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Case Report

Hemiarhinia – A Developmental Disorder - A Case Report

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Abstract

Heminasal aplasia or hemiarhinia is an extremely rare congenital malformation with the exact mechanism or etiology of its development still unknown. The rarity of this anomaly makes its reconstruction a surgical challenge with a diversity and controversy on the timing and technique of such reconstruction. We report a new case of hemiarhinia which to the best of our knowledge is the 71st reported case of congenital defect. The case is a female with absence of the right side of the nose. The clinical and the CT scan manifestations of a case of right-sided hemiarhinia together with the reconstructive plan of it are presented. We recommend early soft-tissue reconstruction using adjacent nasolabial flap without cartilage grafts to provide temporary correction of the disfigurement until the definitive reconstruction can be undertaken.

Keywords: Hemiarhinia – a rare congenital malformation.

Introduction

Nasal hypoplasia ranging from underdevelopment or partial absence of parts to complete arhinia is the most frequently seen nasal anomalies [1]. Heminasal aplasia, hemiarhinia or unilateral aplasia of the nose is a rare congenital malformation in which there is absence of half of the external nose together with a variable degree of abnormality in the internal anatomy of the nose as well as the adjacent facial structures. It imposes a major psychological burden to the parents and may have physiological impact on the patient.

The nose develops from the mesodermal frontonasal process and the two nasal placodes. The frontonasal process appears in the third to fourth week of gestation together with two bilateral ectodermal thickenings known as nasal placodes that grow caudally. During the fifth week of gestation, a central invagination, called the nasal pit, divides each nasal placode into a medial and a lateral nasal process. The nasal pits extend posteriorly to form the nasal cavity, which is separated from the oral cavity by a thin nasobuccal membrane. The nasobuccal membrane eventually ruptures at week 6 to form the posterior choanae. The epithelium around the forebrain thickens to become specialized olfactory sensory cells. The medial nasal processes from both sides fuse, forming the nasal septum and philtrum while the lateral processes develop into the external wall of the nose, the nasal bones, the upper lateral cartilages, the alae, and the lateral

crura of the lower lateral cartilages. The failure of the development of nasal placodes probably leads to the congenital absence of nose [2]. Although the exact mechanism is unknown, several theories for the pathogenesis of arhinia were hypothesized. These theories include 1) failure of the medial and lateral nasal processes to grow, 2) premature fusion of the medial nasal processes, 3) lack of resorption of the nasal epithelial plug, and 4) abnormal migration of the neural crest cells [3,4]. Congenital arhinia may be in part induced by Chromosomal aberrations as some chromosomal change has been reported in several cases. The genetic analysis of five patients with complete arhinia identified a 19 Mb large deletion involving 3q11–q13 in one patient of them [5]. Another patient had and a translocation between chromosomes 3 and 12 [6]. In another report, one case was found to associate with inversion of chromosome 9 and another had mosaic of chromosome 9 [7].

Failure of the development of both nasal placodes results in complete nasal aplasia or arhinia while failure of one placode leads to heminasal aplasia or hemiarhinia [8]. Nasal anomalies rarely occur alone and are frequently associated with other coexistent craniofacial anomalies. They were classified into two major groups, total arhinia, with absence of the nose and both olfactory nerves and partial arhinia with presence of at least one nostril and one olfactory tract. Both groups can be seen withor without other craniofacial malformations [7]. Partial arhinia includes all the range from hypoplasia or absence of only an individual structure to a complete absence of the heminose. Few cases were reported with congenital absence of the columella, with the medial crura of the lower lateral cartilages and their soft-tissue covering were missing, while the remaining septum and other nasal structures were normal [9,10]. Isolated nasal bone agenesis or hypoplasia has also been reported [11,12].

Case presentation

A female (picture 1) presented to us at the age of fourteen with the history of absence of right nostril, swelling over the medial canthus of right eye with watering. The girl was the outcome of full-term normal delivery. She was 2200 grams atbirth. The mother didn't take any medicines during pregnancy, no consanguinity between parents and no such abnormality was found in either of her parents' families. The girl was delivered in a primary health care unit in a village. No history of breathing or suckling problem was reported by the parents who were concerned only by the abnormally disfigured nose and swelling over the medial canthus of right eye. On examination, there was complete absence of the right side of the external nose and right nostril, while the upper bony third was looking normal. There was a soft, cystic welling below the medial canthus of right eye which sometimes bursts and mucopurulent material comes out (according to patient party).There was hypertelorism in the right side. The girl was breathing normally with no respiratory distress or cyanosis. Other examination of the girl did not reveal any other abnormality; neither did the abdominal ultrasonography nor the echocardiography. Blood picture and liver functions were within normal ranges. The anomaly was explained to the parents.

Figure.1 Normal and symmetric nasal bones on both sides

CT scan findings (picture 2, 3, 4and 5) are:

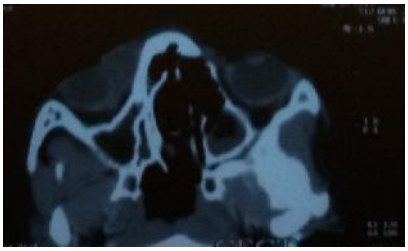

Figure. 2 Single nasal cavity with the nasal septum markedly deviated towards this same side

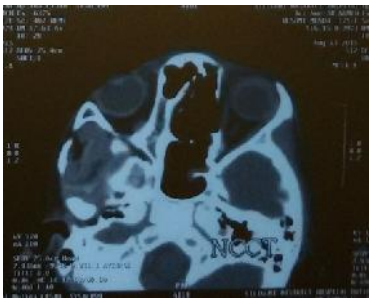

Figure. 3 Well developed ethmoidal, frontal and sphenoid sinuses

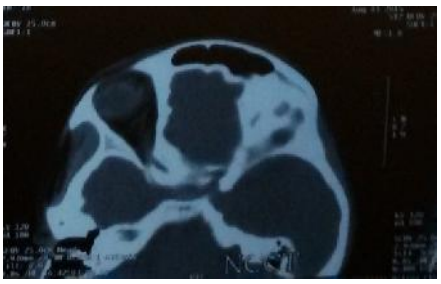

Figure. 4 No orbital abnormality

Figure. 5 Normal nasopharynx and oropharynx

Discussion

To the best of our knowledge, this is the 71st case of hemiarhinia reported worldwide. The previously reported cases of hemiarhinia show strikingly similar features with most of them having concomitant ipsilateral proboscis lateralis [13]. In our case, the patient was a female and the aplastic half of the nose was on the right side, the parents were not relatives and they declined any family history of similar anomaly. There was no proboscis and the only associated extranasal anomaly was the small swelling over the medial canthus of right eye and slight hypertelorism.

The management of arhinia is a surgical challenge and because of its rarity, there is still a controversy concerning the timing as well as the technique of such reconstructive procedures. The ability of the patient to breathe, the adequacy of the air passages and the cosmetic appearance are the main factors determining the timing of surgery. Early surgical intervention in childhood is thought to be better, both from the cosmetic and psychological point of view [14]. Onizuka et al. began their constructive surgery in a case of arhinia at the age of 6 months, and the reconstruction was complete at the age of 18 months [15]. They recommended that operative correction of arhinia to be undertaken as early as possible to achieve a better morphology as long as there is no cerebral anomaly. Muhlbauer et al. started their reconstruction of both the internal and external nose at the ages of 4 months and the reconstruction was completed at the age of 26 months [16]. On the other hand, many authors agree that surgical reconstruction of the nose and its internal cavities should be delayed at least until preschool years [7,17,18,19,20]. Others postpone the nasal reconstruction till the age of 15 years when facial development is nearly complete [21]. We believe that early surgical correction is better particularly in patient with an isolated nasal anomaly.

Conclusion

Hemiarhinia is an extremely rare congenital malformation. A female presented at the age of fourteen with the history of absence of right nostril swelling over the medial canthus of right eye with watering. On clinical examination the right nostril was found absent. There was no breathing problem. There was no other craniofacial anomaly. The surgical indication is only for cosmetic reason. The cosmetic surgery should be done earliest possible for better outcome.

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