DOI: 103126/JNPS.V4113

Case Report

Unilateral Nasal Pyriform Aperture Stenosis Presenting with Respiratory Distress – A Case Report

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Article History
Received on - 2023 Sep 24
Accepted on - 2024 Jan 03

Keywords:

Abstract

Congenital nasal pyriform aperture stenosis is a rare entity and if present unilaterally can be missed. We present a four days old newborn with noisy breathing and worsening respiratory distress. The baby was managed with high humidified flow nasal cannula. Computed tomography confirmed congenital pryiform aperture stenosis and was managed surgically. Failure of passage of infant feeding tube through nares CNPAS needs to be kept in mind. Diagnosis and treatment of unilateral nasal pyriform stenosis is vital and can be life saving.

Online Access

DOI: https://doi.org/10.60086/jnps524

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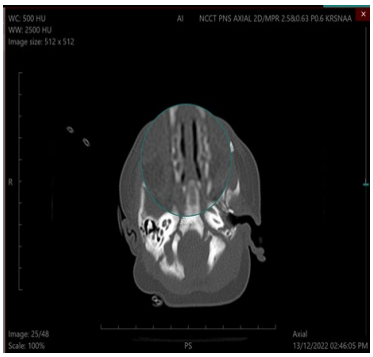
Introduction

Neonatal nasal obstruction due to congenital nasal pyriform aperture stenosis is very rare and uncommon with incidence of 1 in 5,000 to 8,000 live births.[1-5] It typically presents with clinical features that may mimic posterior choanal atresia. It is important to differentiate it from choanal atresia as there are differences in clinical management of both the conditions.[2] The usual clinical presentation is respiratory distress, cyclic cyanosis, apnoea, and feeding difficulties. A bony overgrowth of the maxillary nasal processes is thought to be responsible for this deformity. It has been suggested that a pyriform aperture width less than 8 mm in a term infant is diagnostic of congenital nasal pyriform aperture stenosis[2]. This anomaly has been reported as an isolated feature or can be associated with craniofacial or central nervous system anomalies. Other associated malformations include hypopituitarism, and craniofacial, cardiac, and urogenital anomalies.[6] We are reporting the first case in the northern part of Karnataka, India diagnosed as congenital nasal pyriform aperture stenosis. The baby was managed surgically at Shri BM Patil Medical College Hospital and Research center, Vijayapura, India and has remained symptom free.

Case Report



A 34 week preterm neonate presented noisy breathing, with respiratory distress. The baby was initially started on hood O_2. There was difficulty in passing infant feeding tube no-6 through the right nostril. ENT surgeon opinion was taken and Computed tomography - Para nasal sinuses was advised. In the subsequent course of illness, respiratory distress failed to settle, hence was started on heated humidified high flow nasal cannula along with inotropes dobutamine and adrenaline. 2 D Echocardiography was done which revealed patent fossa ovalis, with grade 1 tricuspid regurgitation. Computed tomography scan done on of paranasal sinuses revealed pyriform aperture stenosis on the right

side. (Figure 1)

Surgery for dilatation of the aperture was planned and was shifted for surgery once the neonate was hemodynamicaly stable. Local infiltration with 2% adrenaline diluted in 2.5ml of distilled water over the mucosal surface of the upper lip. Under microscopic view, 1 cm of sub labial incision was given and mucopricondrial and mucoperiosteal flaps were elevated upto inferior turbinate. After the visualization of narrowed pyriform aperture, 1.8 mm burr aperture was drilled and widened. The aperture was narrowed due to the thick anterior nasal spine and floor of the lateral wall of nose. Nasogastric tube 6 mm could be easily passed through the nasal cavity bilaterally. 2 mm endotracheal tube was passed in both nostrils and was cut and sutured with columella to function as a stent. Incised mucosa was sutured (Figure 2). Nasal suctioning was done hourly. The baby recovered well during the post -operative period.

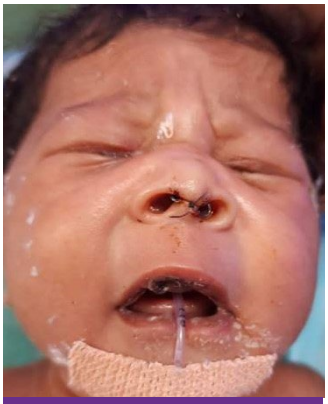


Fig 1: Post op status of the baby

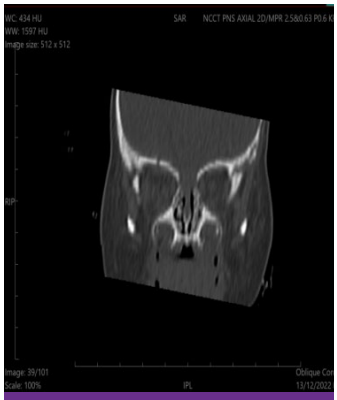


Fig 2: CT scan of the paranasal sinuses

Postoperatively baby was on hood oxygen which was well tolerated. In due course, feeds were started and inotropes were tapered and stopped. Oxygen inhalation along with nebulisations were continued with Endotracheal tube no 2 in situ within the nasal cavity. After a week postoperatively Endo tracheal tube no-2 was removed from both nasal cavity. Oxygen was continued for few more days. Subsequently baby maintained well on room air and direct breast feeding was started.

Discussion

Neonates are obligatory nasal breathers and hence, malformations like congenital nasal pyriform aperture stenosis may present as a life-threatening condition. Immediate intervention after birth is required in congenital nasal pyriform aperture stenosis. Congenital nasal pyriform aperture stenosis is frequently associated with a solitary maxillary central incisor,[10] which has been reported as an isolated morphogenic defect or can be associated with other midline defects such as holoprosencephaly and pituitary deficiencies. The presence of associated brain, pituitary and chromosomal anomalies should be excluded by the mean of magnetic resonance imaging, endocrinologic and genetic evaluation

In general, management of congenital nasal pyriform aperture stenosis depends first on the overall prognosis of the patient, and second on the severity of obstruction. In patients with a good outlook, the choice of treatment is conservative for those with less severe obstruction. However, surgical correction becomes necessary for those with complete obstruction.[8] The diagnosis of this rare condition is based on clinical evaluation. However, confirmatory diagnosis may require nasal endoscopy and especially, computed tomography scans. The inability to pass a 5 French (F) catheter and a radio graphically measured pyriform opening less than 8 to 10 mm in a full-term new born are considered as a relevant stenosis. In turn, the aperture is considered satisfactory when it allows for the passage of an endotracheal tube stent with an inner diameter of 3.5 mm.[8,9] However in our case neonate had a complete obstruction and was managed surgically and nasogastric tube 6 mm could be easily passed through the nasal cavity bilaterally. 2 mm Endotracheal tube was passed in both nostrils and were cut and sutured with columella to function as a stent. Accordingly, in another case series, 0.1 Percent intranasal dexamethasone drops were used successfully for conservative treatment in three out of five patients.[10] However our case did not need conservative management.

Conclusion

Passage of infant feeding tubes through both nares should be practiced to check for nasal patency. Failure to negotiate infant feeding tube through either nares such anomalies congenital nasal pyriform aperture stenosis should be kept in mind and should be differentiated from congenital choanal atresia through clinical examination and passage of infant feeding tubes. Diagnosis should be confirmed through computed tomography. Early detection and surgical intervention to relieve blockage is crucial in management of congenital pyriform aperture stenosis.

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