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Binder's Syndrome – An Unusual Craniofacial Anomaly

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CASE REPORT

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ABSTRACT

Binder's syndrome or maxillonasal dysostosis is a rare congenital condition that primarily affects the mid-face and sometimes the vertebrae. It was named after von Binder who described three cases of hypoplastic maxilla-nasal complex in 1962. It can either occur as a sporadic mutation or may be inherited in an autosomal recessive pattern with incomplete penetrance. Decrease in the naso-labial angle, flat forehead, dish-shaped face, absence of protrusion of nasal tip, absence of nasal flare with triangular or semilunar nostrils, palpable depression in the nasal floor and a class III tendency are characteristic of the syndrome. Vertebral anomalies are seen in some patients owing to the parallel development of the nasal complex and vertebrae in the third month of intrauterine life. Prenatal diagnosis may be done using ultrasonography at 21 weeks of pregnancy. A multi-disciplinary approach towards planning of treatment for individuals with Binder's syndrome includes orthodontic treatment along with osteotomies and grafting to correct the nasal and mid-face defects.

Key words: congenital anomaly, dish face, maxillonasal dysostosis

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INTRODUCTION

Binder's syndrome is a rare genetic condition that is characterised by under-development of the middle third of the face leading to hypoplasia of the nasomaxillary complex.¹ Zuckerkandl in 1882 first described the abnormality of the anterior nasal floor where the anterior nasal spine was replaced by the fossa prenasalis which constituted the inferior margin of the piriform aperture. Later Noyes in 1939 noted a case with flat nasal tip sitting on a retruded maxillonasal base. But it was only in 1962 that it was identified as a separate syndrome after K. H von Binder described three similar cases in unrelated children showing features of maxillonasal dysostosis.^{2,3} Recently in 1997 Posnick and Thompson noted that the syndrome was a result of hypoplasia of fossa prenasalis and a symmetric localised hypoplasia of the maxillary alar ridge region.⁴

The syndrome is known to occur in less than one in 10,000 infants with equal gender predilection, although the estimated prevalence may be underemphasized due to undiagnosed cases in ethnic population like the Japanese.^{2,3,5} The etiology is yet to be identified but

is thought to occur either as an autosomal recessive trait with an incomplete penetrance or as a sporadic mutation with multifactorial genetic and environmental triggers.² Although a majority of Binder's syndrome occur as isolated cases, 36% of the cases in a study by Olow-Nordenram and Valentin occurred with a positive familial history.⁶ An autosomal recessive pattern of inheritance with reduced penetrance may be present.⁷ It has been found that, in siblings with unaffected parents, the rate of recurrence is seven times while it increases to ten times when one parent is affected.⁸

Binder suggested that a disturbance in the prosencephalic induction centre in the third month of intrauterine life could lead to the features of this syndrome. Since the vertebral column also develops during the same time period, a large number of such patients also present with vertebral anomalies.¹ Other causative agents such as birth trauma, vitamin K deficiency in the expectant mother, alcohol and drug abuse, use of lithium, ethanol, warfarin or phenytoin based drugs have been indicated.^{1,3} At the fifth and sixth week of pregnancy, inhibition of the centre of ossification which gives rise to the inferior and lateral borders of piriform aperture



Figure 1. Extra-oral view of the patient showing (a): Depressed nasal bridge, broad non-convergent philtrum, crescent shaped nostrils, bow-shaped incompetent lips and anterior open bite. (b): Flat vertical forehead, acute nasolabial angle, flat nasal tip and lack of nasal tip projection. (c): Hypertelorism

is seen. This leads to localised hypoplasia of the maxilla causing retraction of the columella-lip junction and a lack of the triangular flare of columella seen in normal subjects.⁹ Nasal bone which is also formed during the third month of intrauterine life from the ossification centre adjacent to nasal cartilage is under developed. This along with failure of development of premaxilla causes the characteristic appearance of the syndrome.¹ It is important to differentiate Binder's syndrome from other developmental disturbances of the maxillofacial region such as Down's syndrome, Acrodysostosis, Apert's syndrome, fetal warfarin syndrome, and Stickler syndrome in order to accurately plan a treatment.¹⁰

CASE REPORT

An 18 years old male patient reported to the dental hospital with a complaint of defect in the face since birth. On eliciting the genealogical history, the patient was found to be the third of seven children born to healthy parents in a non-consanguineous marriage. No other family member was known to have similar facial features. On examination, the patient was of average height and normal intelligence. A linear scar from a childhood injury measuring approximately 4 cm in length was seen on the right cheek. He had a flat and hypoplastic mid face with reduced sagittal development of the nose and a broad non-convergent philtrum. Flat vertical forehead, hypertelorism and low-set flat nasal tip with lack of tip projection was evident. The nasolabial angle was acute owing to the convexity of the upper lip and flatness of the philtrum.

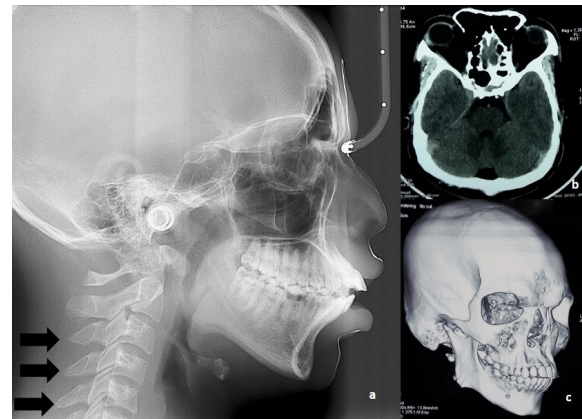


Figure 2. Radiographs of the patient. (a): Lateral cephalogram showing hypoplasia of the anterior nasal spine, thin labial plate of bone covering the maxillary incisors, and triangular shaped spinous processes of the cervical vertebrae (Black arrows). (b): Axial section of the computed tomography image showing retruded maxillonasal base. (c): Three-dimensional reconstruction of CT showing hypoplasia of the anterior nasal spine.

Upper lip was bow shaped and a palpable depression was found at the nasal floor. Classic crescent shaped nostrils were seen (Figure 1). Sense of smell was normal. On audiological examination, bilateral C type immittance with absent acoustic reflex was found. Intraorally, Angle's class I molar relation was seen with anterior open bite. Radiographic analysis of the lateral cephalogram showed hypoplasia of the anterior nasal spine, thin labial plate of bone covering the maxillary incisors, and triangular shaped spinous processes of the cervical vertebrae. Axial sections of the computed tomography image showed a retruded maxillonasal base and three-dimensional reconstruction showed hypoplasia of the anterior nasal spine (Figure 2). Based on the clinical and radiographic features, a diagnosis of Binder's syndrome was made. The patient was advised to undergo pre-surgical orthodontic treatment to favour the reconstruction of his facial profile through camouflage. Esthetic enhancement such as correction of the depressed nasal bridge was planned for a later date.

DISCUSSION

Binder's syndrome, also known as nasomaxillary dysostosis or Binder type maxillonasal dysplasia is a rare congenital malformation of the mid facial region. Although Binder considered it as a mild form of arhinencephaly, researchers have contested this hypothesis since there is usually no evidence of abnormalities of smell.¹¹ Authors have stated that the use of the term 'syndrome' is misleading since the features do not represent a causally defined entity and the word 'dysplasia' should be discontinued as there is no evidence of any histological abnormality in the tissues. They have suggested use of the term

Table 1: Table showing features seen in Binder's syndrome.

Region affected	Features seen
Face	Maxillary hypoplasia, acute naso-labial angle, frontonasal angle of 180°, convex upper lip, concave mid-face (Dish face deformity), small and posteriorly positioned maxilla, strabismus, hypertelorism
Nose	Flat, vertical nose, depressed nasal bridge, short columella
Nostrils	Crescent shaped or triangular, loss of alar flare
Radiological	Short anterior cranial base, decrease of both antero-posterior and vertical dimensions of maxilla, reduced sella-nasion junction,
Intra-oral	Hypoplastic teeth, small maxillary central incisors, congenitally missing incisors or molars, cleft palate, high arched palate, class III tendency (True or Pseudo)
Auditory	Bilateral hearing loss
Skull and vertebrae	Hypoplastic or absent anterior nasal spine, thin labial plate covering maxillary incisors, hypoplastic frontal sinuses, hypoplastic arches or abnormal patterns of ossification of cervical vertebrae, short posterior arch, block vertebrae, thoracolumbar scoliosis, separate odontoid processes, spina bifida occulta
Others (Less commonly seen)	Non-specific congenital heart defects, mental retardation

'association' which affects the maxilla and vertebrae.¹² Features such as arhinoid appearance of the face, abnormally positioned nasal bones, intermaxillary hypoplasia with malocclusion, absent or hypoplastic anterior nasal spine, nasal mucosal atrophy and absent frontal sinuses (not obligatory) are characteristic of Binder's syndrome.¹ The diagnostic feature is the identification of agenesis of anterior nasal spine on laterolateral radiograph.³ Various other features that are seen in the syndrome are listed in Table 1.^{1,3,11} 2D or 3D ultrasonography at 21 weeks of pregnancy can aid in diagnosis by detecting anomalies of nasal bone but it is not confirmatory since various other conditions also present as hypoplastic nasal dorsum at third month of pregnancy.¹³

Since patients with Binder's syndrome lack significant sutural growth potential, correction of the facial defect by orthodontic-orthopedic treatment is difficult. While some researchers claim that an early advancement of the maxilla using extra-oral orthodontic appliance can achieve favourable results, many are of the opinion that orthodontic correction should be postponed until the time of surgical correction since vestibular inclination of the upper tooth roots is inhibited by the thinness of the alveolar bone.¹³ Surgical treatment for Binder's syndrome is usually reserved for correction of the nasal defects since hypoplasia of the mid face allows little space for orthognathic advancement.¹³ Grafting for the hypoplastic naso-chondral scaffolding can be started at 14 years of age. However procedures such as osteotomy of the upper jaw or nose should be planned only after the patient reaches 18 years of age.⁵ Early surgical correction especially of minor cases has been known to jeopardise acceptable occlusal results. It has been seen that such patients have an increased incidence of retruded maxilla, steep occlusal plane angle, large gonial angle and negative apical base angle.¹

Surgical treatment with grafting can improve the convexity of the face and can increase the naso-labial angle to 100-106° from 76-88° that is commonly seen in patients with Binder's syndrome. A common complaint of patients treated with surgery is the resorption of the graft thereby necessitating a second procedure. To overcome this hurdle, L-shaped costal cartilage graft is now being used after immersion in 0.9% sodium chloride solution for 30 minutes.^{1,14} Authors have also suggested insertion of a thin K-wire into the cartilage to prevent its warping. Soft tissue deficiency at the columella is corrected using grafts from the upper lip, free auricular graft, K-Y plasty of the columella or bilateral flaps from the nasal floor.¹ Puttanshetti *et al* used costal graft to augment the mid face, nose and nasal bone and found that the grafts maintained their volumes irrespective of the patient's age or sex.¹⁵ Patients before 16 years of age can undergo onlay bone grafting without osteotomies to help with their self-image.¹ Draf *et al* recommend that screws should be avoided for graft fixation in children in order to protect the roots of permanent teeth.¹⁶ Patients with minor malocclusion can be treated with orthodontics to camouflage the defect and thereby avoid surgery.¹³ Recently allografts have been used by few researchers but it carries with it an additional risk of infections and is not cost effective.¹

CONCLUSION

Binder's syndrome is a congenital disorder that affects the mid-face and sometimes the vertebrae. Early diagnosis helps to support the child patient as well as the parents to cope with the societal as well as psychological issues that may be associated due to the abnormal appearance of the face. Dentists along with paediatricians, orthopaedic surgeons and

plastic surgeons play an important role in the multi-disciplinary approach towards treatment of this rare condition.

CONFLICT OF INTEREST

The authors declare that there were no conflicts of interest related to this case report.

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