CASE REPORT

NASOPHARYNGEAL BRANCHIAL CYST CAUSING STRIDOR IN NEONATE: A CASE REPORT
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ABSTRACT: Neonates may present with various causes of stridor. Nasopharyngeal cysts are rare congenital lesions causing difficulty in breathing. Differential diagnosis of nasopharyngeal cysts include thornwalds cyst, rathke cleft and craniopharyngioma, teratoma, epidermoid cysts, dermoid cyst, nasopharyngeal encephalocele, infected cyst and branchial cyst. Though branchial cysts are very rarely present in nasopharynx, they are easy to diagnose on MRI and fall under the category of preventable causes of neonatal stridor. Their surgical excision is the treatment of choice but sometimes owing to the other prematurities present the neonate period is not always the best time to operate. We report a case of a newborn child with nasopharyngeal branchial cyst presenting with stridor and managed conservatively with aspiration.

KEYWORDS: Nasopharyngeal Cyst, Branchial cyst, Neonate.

INTRODUCTION: Nasopharyngeal cysts are rare lesions, recognized infrequently on routine nasopharyngoscopy or imaging. These entities may be the source for spontaneous CSF rhinorrhea, purulent rhinorrhea refractory to antibiotics as well as steroids, rhinosinusitis, nasal obstruction, seizures and middle ear effusion. Though some of the disorders associated with nasopharyngeal cysts present in adults their development starts in childhood or even prenatally.¹

Ectopic nasopharyngeal cysts may arise from either first or second arch but most commonly it arises from the second arch.² Second branchial cysts are the most common type of branchial anomalies with a classical location either deep or along the anterior border of SCM, and are very rarely found in the nasopharyngeal or parapharyngeal space. Recent experiments with human embryo have shown that gene expression in the head region follows that seen in other species embryo model. The dlx homobox gene codes for regional specification of branchial arches and mutation in these genes is associated with the aberrant branchial arches.³

Nasopharyngeal branchial cysts are also the result of such mutations. Clinical manifestations of nasopharyngeal branchial cysts include rhinorrhea, unilateral middle ear effusion (due to their relatively lateral position), nasal obstruction, vomiting and even an oral mass.¹

Pathology of nasopharyngeal branchial cleft cyst is stratified squamous or columnar ciliated epithelium overlying a rich lymphoid tissue. The cystic contents may be clear, mucinous or seromucinous. MRI is the diagnostic modality of choice. A few dozen cases of ectopic nasopharyngeal branchial cysts have been described worldwide, with a greater incidence among the Asian population.⁴

We report a case of a 2 day old child presenting with nasopharyngeal branchial cyst causing stridor. This report adds another case of this rare pathology that can be a life-threatening condition in the neonate.
CASE REPORT

CASE HISTORY: A term baby 2 days old was referred to our hospital with complaints of intermittent stridor. The stridor was more in supine position and didn’t relieve when the patient was kept prone. The cry of the baby was normal. She could not maintain spo2 so was kept in NICU with oxygen prongs. The infant had a normal prenatal and natal history. Her birth weight was low about 2kgs. She was not able to take breast feeds.

On her ENT check up, soft palate was pushed forward obliterating her oropharynx completely. On palpation a non-pulsatile compressible cystic swelling was felt behind the soft palate. A MRI was done which documented a well-defined, homogenous cyst in the left posterolateral wall of nasopharynx obliterating the airway.

The fluid was isodense to vitreous and CSF T2 and T1 images, hence appearing serous. Surgical excision of the cyst was planned. Owing to LBW, a difficult intubation the anesthesiology department showed concerns regarding the survival of the infant and advised tracheostomy.

To avoid the morbidity in the infant we decided to aspirate the cyst. After aspiration, the baby was relieved immediately. The cytology of the fluid was normal. The baby she weaned of the oxygen within next 24 hours and was able to take breast feeds normally. She was discharged after observing her for one week following cyst aspiration. The patient’s is 1 month old at present and repeat aspiration has not been required till date.

DISCUSSION: Branchial apparatus forms during 3 rd to 7 th week of intrauterine life and contributes to many components of the head and neck region.\(^5\) It consists of 5 paired branchial arches & each separated externally by ectodermal clefts and internally by endodermal pouches.\(^6\) Branchial cysts attribute their occurrence the to the complex development of branchial apparatus which has been widely discussed in the literature.

The Eustachian tube is an endodermal structure opening in the nasopharynx formed from two branchial arches. Its Medial part arises from the first arch whereas lateral part is contributed from the second arch. Nasopharynx is a very rare location for the branchial cyst and they usually originate from the lateral nasal wall with inferior and medial extension. Two theories have been postulated in the evidence of an internal branchial cyst.

The first theory suggests that due to presence of subepithelial tissue the cysts are most likely to derive from ectopic epithelial cells. The second theory suggests that the cysts are derived from the remnant of branchial apparatus.\(^7\) Because the dorsal portion of 2nd branchial pouch blends with first pouch there is still controversy regarding which pouch contributes to the nasopharyngeal branchial cyst.

Shidara and colleagues described 2 cases of nasopharangeal cyst and considered them to originate from dorsal portion of 2nd branchial pouch. Meanwhile Papay and colleagues reported a case of nasopharyngeal branchial cleft and explained that its embryological origin was from the most lateral portion of the 2\(^{nd}\) branchial apparatus.\(^1,2\) Second arch branchial cysts were classified by proctor into 4 types:

Type 1: Lies superficial to ant.border of SCM
Type 2: Beneath the investing layer of deep cervical fascia
Type 3: Cyst passes between internal and external carotid arteries
Type 4: Lies adjacent to nasopharynx\(^8\)
MRI findings of branchial cyst do not differ from those of other cyst. It is a well marginated homogenous cystic mass, signal intensity is isotonc to the vitreous humour and CSF on T 1 and T weighted images. Mucosa of nasopharynx is composed of epithelium, lymphoid tissue and accessory salivary glands therefore d/d include rathke’s pouch, pharyngeal bursa, teratoma, meningocele, abscess, meningocele, ac polyp, mucocele of sphenoid, JNA, pleomorphic adenoma D/d of Thornwaldts cyst and Branchial cyst is based on the anatomical location. Branchial cyst is usually located close to ET while Thornwald cyst lies along midline.

The importance of diagnosis of nasopharyngeal cysts remains of paramount importance especially in neonates, as these may cause splaying of soft palate causing stridor. Though surgical excision is the treatment of choice; aspiration of the branchial cyst with or without sclerosing agent is the best conservative management, relieving the symptoms of nasal obstruction immediately in the neonate with almost no morbidity.

We strongly recommend the consideration of branchial cyst in the differential diagnosis of congenital nasopharyngeal masses leading to nasal obstruction in a newborn child especially in the Asian population. Aspiration of the cyst in NICU under aseptic conditions is a favourable early option for this entity once the diagnosis is confirmed on MRI.

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Image of the neonate showing bulge in oral cavity due to the nasopharyngeal branchial cyst

Radiological images of the neonate showing cyst in nasopharynx in sagittal and axial sections in T2 and in axial section in T1 MRI images
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