CASE REPORT

Hemiarhinia – A Developemental Disorder - A Case Report

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ABSTRACT

Introduction: Aplasia of one half of external nose with nasal cavity is termed as hemiarhinia. This is an extremely rare developmental disorder. The main etiology or mechanism of this congenital disorder is a controversy. The timing and technique of the reconstructive surgical management still create a technical dilemma among the surgeons.

Case report: A female patient presented at our institution with the history of absence of right nostril, swelling over the medial canthus of right eye with watering at the age of fourteen. On clinical examination it was found that the right sided nostril was not developed and there was a single nasal cavity. The breathing was normal. On CT scan there was no other craniofacial anomaly was detected.

Conclusions; The indication of surgery is only for cosmetic reason. Early intervention is necessary for better outcome. Soft-tissue reconstruction using adjacent nasolabial flap without cartilage grafts to provide temporary correction of the disfigurement.

Keywords: Hemiarhinia – a rare congenital malformation, proboscis lateralis, neural crest cell.

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INTRODUCTION

Hemiarhinia or heminasal aplasia is a rare congenital anomaly of nose. This developmental disorder is of two types, partial absence of any parts of nasal cavity with external nose due to underdevelopment and complete absence or arhinia;[1] Of these, unilateral aplasia of nose is the rarest. The heminasal aplasia or arhinia may remain associated with different types of developmental anomaly of the internal anatomy of nasal structure with or without deformity of facial structures. It imposes a major psychological burden to the parents and may have physiological impact on the patient.

The nose develops from a number of mesenchymal processes around the primitive mouth. The nasal cavity is first recognizable in the fourth intrauterine week as the olfactory or nasal placode which are, a pair of ectodermal thickening on the frontonasal process. In the fifth week, the ectoderm in the center of each nasal placode invaginates to form a oval nasal pit. The raised rims of these nasal pits form the lateral and medial nasal processes. The nasal pits extend posteriorly to form the nasal cavity, which is separated from the oral cavity by a thin nasobuccal membrane. This membrane ruptures during the end of sixth week to form the primitive choana.During the sixth and seventh weeks, the maxillary process on either side increase in size and grow medially and fuse with each other to form the intermaxillary process. The intermaxillary process forms the nasal septum and philtrum. The lateral nasal process enlarge to form the nasal alae. They also grow backwards to form the lateral nasal wall. This developing lateral nasal wall shows multiple anteroposterior elevations which form turbinates and corresponding meati.The failure of the development of nasal placodes probably leads to the congenital absence of nose;[2] The etiology of arhinia is not well known. There are several hypothesis regarding the mechanism of this developmental disorder. They are 1) failure of development of medial and lateral nasal processes, 2) early fusion of median nasal processes 3) developmental arrest of nasal placodes 4) abnormality of neural crest cell migration;[3,4] A single nostril results when only one nasal placode forms. The genetical basis of congenital arhinia may be the chromosomal aberrations. There are several cases of chromosomal aberrations already reported, of

which one patient identified with a 19Mb large deletion involving 3q11-q13;[5] Complete nasal aplasia or arhinia results from absence of development of nasal placodes, whereas hemiarhinia results from absence of development of one nasal placode;[6] These developemental anomalies of nose may be frequently associated with other craniofacial anomalies. Complete absence of nose or arhinia usually associated with absence of both olfactory nerves and partial arhinia leads to hypoplasia or absence of atleast one nostril and one olfactory nerve. There were also some cases reported with congenital absence of columella but septum and other nasal structure remained normal;[7]

CASE PRESENTATION

A female (Figure-1) presented in the out patient door of Otorhinolaryngology department of North Bengal Medical College 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 at birth. 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 swelling 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. CT scan findings are shown in figure-2,3,4,5. Figure-1 is the clinical photograph with one nostril (right nostril absent), figure-2 shows single nasal cavity with the nasal septum markedly deviated towards right side, figure-3 shows well developed ethmoidal sinuses and normal orbit, figure-4 shows normal frontal sinus, figure-5 shows normal sphenoidal sinus.

Figure-1: Clinical photograph with one nostril (right nostril absent)

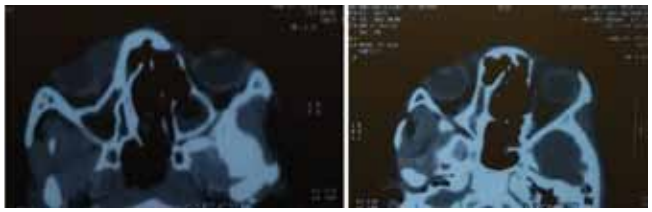

Figure-2: Shows single nasal cavity with the nasal septum markedly deviated towards right side; **Figure-3:** Shows well developed ethmoidal sinuses and normal orbit

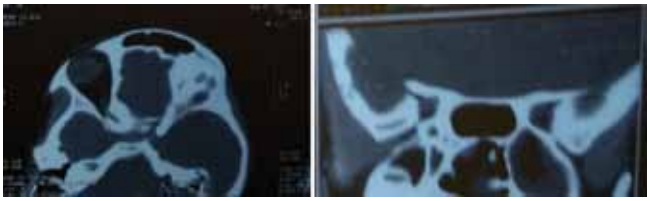

Figure-4: Shows normal frontal sinus; **Figure-5:** shows normal sphenoidal sinus.

DISCUSSION

To the best of our knowledge, this is the 71st case of hemiarhinia reported worldwide. The previously reported cases of hemiarhiniashow strikingly similar features with most of them having concomitant ipsilateral proboscis lateralis;[8] 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 hemiarhinia is based on mainly reconstructive surgery. But the main challenging thing is the timing of surgery and the technique of the surgery. The timing of the surgery depends on the age of presentation, respiratory status of the patient and the cosmetic appearance of the patient after the surgery. Previous study suggests the early surgical intervention is better for cosmetic and psychological value;[9] Oni-

zuka et al showed in their study that the early correction of absence of either partial or complete absence of nostril is better for morphological point of view, if it is not associated with cerebral anomaly;[10] We also recommend reconstructive surgery for our patient to alleviate the psychological stress by improving her cosmetic appearance of face.

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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