Rare Bifid Nose

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Abstract
55 years old poor male farmer in Varanasi visited my hospital who had complete bifid nose right up to the root of nose and the right side is freely hanging and only attached at the root of the nose. Hanging nose is presented with little congenital midfacial disfigurement with hypertelorism . He had no siblings with clefts. He had achieved expected intellectual and physical milestones commensurate with his old age. The physical examination did not revealed any facial cleft with almost normal calvarial and lower facial structures. The patient had a wide nasal radix and right free hanging nasal part with its nare and other part (left portion of nose) is firmly attached to face. The hanging nasal part was bigger in size. There is increased width of nasal radix. The phitral dimple was normal and phitral is columned.

Keywords: Bifid nose, Tessier classification, Aesthetic reconstruction, Hanging nasal nare.

Introduction
Bifid nose is usually a rare condition where the people are born with defective nose and vary in severity among the affected individuals where the nose is divided into two parts to different extent. In some it can hardly be noticeable with just a groove at the nose tip while the severe condition can affect the underlying bones and cartilage resulting in the complete division of the nose. The bifid nose may be the only birth defect presented while in others may have several abnormalities that may affect the growth and function of different parts of the body. That is they can also have genetic defects, which are known as the BNAR syndrome, in which, the rectum, anus, and kidney, have various defects. It may be an inherited condition in some cases or may develop for the first time with no record of its prevalence in the family. They can also have a cleft lip as well as a palate along with this. It has also been observed that 8% of people who have bifid nose are known to have numerous other facial abnormalities that include eyes that are widely spaced (hypertelorism).

Diagnosis
Bifid nose may usually be detected during an examination conducted by a physician. In some cases, the nose abnormality may be mild that appear after adulthood while in other cases, the condition may be evident at birth. It is a rare congenital bifid nose disease which occurs in 4 in 100,000 births. Usually, if the bifid nose occurs with other abnormalities or birth defects, a molecular genetic testing can detect the mutated gene responsible for the cause. A 55 years old farmer man from very poor socioeconomic class from a village in Varanasi, India presented to our hospital for other ailment but was diagnosed for this also by me but inspite of free treatment offered, he refused treatment as he had no other symptomatic problem except the aesthetic reason which was no concern for him. He had complete bifid nose right to the root radix of nose and the right side was only attached at root of nose and freely hanging. Patient refused even the free CT Scan as he feared any treatment like reconstructive surgery to be prescribed by us. Hanging nose presented with little congenital midfacial disfigurement with hypertelorism. He had no siblings with clefts. He had achieved expected intellectual and physical milestones commensurate with his old age. The physical examination did not revealed any facial cleft. He had apparently normal calvarial and lower facial structures. The patient had a wide nasal radix and right free hanging nasal part with its nare. The other part of nose is firmly attached to face. The hanging nasal part was bigger in size. The nares were separate. The phitral dimple was normal and phitral is columned.

Fig. 1: Bifid nose with hanging right part.

Fig. 2: Bifid nose

Discussion

Midline facial clefts are rare deformities with a wide range of clinical findings from a simple midline vermillion notch to major skeletal malformations, including orbital hypertelorism. The bifid nose is a relatively uncommon malformation that is frequently associated with hypertelorbitism and midline clefts of the lip. The presentation of a bifid nose ranges from a minimally noticeable midline nasal tip central groove to a complete clefting of the osteocartilaginous framework, resulting in two complete half noses. Craniofacial clefts cause severe facial disfigurement even in minor forms. The surgical reconstruction is imperative to restore function and appearance of facial structures. Orofacial cleft is a failure in embryonic facial development during the first 8 weeks of life. The cause of craniofacial clefts is not clear. However, some theories about orofacial cleft have been submitted such as failure of fusion theory and the failure of mesodermal penetration theory. Various risk factors (radiation, infection, maternal metabolic imbalances, and drugs and chemicals) have been published. Craniofacial clefts were classified by Tessier, according to their anatomical basis. This classification system contains numbered clefts from 0 (midline cleft of the lip and nose) to 14 and 30 (mandibular). Presentation of median cleft may be varied from minimal changes on median facial structures such as lip, vermilion, and nose to wide clefts dividing all median craniofacial structures. The median craniofacial skeleton (crista galli, ethmoid, vomer, nasal, and premaxillary bones) and the cartilaginous septum can be affected in severe clinical presentation of patients. This has been almost always enough for a diagnosis of the craniofacial clefts and computed tomography is of great help. Tessier number: 0-cleft may cause death if it is associated with holoprosencephaly. CT scans help the surgeon to plan surgery. Minimal malformations on median facial structures can be corrected with local flap options such as z-plasties and v-y advancement flaps. However, severe cases require reconstruction of midline of the craniofacial skeleton. Surgical correction of bifid nose can be challenging and requires multiple surgical procedures. A few articles related to surgical correction of this unusual malformation have been published.

Treatment

The bifid nose can be extremely disheartening for the patients but with a proper treatment like molecular genetic testing and reconstruction surgery, doctors can easily get rid of it.

Source of funding

None.

Conflict of interest

None.

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