The Role of Computed Tomography in Diagnosis of Unilateral Congenital Choanal Atresia in a Teenager: A Case Report

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***Abstract*:** Choanal atresia is a rare malformation in newborns and infants. It could be a life threatening condition in neonates. We present a 15 year old female with history of recurrent right sided nasal discharge since birth. With Computed Tomography (CT) and CT Choanography, she was diagnosed to have unilateral (right sided) membranous choanal atresia which was treated by simple puncture transnasally with post-op stenting.

***Index Terms*:** choanal atresia, girl, computed tomography, choanography

I. INTRODUCTION

Choanal atresia is the developmental failure of the nasal cavity to communicate with the nasopharynx[1]. It is a rare malformation that causes airway obstruction in newborns and infants. It has an incidence of 1 in 5000-8000 live births. Females are more affected than males with a M: F of 1:2 and it is frequently unilateral and right sided[1]. The atresia could be bony, membranous or mixed[2]. The clinical presentation depends on whether it is unilateral or bilateral. Bilateral atresia is a life threatening condition as neonates are obligate nasal breathers and is one of the leading causes of sudden infant death syndrome. Unilateral atresia may present late with recurrent one sided nasal blockage and rhinitis[1, 3]. The diagnosis is established by computed tomography (CT) and CT choanography. This case is reported because of its rarity and late presentation and to emphasize the need for inclusion of CT in evaluating patients with recurrent nasal blockage.

II. CASE REPORT

KS is a 15 year old female student who presented to the ear, nose and throat unit (ENT) of Usmanu Danfodiyo University Teaching Hospital Sokoto with complaints of persistent right nasal blockage since birth. There was history of recurrent right nasal discharge and headache. No cough and no fever. She was treated on several occasions in a secondary health center with no satisfactory result and decided to seek medical care in tertiary health center. There was no family history of choanal atresia.

On physical examination she was fully conscious, not in obvious respiratory distress, not pale, anicteric and acyanosed. Examination of the nasal cavities showed that the right nasal cavity was filled with thick tenacious mucoid discharge. Nasogastric tube size 8 could not be passed beyond 4cm. The left nasal cavity was patent. A provisional diagnosis of suspected choanal atresia was made and she was referred to radiology department for computed tomography (CT) of the nose and paranasal sinuses.

The non contrast CT images revealed an isodense septum in the posterior nasal passage on the right extending from the medial wall of the right maxillary antrum to the vomer. No bony component was demonstrated in it. An isodense collection with air-fluid level is demonstrated anterior to the septum in keeping with the tenacious mucoid discharge seen on physical examination. There was thickening of the vomer, medial deviation of the medial wall of the right maxillary antrum and associated narrowing of the choanal air space distance on the right side measuring about 2.5mm (Fig.1). The left nasal cavity was normal. There was mucosal thickening involving both maxillary antra (Fig. 2) with fluid collection on the left antrum (Fig 3). The remaining paranasal air sinuses were clear. Images of CT choanography acquired after instillation of non ionic contrast medium into the right nasal cavity showed abrupt termination of the contrast column in the posterior choanal area with failure of the contrast to pass into the nasopharynx (Figs.2 and 3). A diagnosis of unilateral membranous choanal atresia was made.

She had nasal toileting with subsequent repair of the atresia using simple puncture transnasally with post-op stenting. Her symptoms greatly improved with no re-stenosis on follow up visits.

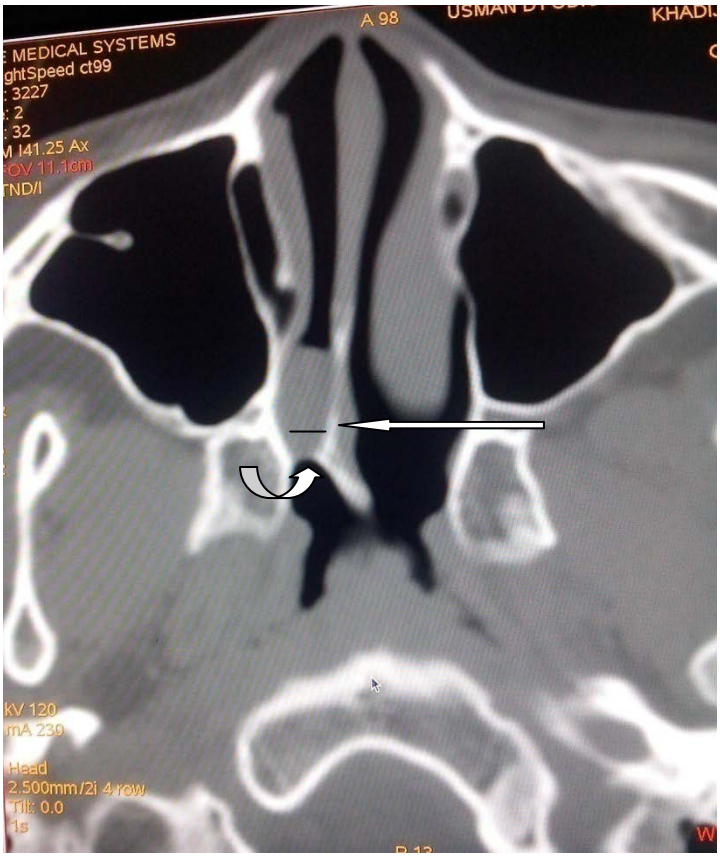

FIGURE 1: Non contrast axial CT showing an isodense septum at the posterior nasal space on the right side due to atresia (curved arrow) with collection and air fluid level anteriorly. Note the thickening of the vomer, medial deviation of the posterior right maxillary antrum and narrowing of the right posterior nasal air space (straight arrow).

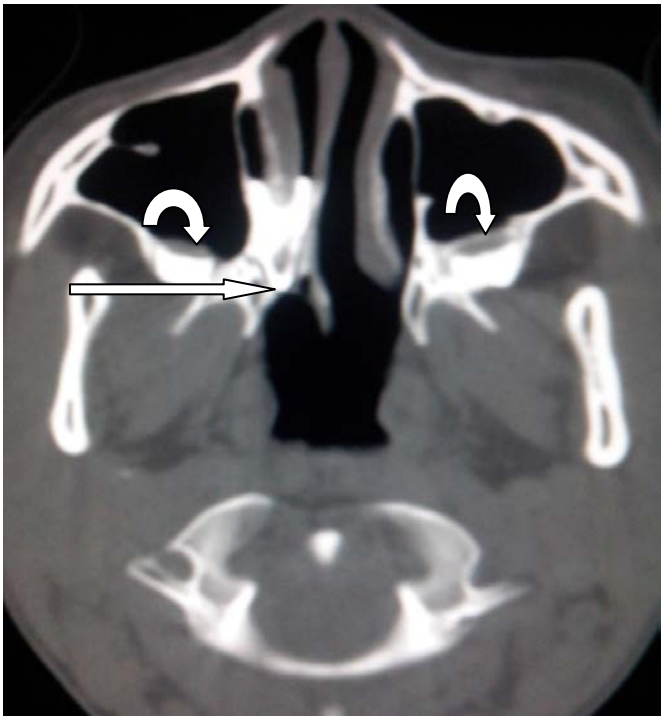

FIGURE 2: Axial CT choanogram showing termination of contrast column at the atretic site (arrow) with air-contrast level anteriorly. The filling defect is due to enlarge turbinate. Note the mucosal thickening in both maxillary antra (curved arrows).

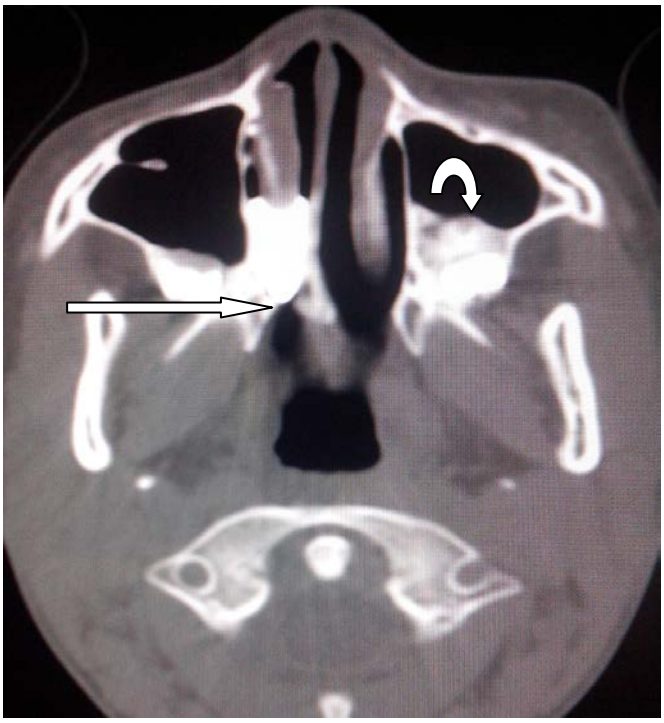

FIGURE 3: Axial CT choanogram at different level showing termination of contrast column at the atretic site (arrow) with air-contrast level anteriorly. Note the collection in the left maxillary sinus (curved arrow).

III. DISCUSSION

Choanal atresia (CA) is a rare congenital disease of the nasal airway where no connection exists between the nasal cavity and the nasopharynx. It was first described by Johann Roderer in 1755[2]. It is commoner in female and unilateral atresias are more common than bilateral[4] as seen in this patient. In more than 90% of cases of CA, the abnormality is partly or completely osseous. Pure membranous atresia is rare[4]. Isyaku et al reported a case of bilateral CA in Ibadan being membranous on the right side and bony on the left[5]. This case is unilateral (right sided) and of the rare membranous type.

The cause of choanal atresia is unknown; however classic theories of embryogenesis of posterior choanal atresia implicate persistence of either the nasobuccal membrane or failure of the buccopharyngeal membrane to recanalized[6]. Most cases of CA are isolated malformation, but association with other congenital deformities is not exceptional and has been reported in literature as in the 'CHARGE' syndrome (Colobomatous blindness, Heart diseases, Atresia of the choana, Retarted growth, Genital hyperplasia in males, Ear deformity)[2,6]. No other congenital anomalies were detected in the case here presented.

The clinical presentation of this condition depends on whether it is unilateral or bilateral. The bilateral complete type is incompatible with life as newborn human babies are obligatory nose breathers. However if it is incomplete the neonate presents with nasal obstruction and cyanosis soon after birth. Unilateral atresia may go unnoticed or patient may have recurrent one sided nasal blockage and rhinitis only to be diagnosed later in life[6]. Cases of adolescent and adult CA have been reported[4, 6]. The index case is a unilateral CA diagnosed at adolescence.

Plain radiography is an imaging method for initial radiologic assessment of patient suspected to have CA. An NG tube with a radio-opaque tip may be passed into the nasal cavity and advanced to the furthest it can go and then a radiograph (lateral) is taken. This can be done before choanography and computed tomography[5]. Conventional choanography has been the traditional method of diagnosis of choanal atresia. However CT is the current investigation of choice and when combined with choanography can differentiate stenosis from atresia. Axial scans are best for assessment of choanal atresia and may show a unilateral or bilateral posterior nasal narrowing with an obstruction. The airway is less than 3mm measured at the level of the pterygoid plates in axial plane. There may be presence of air-fluid level above the obstruction point. It may also show thickening of the vomer and medial bowing of posterior maxillary sinus[4, 5, 7]. Most of these features have been demonstrated in this case (Fig.1).

The differential diagnosis of this condition includes nasolacrimal duct mucocele, pyriform aperture stenosis, sinonasal mucoid impaction and nasal polyp. These are usually differentiated from CA based on its described imaging features[3, 8].

The treatment of membranous CA is by endoscopic perforation. Osseous atresia may require choanal reconstruction or transpalatine resection of thick vomer[6]. The success rate following endoscopic treatment was reported as 62- 100%[9]. This patient had unilateral membranous atresia and was treated successfully by endoscopic puncture with post-op stenting.

IV. CONCLUSION

Using Computed Tomography (CT) and CT Choanography, a 15 year old girl presented with history of recurrent right sided nasal discharge since birth was diagnosed to have unilateral (right sided) membranous choanal atresia. She was treated by simple puncture transnasally with post-op stenting.

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