Binder’s Syndrome: A Comprehensive Surgical and Orthodontic Treatment of a Case

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ABSTRACT

Binder’s syndrome or maxillonasal dysplasia is a rare clinical entity characterized by abnormal development (dysplasia) of the nasomaxillary region. Management of the syndrome depends on the severity of facial anomalies present and association with other anomalies. Interdisciplinary team management of this malformation is essential. This case report describes a patient with Binder’s syndrome and the treatment aspects in detail.

INTRODUCTION

Binder’s syndrome or maxillonasal dysplasia is a rare clinical entity characterized by abnormal development (dysplasia) of the nasomaxillary region. In 1962, Binder reported three cases and described it as a distinct clinical entity with six characteristic features which include arhinoid face, abnormal position of nasal bones, intermaxillary hypoplasia, reduced or absent anterior nasal spine, atrophy of nasal mucosa and absence of nasal bones, intermaxillary hypoplasia, reduced or absent nasal bones, atrophy of nasal mucosa and absence of frontal sinuses (not obligatory) and malocclusion.

The exact etiology of the syndrome is not clear but Binder suggested that there is disturbance of the prosencephalic induction center during embryonic development. The induction process for both the prosencephalic area and vertebrae is common, accounting for increased vertebral anomalies associated with the syndrome. It was suggested that inheritance could be autosomal recessive with reduced penetrance or multifactorial. Gorlin emphasized that Binder’s syndrome is an abnormality of the nasomaxillary complex and familial examples are as a result of complex genetic factors. Although most of the cases involve only the nasomaxillary complex, a variety of other anomalies have been recorded like cervical vertebrae, skeletal defects, cardiac anomalies, orofacial clefting, strabismus and mental retardation. Other factors like birth trauma and familial history have also been reported. Gorlin et al suggested that familial factors involve complex genetic factors, similar to those involved in producing a malocclusion.

Management of the syndrome varies with the severity of facial anomalies present and association with other anomalies. Interdisciplinary team management of this malformation is essential. Severe maxillary hypoplasia can be treated surgically with Le Fort I and Le Fort II osteotomies. Nasal or columella lengthening, dorsal augmentation with costal cartilage graft are often required for nose reconstruction. Orthodontic expansion of the deficient maxillary arch with correction of impacted and misaligned teeth helps in complete rehabilitation of the syndromic patient.

CASE REPORT

A 22-year-old patient presented with a complaint of irregularly placed teeth and poor facial esthetics. There was no history of prenatal or natal trauma, long-term maternal drug intake or any familial history of a similar condition. Previous surgical history of palatal closure, lip surgery, multiple rhinoplasty and ear cartilage transplantation were reported.

On extraoral examination there was no gross facial asymmetry detected. The patient had a concave profile due to midface deficiency leading to relative mandibular prognathism. There was localized hypoplasia of the premaxilla, nasal, paranasal and alar base region and hypertelorism. The nose was flattened with compressed nostrils. Smell sensation was normal. The mandible showed normal width with increased gonial angle. Depression in ala-nasal and maxillary sinus region was noted. The patient demonstrated macrostomia with an everted lower lip and hypoplastic upper lip with sparse hair in the eyebrow region along with polydactyly (Fig. 1).

On intraoral examination there was a narrow and high palate and V-shaped arch with maxillary lateral incisor missing, over-retained deciduous left canine and a transmigrated incisor in the maxillary arch. The mandibular central incisor was absent on left side. Class I molar relationship present bilaterally (Fig. 2).

The lateral cephalogram (Fig. 3) revealed a decreased anterior cranial base dimension, hypoplastic anterior nasal spine and premaxilla. The maxilla was retrognathetic with increased lower anterior facial height and an increased
mandibular plane angle. The nasomaxillary angle was also increased. Based on clinical and radiographical features, the diagnosis of Binder’s syndrome was concluded. An Abbe flap was recommended for philtrum reconstruction. After release of the flap, orthodontic treatment was initiated with extraction of one lower incisor. Following coordination of the upper and lower arches, the malformed incisor was built up with composite. A forehead flap with cartilage augmentation was performed to increase nasal tip projection.

**DISCUSSION**

The major skeletal abnormality is small maxilla positioned posteriorly on a short anterior cranial base and flat vertically deficient nose. The characteristics of the syndrome are typically visible including hypoplasia of variable severity of the cartilaginous nasal septum and premaxilla. There are associated anomalies of muscle insertions of the upper lip, nasal floor and of the cervical spine. In addition patients have an underdeveloped upper jaw, a relatively protrusive lower jaw with anterior mandibular vertical excess and a Class III skeletal and dental (reverse overjet) profile. The frontal sinus is small with global facial imbalance. In most reports on Binder’s syndrome, the local maxillary hypoplasia of the floor and walls of piriform aperture have been corrected with bone grafts. Various methods of correcting the deformity associated with the Binder’s syndrome have been mentioned in the literature, although no rigid protocols for treatment are followed. Midfacial hypoplasia is the major concern during the treatment of Binder’s syndrome, various treatment options include malar augmentation, Le Fort I and Le Fort II osteotomies, paranasal onlay grafting or has been described for the correction of the midface hypoplasia and malocclusion. The nasal deformity can be corrected with bone grafts, cartilage grafts or alloplastic materials.

In this patient, upper lip reconstruction was performed by building up the philtrum by repositioning the Abbe flap from the lower lip. The flap was developed by the American plastic surgeon Robert Abbe. It is based on the main artery of the orbicularis oris, the labial artery. A portion of the uninvolved lip (either upper or lower) is rotated across the mouth and into the defect of the involved lip while maintaining the blood supply from the labial artery. After 10 to 14 days, the blood supply of the flap get established to the point where the artery can be divided. The Abbe flap has an excellent cosmetic result when it is used to replace the entire philtrum of the upper lip as in the our case.

One supernumerary tooth and over-retained deciduous canine were present on the right side in the maxillary arch and one central incisor in the mandibular arch was missing as seen in occlusal and panoramic radiographs (Fig. 4).

The maxillary right deciduous canine and supernumerary tooth were removed and the permanent canine was distalized and brought into the arch.

The single mandibular incisor was extracted and the teeth were levelled and aligned utilizing the space gained by extraction (Fig. 5).

The degree of mandibular anterior dental crowding, existing mandibular tooth-size excess, and the dental midline discrepancy justified the extraction of one mandibular incisor.

The nasal cartilage was reconstructed by utilizing an auricular and forehead flap with costal cartilage for nasal augmentation (Figs 5 and 6).
Upper and lower part of cartilaginous graft was fixed with soft tissue using nonabsorbable suture. The lower part of columella strut was fixed with upper part of orbicularis oris with prolene suture.

CONCLUSION
Binder’s syndrome or maxilla-nasal dysplasia is an uncommon clinical entity, but the exact birth prevalence remains unknown. It is important to understand that Binder’s syndrome has a variable presentation, and therefore needs to be tackled with various treatment strategies. A careful preoperative assessment and evaluation to determine which treatment modality is essential to provide maximum possible benefit, both esthetically and functionally. Depending on the facial correction needed and patients’ demands, the treatment strategy may include orthognathic surgery or onlay augmentation with autogenous grafts or alloplastic materials.

REFERENCES