A RARE CONGENITAL ANOMALY OF NOSE - ACCESSORY NOSE : CASE REPORT

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Abstract: We describe a 21 year old man who was found to have an opening on his external nose since birth. No other clinical and radiological abnormalities were demonstrable elsewhere in his body. The opening is on the lower part of the nose one cm. away from the midline on right side, clinically looking like a sinus. Histological examination of the excised tract showed an organoid structure that was consistent with an "Accessory nose."

Key words: accessory nose, congenital anomaly nose, supernumerary nostril.

INTRODUCTION

Accessory nose, also known as supernumerary nostril, is characterized by the congenital presence of accessory nostril usually close to the midline. The nostril appears similar to the natural ones with vibrissae. It may or may not communicate with the cavity of the nose. It is usually associated with other congenital malformations like submucous cleft palate, incomplete or complete cleft palate or cleft lip, Proboscis lateralis etc. It is believed to be a rare congenital anomaly, occurring during the development of nose in 3rd or 4th week of embryonic period. Adequate knowledge and description of the etiology, pathogenesis and management is lacking in the world literature due to the rarity of the anomaly.

CASE REPORT:

A 21 year old male presented to the out patient department of our institute with the complaint of presence of an opening on his nose situated 1 cm. lateral to the midline in the lower half of the nose on right side associated with the history of recurrent mucopurulent discharge from the opening (Fig.1). There is no history of bleeding or continuous watery discharge from the opening. Patient's birth history revealed that he was delivered by full term normal vaginal delivery at home. There was no history of consanguinity. No significant finding is present in the maternal history. There is no similar or significant illness or congenital anomalies in the family. He is a non-smoker and non alcoholic.

He has no other significant illness and is fruit vendor by profession. On examination the opening was present on the lower half the nose, approximately 1 cm. away from the midline. No active bleeding or discharge was noted. There was no other mass or swelling on the nose or anywhere else in the body. Prominent viable hairs noticed near the sinus opening. Skin around the opening is normal. On palpation there was no tenderness. No mass was palpable. Anterior rhinoscopy and posterior rhinoscopy revealed no abnormality. Transillumination through the sinus opening did not reveal continuity of the sinus into the nasal cavity. On probing with Jobson Horne probe the sinus tract was found to end blindly 2cm superiorly from the opening.

Diagnostic nasal endoscopy was done which revealed no abnormality. The scope could not be passed in the sinus opening due to smaller size of the opening. Sinogram was performed to know the extent of the tract, which showed the tract ending blindly in the soft tissues of the nose, without any communication to the intracranial structures (Fig.3).

Surgery was undertaken to excise the tract and correct the anomaly (Fig.2), after explaining the patient, the course, prognosis and complications. A circular incision was given around the opening and tract dissected out and sent for histopathological examination, which revealed well formed organoid structure consistent with accessory nose.

Fig.1: Patient with the accessory nose

Fig.2: Intraoperative picture of the tract

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Fig.3: Sinogram showing the extent of the tract

Fig.4: Post operative picture

DISCUSSION

Embryologically, by week 4 of intrauterine life, precursors of nasal cavities and the lenses of the eyes are formed. The nasal placodes (olfactory placodes) arise from the medial aspect of the lower portion of the frontal prominence, and the lens placodes arise from the lateral aspect of the lower portion of the frontal prominence. At week 5, the mesenchyme, which covers the caudal surface of the forebrain, proliferates with the surface ectoderm to form the frontonasal process. Two ectodermal thickenings (the nasal placodes) arise on each side of the dependent part of the frontonasal process. Subsequently, a depression develops in the surrounding mesenchyme on each side of the two nasal placodes, thereby forming two olfactory pits or gutters. These olfactory pits separate the frontonasal process into a medial nasal process and two lateral nasal processes. The lateral nasal processes subsequently form the alae of the nose. As the olfactory pits progress deeper to form the primitive nasal cavity, the medial nasal process thins out gradually to form the primitive nasal septum. At this stage, the maxillary process develops from the cephalic side of the dorsal part of the mandibular arch. Each maxillary process grows ventromedially to meet and fuse with the lateral nasal processes, and each ultimately fuses with the medial nasal process, thus forming the external opening of the primitive nasal cavity and the upper lip.

Supernumerary nostrils are exceedingly rare congenital anomalies of unclear etiology. In 1962, Erich⁵ reported a case of double nose. He also supported Lindsay's⁴ theory of dichotomy by atavism or parallel evolution, and he further speculated that if the accessory nasal pit is located too laterally, the fusion of the lamina is not affected, which leads to the formation of a supernumerary nostril. In 1972, Onizuka and Tai⁶ reported the case of a single accessory nostril that had developed above the nasal ala. In 1987, Nakamura and Onizuka⁹ reported a similar case, and they hypothesized that the cause was probably a localized defect in the lateral nasal process. In 1992, Chen and Yeong⁷ described a case of bilateral supernumerary nostrils that were situated below the normal nasal openings, and they proposed treating such anomalies by staged corrective surgery. Another case has been reported by Kurul.S³ et al, in 1995, where a 6 year old Turkish girl presented with accessory nose and incomplete cleft palate in 2001, Hallak et al⁸ reported a case of supernumerary nostril in which a

blind cavity was present in a normally developed nose. They advocated that corrective surgery be performed at an early age to prevent any possible alar deformity. Recently, in 2004, G.Cuervo De Lacalle et al¹⁰ reported a case of supernumerary nostril with ear deformity.

Most reported cases of supernumerary nostrils have been unilateral, and most were associated with other craniofacial malformations, such as a facial cleft¹⁰. A supernumerary nostril may or may not communicate with the ipsilateral normal nasal cavity, depending on the extent of the anomaly's embryologic progression. Dermoid cyst with sinus can be considered in the differential diagnosis of the accessory nose, but dermoid is usually in the midline, and presents as mass on the nose with small punctum containing hair follicle, and it communicates with intracranial structures. Other midline congenital masses like encephalocele, meningocele, meningocele are not relevant to the case reported here.

To best of our knowledge, this is the first case report of isolated anomaly of accessory nostril, in this part of the world.

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Clinical Report**A CASE STUDY OF PLUMMER-VINSON SYNDROME**

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Abstract: The case series presents the pattern of association of different co-morbid conditions of Plummer Vinson's Syndrome. The commonest morbidity is web at post cricoid level. The symptoms of dysphagia are more common and there appears to be a female preponderance. The presenting age is usually 5th decade. Although association with post cricoid carcinoma has been reported but only 10% of cases in the present series had carcinoma at the time of presentation. The effect of iron and B-complex supplementation has been clinically appraised and good symptomatic relief has been seen. Maybe this measure can prevent the progression to malignancy. Bouginage was effective in relieving symptoms in all the cases it was tried

Keywords : Plummer Vinson Syndrome, Postcricoid web.

INTRODUCTION

Plummer-Vinson syndrome or Patterson-Kelly syndrome is characterized by dysphagia which is usually due to upper esophageal web, anemia, splenomegaly and other features like angular chelitis and glossitis. A prospective study was conducted in BJ medical college, Ahmedabad with an aim to find out the age and sex incidence of this syndrome. To evaluate the results of iron and B-complex supplementation with esophageal dilation and find out relationship with post-cricoid carcinoma, if any.

MATERIALS AND METHODS

25 confirmed cases of Plummer-Vinson syndrome were included in the study after taking detail history and detail examination, all cases were investigated for all routine blood parameters including blood indices and radiological investigation barium swallow and other indicated investigations. The patients are subsequently put on iron and B-complex therapy. Rigid endoscopy was done, webs and strictures were dilated and biopsy of suspicious lesions was taken. The patients were adequately followed up and any needed further management was instituted.

OBSERVATIONS

Out of 25 cases of the present series, 22 patients i.e. 88% were females and the rest were males. The age distribution is as shown in table A. 12 patients were in the 5th decade of life. Next highest was 7 cases in 4th decade of life. The remaining cases were sparsely distributed over the other decades. The youngest was 20 year old and the oldest was 65-year old. 2 patients had a past history of oesophagoscopy and dilatation, whereas 2 patients had a history of surgery for acid peptic disease. The symptomatology is as shown in Table B. All cases had dysphagia

other symptoms like soreness of mouth, easy fatigability, palpitation, parasthesia, giddiness, dyspnea and malena were present in varying proportions.

Grade I dysphagia where in patient has only difficulty in swallowing solids was present in 52% cases. Grade II dysphagia where in patient can take only semisolids was present in 36% patients. Grade III dysphagia where in patient can take only liquids was present in 8% cases. Difficulty to swallow even saliva i.e. grade IV was present in 4% cases. Among clinical features signs of iron deficiency, anemia like bald tongue, pallor, koilonychia were present in all cases. Angular chelitis and glossitis was present in 80% and 60% cases respectively. Splenomegaly was found only in 4 cases.

All patients had hemoglobin less than 10gm% and 8 cases were severely anemic with hemoglobin less than 5gm%. Blood indices like PCV, MCV, MCHC, serum iron and total iron binding capacity revealed microcytic hypochromic anemia with iron deficiency in all cases. Urine analysis was normal in all cases. Stool examination revealed occult blood in 2 cases. Gastric analysis revealed achlorohydria in 3 cases. Radiological investigation revealed post cricoid web in 4 cases, benign stricture in 1 case, malignant stricture in 2 cases & surprising normal finding in 12 cases. All cases in spite of barium swallow findings underwent rigid oesophagoscopy under general anesthesia which showed post cricoid web in 15 cases. These webs were commonly situated on lateral wall. Stricture was present in 4 cases. Exophytic growth was seen in 2 cases which proved to be malignancy on biopsy and histo-pathological examination during endoscopy full dilation with gum elastic bougies was carried out in 19 cases. All patients except those with malignant growth were started on full diet next day itself iron and vitamin supplements was instituted in all cases.

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