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ORIGINAL ARTICLE

Congenital nasal pyriform aperture stenosis: The report of four rare cases

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KEYWORDS

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Abstract Congenital nasal pyriform aperture stenosis (CNPAS) is an unusual cause of upper airway obstruction that may happen because of respiratory distress, cyclic cyanosis or feeding problems. Bony overgrowth of maxillary nasal process sounds responsible for this deformity. Diagnosis is based on clinical evidence and radiologic findings. Failure to conservative management, respiratory distress and feeding problems are all indications of surgical treatments. In our experience, the sublabial approach is known as the most effective to these patients at any age. Otolaryngologists and pediatricians should therefore consider CNPAS as a rare but treatable cause in the patients with upper respiratory problems.

1. Introduction

Nasal obstruction is a common disorder encountered during childhood. Nasal discharge, foul smelling, open-mouth breathing and some evidence of rhinosinusitis are some manifestations of nasal obstruction.[1] Congenital nasal pyriform aperture stenosis (CNPAS) is an unusual cause of nasal obstruction leading to respiratory distress in neonate. CNPAS can be manifested with apnetic episode, cyclic cyanosis, sudden total airway obstruction which clinically is similar to choanal atresia or stenosis. CNPAS can occur isolated or associated with other anomalies. Radiologic imaging confirms the diagnosis and is useful in surgical planning. In this experiment, we represent four cases with CNPAS nasal obstruction unresponsive to conservative medical management.

1.1. Case 1

A 3-year-old boy was referred to the Otolaryngology department of Pediatrics hospital affiliated to Tabriz University of Medical Sciences, Iran with chronic mucopurulent nasal discharge, difficulty in nasal breathing, foul smelling from nares and some sleep disorders such as open-mouth breathing and snoring. The patient also had passed several courses of rhinosinusitis with a temporary response to medical management. The patient was normocephal with normal development and no similar familial history.

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Examination revealed very narrow and asymmetric nares, as the lateral nasal wall was medially displaced which was in close proximity to the septum. It made it difficult to view the nasal cavity. To rule out the choanal atresia we attempted to pass a 2 mm polyethylene catheter, but it passed through no more than 1 cm in both nares. He had no other abnormality except solitary incisor teeth. A CT scan with fine contiguous sections demonstrated a narrow pyriform aperture in combination with a small nasal cavity, because nasal walls were medially displaced mostly in anterior segments related to the maxillary bone. The maxillary sinuses were also hypoplastic. No other craniofacial disorders such as choanal atresia or any evidence of central nervous system defects were seen. (Fig. 1) Brain MRI was normal, too. Endocrine examinations showed no abnormalities. Because of recurrent episodes of chronic rhinosinusitis unresponsive to multiple courses of medical management, surgical treatment was recommended.

1.2. Case 2

A 3-month boy with nasal obstruction and after-birth feeding problem was referred to the ENT Clinic of Tabriz Pediatrics Hospital with the probable diagnosis of choanal atresia. Initial examinations proved normal choans and a bilateral pyriform aperture stenosis with solitary incisor teeth. The patient had no other abnormalities. (Fig 2) The conservative treatment was hardly successful. The authors recommended surgical operation which was postponed due to some financial reasons.

1.3. Case 3

A newborn female was referred to the ENT Clinic of Tabriz Pediatrics Hospital with symptoms of mesomelic upper limbs, short stature and a premature dentition. She suffered from a severe nasal obstruction. With no similar familial history and solitary incisor teeth, the patient was normocephalic with hypertrophic alveolar bone due to a premature dentition. Initial examinations to rule out other organ abnormalities were

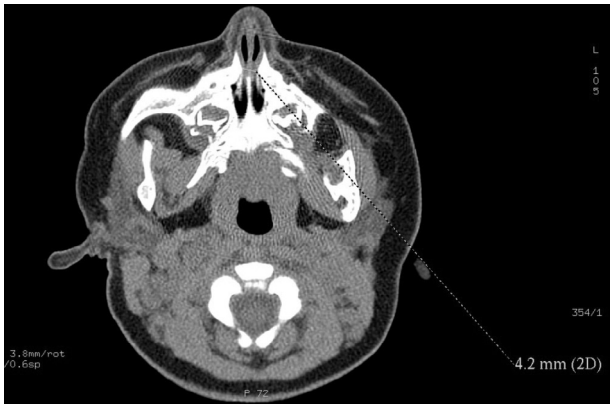

Figure 2 An axial section shows narrow 4.2 mm diameter of pyriform aperture.

conducted. The patient's plain radiography displayed mesomelic upper limbs, hemi vertebrae and deformity of ribs. (Fig 3) A CT scan of the nose showed severe CNPAS. The conservative treatment for nasal obstruction failed, hence a surgical treatment was recommended.

1.4. Case 4

A 6-year-old girl was referred to the department of chronic open mouth breathing and recurrent sinusitis at Tabriz Pediatrics Hospital. She had the history of nasal trauma at the age of 3. On her physical examination, she showed the long face syndrome due to chronic mouth breathing, anterior nasal stenosis, solitary incisor teeth, and septal deviation. A CT scan of the nose proved the findings. Considering the long face syndrome, conservative septoplasty and CNPAS surgery were recommended.

2. Surgical techniques

A sublabial approach to premaxilla was selected. An incision was made in the mucosa preserving 2 mm of the mucosa on

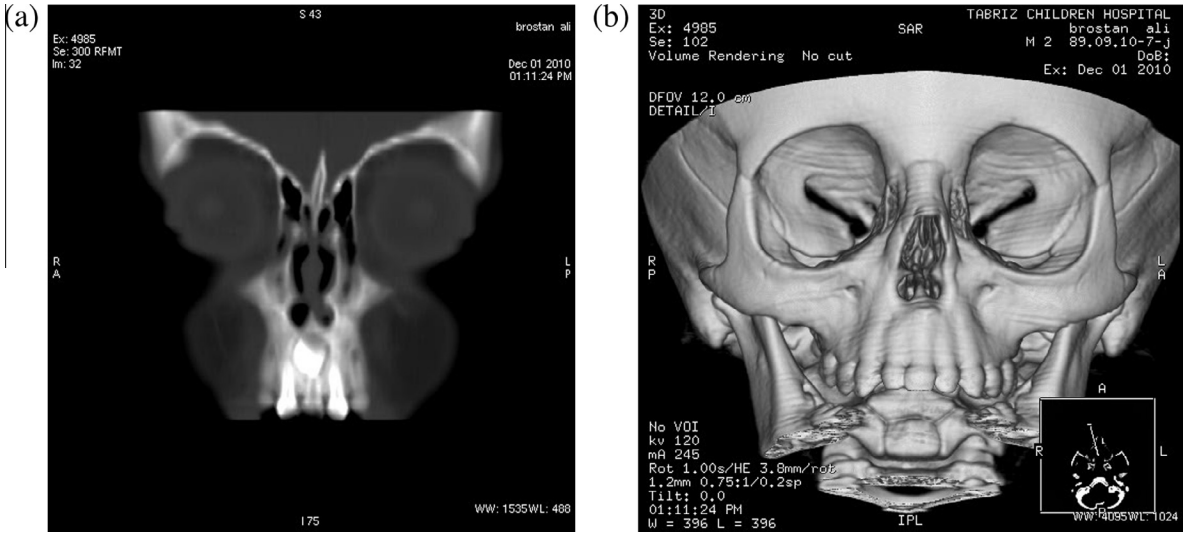

Figure 1 (a) Coronal CT scan shows medial approximation of nasal process of maxilla causing pyriform narrowing and (b) 3D-CT shows pyriform stenosis.

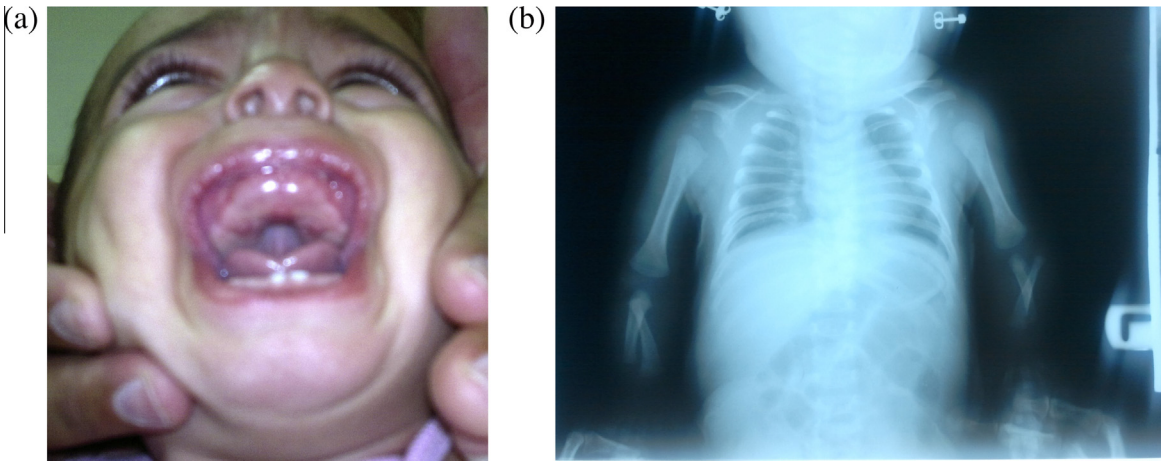

Figure 3 (a) A newborn female with premature dentition and severe nasal obstruction (b) plain radiography shows hemi vertebrae and deformities in ribs.

gingival side and dissection in subperiosteal plane to identify the anterior nasal spine and pyriform aperture. Under magnification, lateral aspects of aperture were enlarged with diamond burrs, preserving the nasal mucosa. Nasolacrimal ducts were preserved bilaterally with the dissection remaining anterior to inferior turbinate. Drilling on the nasal floor was avoided to prevent possible injuries to teeth buds. A releasing mucosal incision was made in the nose floor. The beveled edge of nasotracheal tubes (its size is decided upon the patient's age) was introduced bilaterally to secure their sublabials. The postoperatively parenteral antibiotics were administered. Parents were instructed for saline irrigation and suctioning of the tubes. Patients were discharged 2 days post operatively, while kept stents for 10 days. Saline irrigation of the nose was continued after extraction of tubes. One of our patients rejected the recommended surgery, while the other 3 patients underwent the planned surgical technique. All 3 patients were discharged 2 days after surgery on oral antibiotics (azithromycin 5 mg/kg BID) for 5 days.

After 2 weeks, one of the patients was infected with acute purulent sinusitis due to disregarding the nasal wash and hygiene. The patient's symptoms were resolved after receiving antibiotics and regular cleaning. In serial follow-ups and the final follow-up after 6 months, patients had no obstructions. Their breathing was normal and through the nose. Parents reported no snoring by the patients.

3. Discussion

Congenital nasal pyriform aperture stenosis (CNPAS) is a rare but life threatening nasal obstruction in neonates as they are nasal obligatory in 6–8 weeks of life.[1] Bony obstruction of nasal cavity either unilaterally or bilaterally is commonly due to choanal atresia.[2] CNPAS occurs at a frequency of about one-fourth to one-third of choanal atresia.[3] Some other causes can lead to nasal obstructions including meningocele, meningoencephalocele, dermoid and epidermoid cysts, and sinonasal tumors of any origin, septal dislocation and/or hematoma, choanal atresia and nasal hypoplasia due to warfarin teratogenicity or chondrodysplasia.[4] These causes should be considered in differential diagnosis of CNPAS.

Anatomically, pyriform aperture involves nasal and maxillary bones. CNPAS is hypothetically caused by the uncontrolled growth of the maxillary nasal process bone that occurs within the first 4 months of fetal development.[1,4–6] CNPAS can occur isolated or in association with other abnormalities such central nervous system, endocrine and craniofacial abnormality.[5] The most likely hypothesis is that CNPAS is a microform holoprosencephaly (malformation caused by incomplete cleavage embryonic holoprosencephalon).[6]

Holoprosencephaly, as the most common developmental anomaly in human forebrain occurs in an incidence of 1:16,000 live births. Clinical presentation of holoprosencephaly includes hypotelorism, midline cleft lip, cerebral malformations, learning disabilities, hypopituitarism, and a single maxillary central incisor.[7] To exclude associated abnormalities, the patients should undergo evaluation by magnetic resonance image, endocrine and genetic assessment.[4,8,9]

CNPAS is clinically presented with unspecific symptoms such cyclic cyanosis that becomes worse with feeding and better with crying, aspiratory strider, sternal retraction.[6,10,11] Feeding problems and sleep difficulties are also associated.[12] Diagnosis might be delayed in less severe cases, as it was observed in our own patients even by the age of 6 years-old. At this age, midface bone deformities such as adenoid face have occurred.

The diagnosis is based on clinical evaluations including nasal endoscopy and imaging. The recommended imaging option is CT scan applying fine contiguous midface sections (1–3 mm) in the axial mode.[4] CT scan provides an accurate diagnosis and rules out other such causes as choanal atresia.[13] The presence of an entry in the narrowed anterior nasal and overgrowth maxillary nasal process confirms the diagnosis.[4] Each pyriform aperture width less than 3 mm, or a whole pyriform aperture width less than 8 mm confirms the CNPAS diagnosis.[3]

Conservative or surgery treatment should be performed based on the prognosis of patients and the severity of obstruction.[5] If the nasal obstruction is mild, topical nasal decongestants, humidification and suction are possibilities.[1]

Lee and colleagues presented two patients with nonsurgical treatment and reported favorable outcomes. They suggested that if the patient could tolerate nonsurgical therapy, symptoms would be improved within 6 months of birth. Significant

indications for surgical intervention are respiratory distress, or feeding problems.[14] Although it seems better to postpone surgical procedures until the infant reaches 10 lb, 10 weeks of age and/or 10 mg/dl of hemoglobin (the rule of 10), but If the patient's respiratory condition is not stable and does not respond to medical management, surgery within 10–15 days can be advocated.[1] The most appropriate approach is sublabial incision, aperture dissection and widening the lateral wall with diamond burrs under magnification as described earlier.[15] The transnasal approach increases the risk of soft tissue trauma.[11] Our experience with 3 patients who underwent surgery proved that sublabial approach can be done safe at any age. Attention must be paid not to injure soft tissues and the nasolacrimal duct. This procedure has little complication and is effective enough. Even in syndromic cases, surgery improves the nasal breathing and prevents adverse effects of obstruction.

4. Conclusion

CNPAS is a rare complication that causes nasal obstruction or respiratory distress which demands intensive care and management. Although medical management may be beneficial for mild cases, respiratory distress, feeding problems and unresponsiveness to conservative treatments are obvious indications of surgery. Surgery can be an attempt as early as 10–15 days via a sublabial approach to enlarge the narrowed pyriform aperture.

CNPAS is difficult to diagnose in patients with less symptoms and possible to get delayed. Considering this diagnosis and performing proper evaluation in patients with recurrent rhinosinusitis or history of breathing problems can be helpful. However, pediatricians and otolaryngologists should consider CNPAS as a diagnosis in patients with respiratory problems.

Conflict of interest

None declared.

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