G2P1L1 with a 35 week gestation pregnancy complicated by gestational diabetes with polyhydramnios (Amniotic Fluid Index– 30) for which she underwent caesarean section. Her antenatal examinations were reported as normal. In view of respiratory distress at birth with desaturation, baby was transferred to neonatal ICU. Baby continued to show severe retractions with stridor. Choanal atresia was ruled out clinically and baby was placed on oropharyngeal airway with continuous positive airway pressure support. Owing to high degree of suspicion, baby underwent a CT scan which showed a nasopharyngeal mass causing luminal obstruction.

Subsequently, MRI- face and neck with gadolinium contrast was done for better soft tissue delineation and vascularity which revealed a well defined solid cystic lesion measuring 2.2 (CC)×1.9 (AP)×1.7 (TR) cm within the nasopharynx causing almost complete luminal obstruction and inferiorly depressing the soft palate with no obvious intracranial extensions. No calcification or fat density noted. Mass was isointense/mildly hypointense on T1W images and hyperintense on T2W/ STIR images with delayed heterogenous enhancement seen post contrast suggesting thick and thin internal septations. Refer Figure 1 A and B for MRI images of tumor and its extent.

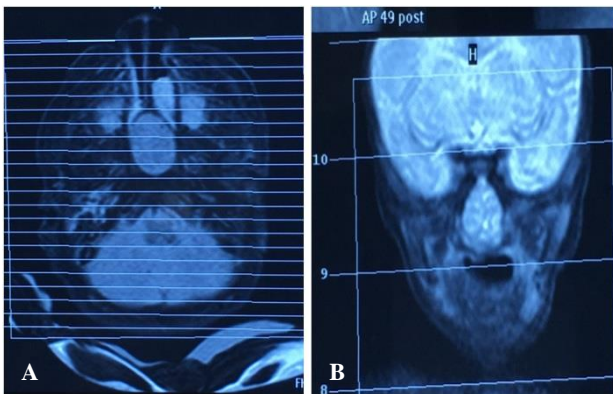


Figure 1 (A and B): MRI images of tumor and its extent.

With high suspicion of the mass being a teratoma, preoperatively, serum alphafeto protein (AFP) was done which showed raised AFP with 116177.120 ng/ml (normal <9.0 ng/ml). Patient was planned for surgery using novel coblation technique. In view of suspected tumor vascularity and low blood volume of a 5 day old baby of weight 2.45 kg, coblation was preferred.



Figure 2: Endoscopic view of tumour occupying nasopharynx.

Under general anesthesia, child was planned for removal of nasopharyngeal mass using endoscopic surgical approach by coblation technology. Refer Figure 2 for endoscopic view of tumor occupying nasopharynx. Child was placed in supine position with shoulder extension. Regular boyle davis mouth gag could not be used owing

very young age of child. Dingmans mouth gag was applied and soft palate retracted using nasal catheters. Nasal catheter could be passed only from one side as the other was completely obstructed by the tumour. Using Procize EZ wand and aided by visualisation by 70 degree endoscope, the tumour was slowly debulked starting from the center of the lesion creating an overall effect of the tumour collapsing on itself. Refer Figure 3 A and B for intraoperative view of tumour removal using coblation Procize EZ wand.

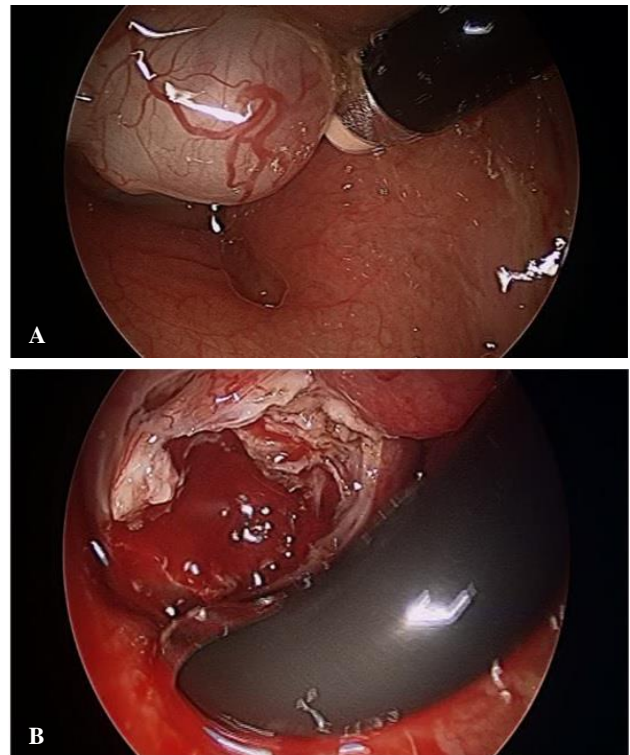


Figure 3 (A and B): Intraoperative view of tumour removal using coblation Procize EZ wand.

We took adequate specimens for biopsy for assessment of tumor margins. Superiorly the tumour was seen reaching up to the roof of sphenoid and could be completely removed without breaching the intra-sphenoidal mucosa. The tumour was noted to be attached to a shelf of atretic bone arising from the vomer which was removed. Laterally the tumour was free from any attachments to the tubal orifices and could be removed totally. As the tumour was adherent to the soft palate, minor injury to the soft palate was seen which was repaired primarily.

Post-operatively, child did not need any intubation and maintained oxygen saturation with the assistance of oropharyngeal airway on room air. Child was started oral feeding on post-operative day 3 and could tolerate oral feeds. Post operatively serum alpha fetoprotein was repeated on day 4 which reduced to 64661 ng/ml (normal 0-13 days: 4132.23-86776.85 ng/ml) indicating complete removal of the tumour. Refer Figure 4 for postoperative endoscopic view after complete removal of tumour.



Figure 4: Postoperative endoscopic view after complete removal of tumour.

On final histopathological examination, microscopy revealed a tumor composed of many cystic spaces lined variably with respiratory, stratified squamous and cuboidal to mucinous epithelium with salivary gland, cartilage and smooth muscle tissue also being observed. Prominent areas of neuroepithelial component consisting of proliferating immature round to oval cells present as neural rosettes and nests in a neurofibrillary background are noted with areas of hemorrhage compatible with a diagnosis of nasopharyngeal teratoma.

DISCUSSION

Establishing diagnosis in neonatal stridor is a challenge. Tumors of the head and neck region are among the more unusual causes of airway obstruction in the neonatal period. Teratomas are one of the main tumor types encountered in the newborn period out of which teratomas of the nasopharynx is a rare entity comprising only 10% of them.¹ Teratomas are true neoplasms originating from pluripotent cells and are composed of tissues from all three germinal layers. In childhood, these teratomas are usually reported as benign and seen most commonly in association with polyhydramnios as seen in our case.² Symptoms of teratoma of the nasopharynx are dependent on the size and extent of the lesion. Typically, it induces airway obstruction which requires rapid airway management especially in the neonatal period. Other symptoms include failure to thrive, difficulty in feeding, dyspnea.¹

Imaging studies are helpful in aiding diagnosis in addition to showing location and extent.³ In the present case, both CT scan and MRI with contrast, identified the nasopharyngeal mass and differentiating it from other tumours and in addition provided valuable information about skull base extension and suspected vascularity.⁴

The main treatment of teratoma is complete surgical resection with rare recurrences as reported in literature.⁵ Coblation technique has been traditionally used for varied conditions like adenoidectomy, tonsillectomy, channelling of palate. At the same time, coblation has been now proved to be an effective method for resection

of selected head and neck malignancies.⁶ Coblation creates plasma mediated dissociation of molecular bonds within the tissues with minimal thermal damage with additional advantage of coagulation so as to limit blood loss.⁵ Precise Plasma Wand used in coblation technique has excellent features which includes an enhanced flat-screen active electrode configuration that enables efficient bipolar ablation and coagulation during surgical procedures and a long, low-profile malleable wand shaft that allows increased surgical field visualization, adjustable to individual anatomy.⁵

As nasopharyngeal teratomas are often encapsulated or pseudo-encapsulated and not infiltrating, coblation technique was chosen which facilitates dissection of the teratoma from surrounding tissue structures.⁶ Bycoblation, extensive dissection of tissue planes could be avoided which is a tough prospect in a 5 day old baby.²

Hwang et al used coblation for removal of nasopharyngeal teratoma, but reported limitations in assessment of tumour margins.⁷ We took adequate specimens as one of the limitations of using coblation is described as difficulty in assessment of tumour margins.

Alpha fetoprotein is an important tumour marker for identifying recurrences and residual disease.⁸ In our case AFP levels were raised well above normal but post-operatively fell to normal levels indicating absence of residual disease.

CONCLUSION

Teratoma of the nasopharynx is a very rare lesion which can potentially cause fatal airway obstruction in the neonatal period. These tumours should be kept in mind especially in neonates with respiratory distress. We describe the successful identification of tumor at birth, challenges faced in diagnosis and surgical management of tumor and successful removal of such a teratoma in a site with difficult access using coblation technique in the youngest baby ever reported in literature.

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Ethical approval: Not required

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