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CASE REPORT

BINDER'S SYNDROME: A CASE REPORT

Rohit Mehrotra

Department of Oral pathology and Microbiology

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ABSTRACT

Binder's syndrome or nasomaxillary hypoplasia or maxillonasal dysplasia is an uncommon developmental disorder predominantly affecting the maxillary anterior region and the nasal complex. A concave profile, broad and flat nose, short columella, convex upper lip, and malocclusion are the major clinical findings. The characteristic features of this syndrome are developmental failure in the premaxillary area, with concomitant defects of the nasal skeleton. The current article presents case report of 16yr old patient of binder's syndrome describing its features, surgical intervention and postop results.

INTRODUCTION

Binder's syndrome or maxillonasal dysplasia or maxillonasal dysostosis is an uncommon developmental disorder predominantly affecting the maxillary anterior region and nasal complex. Arhinoid face, abnormal position of the nasal bones, reduced or absent anterior nasal spine, atrophy of the nasal mucosa, intermaxillary hypoplasia, with associated malocclusion, and absence of the frontal sinus (not mandatory). Individuals with Binder's syndrome have a characteristic appearance, which shows an underdeveloped midface (midfacial hypoplasia), with an abnormally short nose and flat nasal bridge, underdeveloped upper jaw, and a relatively protruding lower jaw.

Case Study

A 16 year old patient presented to our hospital with chief complaints of nasal obstruction since past three months. Clinical examination revealed deviated nasal septum right side with depressed dorsum of nose and nasal tip with depressed maxilla. Diagnostic nasal endoscopy and NCCT nose and PNS were done which confirmed the same along with inferior turbinate hypertrophy on both sides.

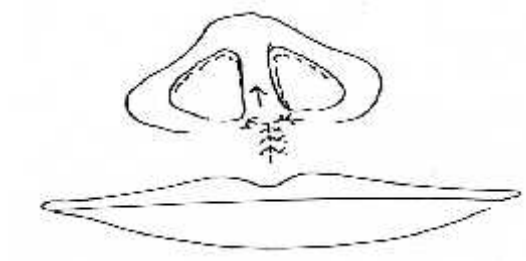
After thorough preliminary blood investigations and pre anaesthetic checkup (PAC), we planned for reconstructive surgery under general anaesthesia.

Extracorporeal Rhinoplasty With Rib Cartilage Interposition At Maxilla With Screw And Cartilage At Nasal Dorsum And Tip Was Performed Under General Anaesthesia.

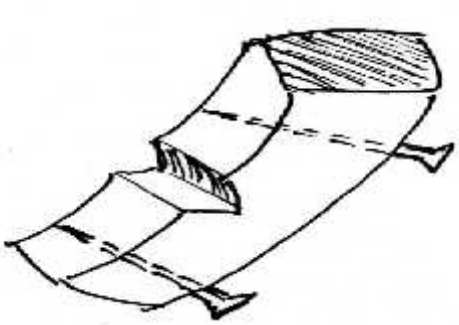
Diagrammatic Pictures of Operative Assembly

Incision Line

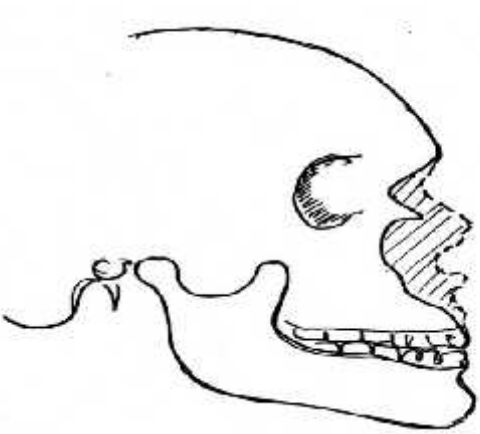
V Y Plasty



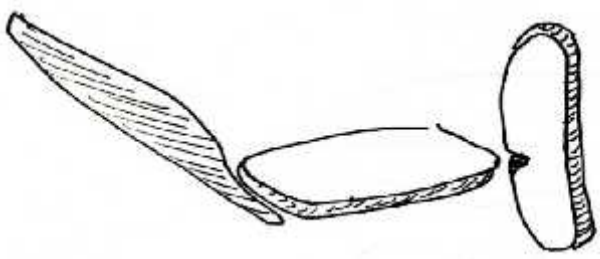
Costal Cartilage With Notch For Nasal Spine



Deficient Pre Maxilla



Nasal Cartilage Assembly



Two major problem in nasal deformity correction in Binder's syndrome is Premaxilla depression and very short columella. We need large amount of graft material which can only be found from costal cartilage. This cartilage is kept in saline otherwise it tends to bend. Premaxilla is augmented by keeping a large oval piece with a depression created in midline so that it is fixed over nasal spine with screws, through sublabial incision. Nasal floor mucosa is elevated and septum is chiseled out from the floor to separate nasal structures from floor so that it can be augmented easily. Columella lengthening is done by large V-Y plasty at the expense of midline lip skin.

Postoperatively patient was given injectables of antibiotics and pain killers along with long acting steroids and anti allergics. Patients was discharged on day 2 post op with oral antibiotics, steroids and intra nasal sprays. Patient was followed up after three days and then weekly basis. In follow up suction and cleaning of nose was done, all crusts were removed and patency was maintained. X ray nose and PNS waters view was done and revealed intact screws in maxillary region.

Patient symptom of nasal obstruction was resolved and no fresh complaints were present. Patient nasal tip and dorsum was fine and well elevated and maintained compared to pre op state.

DISCUSSION

The craniofacial deformity with hypoplasia of middle third of face associated with congenital absence of anterior nasal spine and depression of nasal bones with flattened nasal alae has been described as **Binder's syndrome** or Maxillo-nasal dysplasia. The use of terms syndrome and dysplasia is misnomer as has been discussed by Quarrell *et al*.[1] because of varied clinical presentation and because of absence of any reported hisopathological description.

Binder's Syndrome/Binder Syndrome (Maxillo-Nasal Dysplasia) is a developmental disorder primarily affecting the anterior part of the maxilla and nasal complex (nose and jaw). It is a rare disorder and the causes are unclear. Hereditary and vitamin D deficiency during embryonic growth have been researched as possible causes.[2]

The characteristics of the syndrome are typically visible. The syndrome involves hypoplasia of variable severity of cartilaginous nasal septum and premaxilla. It includes complete total absence of the anterior nasal spine. There are also associated anomalies of muscle insertions of the upper lip and the nasal floor and of the cervical spine. Affected individuals typically have an unusually flat, underdeveloped midface (midfacial hypoplasia), with an abnormally short nose and flat nasal bridge. They have an underdeveloped upper jaw, relatively protruding lower jaw with anterior mandibular vertical excess and a Class III skeletal and dental (reverse overjet) profile. They have a small frontal sinus and global facial imbalance.

Hopkin, in 1963, reported five cases, and he proposed that Binder's syndrome was the result of a developmental abnormality. [3] In 1987, Narcy *et al*. reported a case of Binder's syndrome in association with esophageal achalasia and abnormal autonomic reflexes, suggesting that the phenotype could be the result of an abnormal migration of neural crest cells. [4],[5] There was evidence that vitamin K-deficiency, during human pregnancy, caused by some chemicals such as lithium and ethanol or the therapeutic use of warfarin or phenytoin, could be a causative factor. Pregnancy histories of three cases of Binder's syndrome were reported earlier, associated with warfarin and phenytoin exposure and alcohol abuse. [6] Individuals with Binder's syndrome have a characteristic facial appearance that is easily recognizable. Binder's syndrome is characterized by the absence of the nasofrontal angle with a hypoplastic nose, with a small tip and mild hypertelorism.

Although, Binder had suggested the defect to be of archinecephalic origin, along with disturbance of prosencephalic induction center during embryonic growth.[8,10] Noyes[7] suggested birth trauma to be one of the causative factors though in five cases reported by Hopkin[9] none had any birth trauma. Ferguson and Thompson [12] had thought of genetic etiology, which was also supported to some extent by others. It was suggested that inheritance could be autosomal recessive with reduced penetrance or it could be multifactorial in heritance. Gorlin *et al*. [11] finally emphasized that Binder's syndrome is a non-specific abnormality of nasomaxillary complex and familial examples are a result of complex genetic factors. The syndrome as reported by Binder [8] had few characteristic features such as arhinoid face, abnormal position of nasal bones, inter-maxillary hypoplasia with associated malocclusion, reduced or

absent anterior nasal spine, atrophy of the nasal mucosa and absence of frontal sinus.

Treatment is encouraged as early as possible with posteroanterior traction on the maxilla and, at about age 8, reinsertion of the nasolabial muscles onto the anterior border of the cartilaginous system. Many who have a severe case of the disorder undergo plastic surgery or orthodontic treatment for cosmetic reasons. The treatment of this complex maxilla nasal deformity would mainly require orthodontic and surgical intervention after assessing the degree of this complex maxilla-nasal deformity. LeFort I and II osteotomy with nasal grafting can be carried out for severe cases.

CONCLUSION

Despite the fact that malocclusion is closely related to maxillo-nasal dysplasia traditionally patients with this condition have been treated by plastic surgeons alone. Nevertheless, the presence of a variety of dental and facial deformities necessitates an inter-disciplinary approach for management of these cases, along with a proper treatment planning

Conflict of Interest*:* Since there are no funding issues all three authors declares that there is no conflict of interest.

Ethical Approval*:* Procedure performed in the study involving human participants were in accordance with the ethical standards of the institution and with the 1964 Helsinki declaration and its later ammendments or comparable ethical standards.

Informed Consent*:* informed consent was obtained from the patient included in the study.

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