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Case Report

Congenital nasal pyriform aperture stenosis in association with solitary median maxillary central incisor: unique radiologic features

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ABSTRACT

Solitary median maxillary central incisor (SMMCI) coexists in 34%-65% of patients initially diagnosed with congenital nasal pyriform aperture stenosis. SMMCI, a genetic syndrome, warrants consideration for further screening because of its high prevalence of other diagnostic possibilities—specifically central defects, like nasal obstruction and hypothalamo-pituitary axis abnormalities. We report on a presentation of SMMCI with congenital nasal pyriform aperture stenosis which highlights the unique radiologic features and notes the relationship between these two central associated findings in the literature.

Introduction

Respiratory distress in infants presents as nasal flaring, rapid and shallow breathing, cyanosis, and failure to thrive secondary to feeding difficulties. Infants are preferentially nasal breathers; therefore, nasal obstruction can result in respiratory distress [1]. The more common causes of nasal obstruction include rhinitis, adenoid hypertrophy, deviated nasal septum, and polyps [2]. However, anatomic causes must also be ruled out such as choanal atresia, midnasal stenosis, and congenital nasal pyriform aperture (PA) stenosis (CNPAS).

CNPAS is a rare cause of nasal obstruction that occurs around 1 in 25,000 births [3]. It is thought to arise from disrupted interaction between the nasal placode and the nasal processes that form the central maxilla and help establish the internal nasal aperture at the PA [4,5]. The excessive bone growth in the nasal processes of the maxillary bones results in stenosis of the PA [2]. The diagnosis is made with computerized tomography (CT) images. Belden et al. established diagnostic measurements on CT images of the PA taken parallel to the hard palate with 11 mm or less width being diagnostic of CNPAS. Furthermore, in looking at age cohorts with CNPAS,

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width between the medial aspect of the maxilla at the level of the inferior meatus were measured on axial CT images to be 4.8, 7.0, and 6.0 mm in patients with CNPAS in the age ranges of 0-3, 4-6, and 10-12 months, respectively. This compared with 13.4, 14.9, and 15.6 mm in control subjects in the same age ranges [3]. Wormald et al. [6] described in their literature and retrospective case review that a PA width of 5.7 mm or less required surgical intervention.

During the diagnostic workup for CNPAS patients, there can be associated findings—seen incidentally on CT scan or physical examination—that warrant further exploration for the possibility of a genetic syndrome. Particularly apparent when evaluating the PA on imaging are the unerupted maxillary incisors. When evaluating a child for nasal obstruction, the physical examination, CT, or dental x-ray, will clearly show the maxillary dentition presence of solitary median maxillary central incisor (SMMCI; Fig. 1). Of note, 90% of infants with SMMCI will have CNPAS, choanal atresia, or midnasal stenosis. CNPAS and SMMCI seemingly coexist in a higher percentage than previously reported historically in the literature. Cases seen in isolation should be given consideration for further screening, specifically when a SMMCI is noted.

Case report

A 9-month-old female child presented to the emergency department from the pediatric office for failure to thrive, repeated bouts of cyanosis, worsening difficulty feeding, and recurrent "sinus" infections. Otolaryngology consultations were obtained. On initial evaluation, anterior rhinoscopy with an otoscope showed bilateral yellow drainage and extremely swollen mucosa at the PA. A pediatric nasal fiberoptic endoscope (2.7-mm diameter) was not able to be passed through either nares. A lubricated 5-French plastic catheter was similarly not able to pass. The child was not in acute distress with stable vitals and saturation of 98% in room air. Afrin (oxymetazoline) sprays and nasal saline mist were recommended along with cool mist mask next to the child in the crib. Calorie count, strict ins and outs, and observed feeding was planned. The child was placed on a monitored unit and a noncontrast maxillofacial CT scan with 1-mm cut through the nose was obtained.

CT scan revealed a PA width <5-mm bilaterally and no apparent midnasal stenosis or choanal atresia (Figs. 2, right image and 3, right image). On review of the CT scan for surgical planning, a single maxillary central incisor was noted (Fig. 1 middle and right image). Endocrine and genetic consults were ordered, and the child was screened for other associated abnormalities and magnetic resonance imaging (MRI) of the brain was ordered. Once workup was completed, noting the child showed no central abnormalities, the family was counseled on PA surgical repair. The surgery was performed through a sublabial incision elevating the mucosa off the nasal floor and medial maxilla. The medial maxillary prominences were drilled down until size 5 endotracheal tubes were able to be passed with minimal resistance. These size 5 endotracheal tubes were then trimmed to size and left in place as stents. These stents were removed on postoperative day 2, and nasal hygiene rinses were started. After discharge, the patient's nasal obstruction was improved, and further surgery was not indicated.

Discussion

Diagnostic workup of a CNPAS patient may unearth an incidental finding of a median maxillary incisor on CT imaging. This characteristic finding is part of a syndrome called SMMCI, a genetic syndrome that occurs in 1:50,000 live births and represents a spectrum of midline facial defects and other possible congenital anomalies including intellectual disability in up to 50% of patients [4,7]. Phenotypic characteristics at birth include an arch-shaped appearance of the upper lip, prominent maxillary alveolus, absent labial frenulum, narrow nose, and "V"- shaped palate with a prominent narrow ridge along the midpalatal suture [4]. Case studies have also correlated the presence of SMMCI with conditions such as microcephaly, cardiac defects, holoprosecenphaly (HPE), VACTERL, CHARGE, and velocardiofacial syndrome [4,7].

These phenotypic characteristics of SMMCI are thought to arise due to an interruption in development during gestational days 35-38 [4,8,9]. Interaction between the nasal placode and the nasal processes that form the central maxilla are disrupted leading to cranial facial, dentition, and nasal airway malformations. CNPAS may be an isolated anomaly. However,

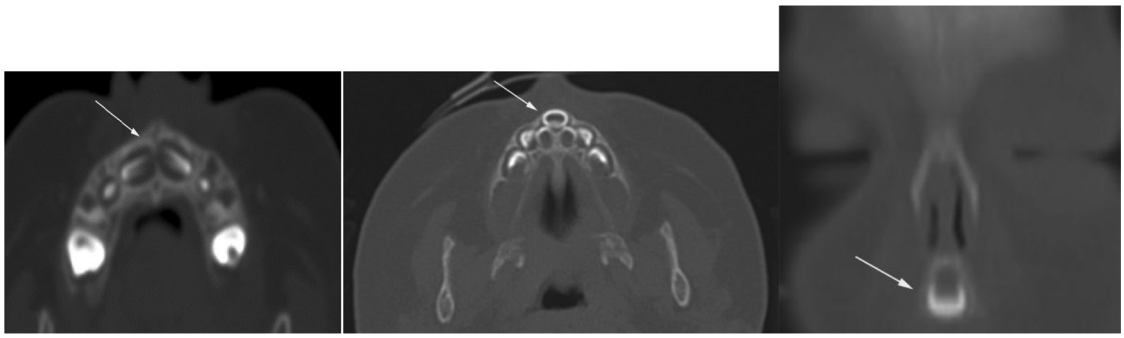

Fig. 1 – CT images through the hard palate in two 9-month-old females. An axial view of maxillary segment with normal tooth crowns is shown on the far left (arrow). In contrast with an axial view showing a solitary median maxillary central incisor (middle image, arrow). On the far right is a coronal view of a solitary median maxillary central incisor (arrow).