Online Journal of Health and Allied Sciences
Peer Reviewed, Open Access, Free Online Journal
Published Quarterly : Mangalore, South India : ISSN 0972-5997
Volume 10, Issue 2; April-June 2011

Case Report:

Binder's Syndrome.

Subhas Babu G, Prof. and Head, Department of Oral Medicine and Maxillofacial Radiology, AB Shetty Memorial Institute of Dental Sciences, Mangalore,
Kumuda Arvind Rao HT, AB Shetty Memorial Institute of Dental Sciences, Mangalore,
Shishir Ram Shetty, Assistant Professor, Department of Oral Medicine and Maxillofacial Radiology, AB Shetty Memorial Institute of Dental Sciences, Mangalore,
Renita Lorina Castelino, AB Shetty Memorial Institute of Dental Sciences, Mangalore.

Address for Correspondence:
Dr. Kumuda Arvind Rao HT,
AB Shetty Memorial Institute of Dental Sciences,
Mangalore, India.
E-mail: kumuda_arht@rediffmail.com

Citation: Babu SG, Rao KAHT, Shetty SR, Castelino RL. Binder's Syndrome. *Online J Health Allied Scs.* 2011;10(2):20
URL: http://www.ojhas.org/issue38/2011-2-20.htm
Open Access Archives: http://cogprints.org/view/subjects/OJHAS.html and http://openmed.nic.in/view/subjects/ojhas.html

Submitted: Apr 15, 2011; Accepted: Jul 15, 2011; Published: Jul 30, 2011

Abstract: Binder's Syndrome also known as 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. It is an uncommon condition characterized by a retruded mid-face with an extremely flat nose. Hereditary factors and vitamin D deficiency during embryonic growth have been researched as possible causes. Morphological characteristics of the syndrome are of fundamental importance for the correct diagnosis and treatment planning of these patients. We hereby report to you a rare case of Binder's syndrome with clinical, radiographic features and discussed the treatment options.

Key Words: Maxillo-Nasal Dysplasia; High arch palate; Hypertelorism

Introduction:

Binder's syndrome (BS) is a congenital malformation characterized by nasomaxillary hypoplasia due to an underdevelopment of the mid-facial skeleton. Binder type maxillonasal dysplasia(MND) is a rare condition characterized by abnormal development (dysplasia) of the nasal and upper jaw (nasomaxillary) regions. Many researchers suggest that Binder type maxillonasal dysplasia does not represent a distinct disease entity or syndrome, but, rather, is a nonspecific abnormality of the nasomaxillary regions. In most cases, the condition appears to occur randomly for unknown reasons (sporadically); rare familial cases have also been reported.

Case Report:

A 10-year old female patient reported with a chief complaint of mal-aligned teeth since 3-4 years and was dissatisfied by poor esthetics. There was no relevant history of pre-natal, natal disorders or long term maternal drug intake. The patient's family history was also non-contributory.

On extra oral examination, no gross facial asymmetry was detected. Concave profile was seen due to midface deficiency. Flattening of right and left cheek was noticed. Localised hypoplasia was noticed in the alar basal region. Premaxilla was hypoplastic with flattening of maxillary base and sagittal shortening of the maxillary arch. Nose was hypoplastic, with flattened alae and the columella was short. Nasal bridge was flattened. Fronto nasal angle was absent. Nostrils were compressed giving it a triangular shape when viewed from below. Sense of smell was normal. Hypertelorism was noticed. Philtral crests were poorly developed, bow shaped and rose vertically without convergence. Mandible showed normal width and increased gonial angle. Relative mandibular prognathism was seen due to maxillary shortening. A palpable depression was present in the ala-nasal floor and maxillary sinus region. Macrostomia with everted lower lip and hypoplastic upper lip and sparse hair in the eyebrow region were also noticed.

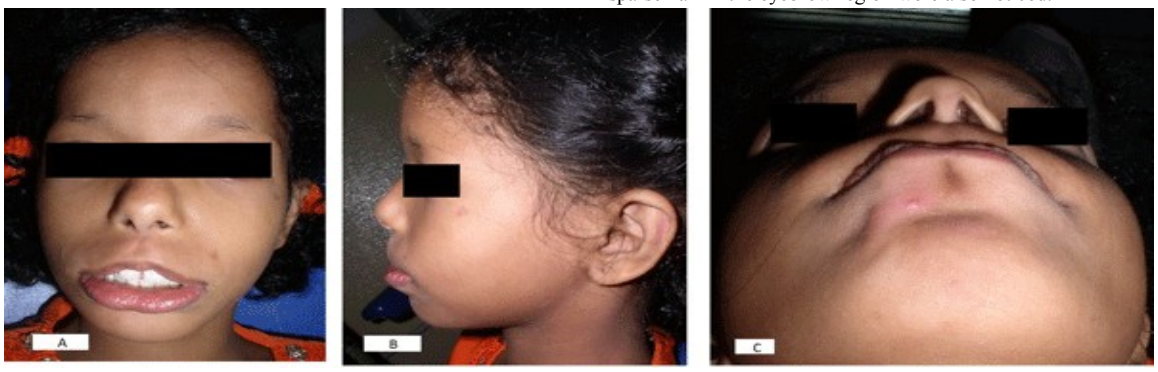

Figure 1: A- Showing malar hypoplasia and macrostomia; B- Showing mid-face hypoplasia, depressed nasal bridge, relative mandibular prognathism; C- showing triangular shaped nostrils.

On intra-oral examination narrow, high arched, V-shaped palate was observed. Macroglossia was present. Patient presented with a class I malocclusion. Spacing was seen in relation to upper anteriors. The tooth eruption pattern appeared normal.

Lateral cephalogram revealed hypoplastic anterior nasal spine and thinning of labial plate of the alveolar bone over upper incisors. The maxilla was retrognathic. The lower third face height was increased. The naso maxillary angle was increased. Cephalometric studies revealed increased gonial angle and proclination of incisors. Decreased anterior cranial base measurements, smaller maxilla vertically and antero-posteriorly were also noticed. Based on clinical and radiographic features, a diagnosis of Binder's syndrome was arrived at.

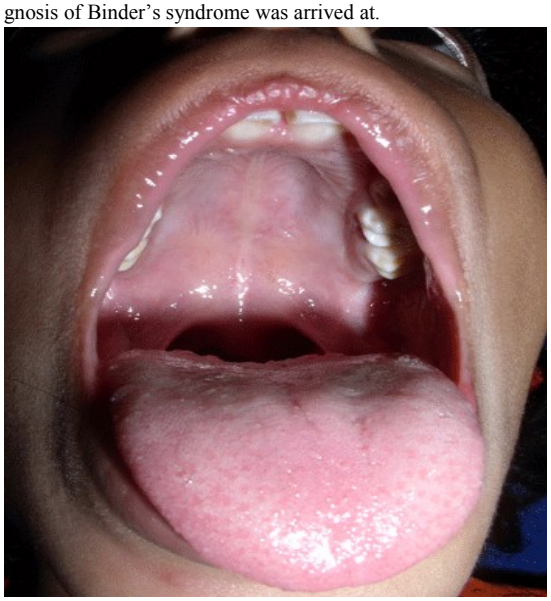

Figure 2: Showing macroglossia and high arch palate.

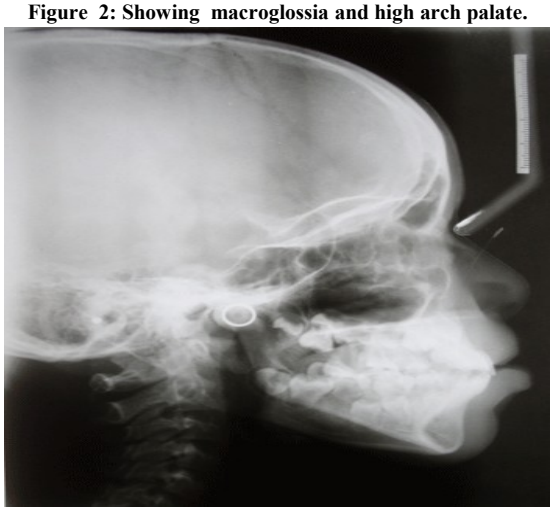

Figure 3: Lateral cephalogram showing hypoplastic anterior nasal spine.

Patient was referred for orthodontic and surgical evaluation for effective management.

Discussion:

The essential feature of MND was initially described by Noyes in 1939, although it was Binder who first defined it as a distinct clinical entity in 1962.[1] He reported on three cases and recorded six specific characteristics: Arhinoid face, abnormal position of nasal bones, inter-maxillary hypoplasia with associated malocclusion, reduced or absent anterior nasal spine, atrophy of nasal mucosa, absence of frontal sinus (not obligatory).[2] The orthodontists and surgeons are more closely associated with these patient's due to the malocclusions and facial abnormalities, and the information regarding BS is very sparse in the literature.

MND is a nonspecific abnormality of the nasomaxillary complex. The familial examples are a result of complex genetic factors, similar to those involved in producing a malocclusion.[3] Although most cases involve only the nasomaxillary complex, a variety of other anomalies have been recorded including especially cervical vertebrae anomalies, but also various other skeletal defects, cardiac anomalies, orofacial clefting, strabismus, mental retardation, and other abnormalities.[4,5] However these features were not observed in our case. The affected patient's show mid-face profile which is hypoplastic, flattened nose, convex upper lip with a broad philtrum, typically crescent or semi-lunar in shaped nostrils due to the short collumela, and a deep fold or fossa occurring between the upper lip and the nose, resulting in an acute nasolabial angle.[6] All but the last features were observed in our case.

The maxilla was hypoplastic in both antero-posterior and vertical directions in early childhood and the latter showing some improvement with age.[7] This causes the feature of relative mandibular prognathism although mandibular length may be greater than normal, suggesting true prognathism in some cases.[8,9]

Binder believed that his patients had a mild form of arhinencephaly but there have been no reports of difficulties with the sense of smell to support this hypothesis.[2]

Similar facial features may be seen in other well defined conditions, including warfarin embryopathy, acrodysostosis, and Stickler's syndrome which may be included under differential diagnosis. It should be possible to distinguish each of these syndromes on the basis of additional historical and clinical features.[10]

After assessment of the degree of facial bone abnormality, orthodontic and surgical procedures can be planned.[11] More severe cases require a Le Fort I or II osteotomy with nasal grafting.[12] As the degree of malformation in BS varies significantly, surgical correction needs to be individually tailored.[13]

Conclusion:

The characteristics of the Binder's syndrome are typically visible since a young age. It can also be present in combination with other malformations. In such severe cases, the syndrome requires combined orthodontic and surgical treatment to achieve an adequate facial profile.

References:

1. Noyes FB. Case report. *Angle Orthod* 1939;9:160–165
2. Binder KH. Dysostosis maxillo-nasalis, ein archinencephaler Missbildungskomplex. *Deutsche Zahnarztuche Zeitschift* 1962;17:438–444.
3. Gorlin R, Pindborg JJ, Cohen M Jr. Maxillonasal dysplasia (Binder syndrome). Syndromes of the head and neck. 2nd ed. New York: McGraw-Hill; 1976.
4. Delaire J et al. clinical and radiographic aspects of maxillonasal dysostosis (Binder's syndrome). *Head Neck Surg* 1980;3:105-122.
5. Olow-Nordenram MA, Radberg CT. Maxillonasal dysplasia (Binder's syndrome) and associated malformations of the cervical spine. *Acta Radiol (Diagn)*1984;25:353-360.
6. Dyer FMV, Willmot DR. Maxillo-nasal dysplasia, Binder's syndrome: review of the literature and case report. *Journal of Orthodontics*. March 2002;29(1):15-21,
7. Horswell BB, Holmes AD, Levant BA, Barnett JS. Cephalometric and anthropomorphic observations ofBinder's syndrome: A study of 19 patients. *Plast Reconstr Surg* 1988;83:325-335.

8. Olow-Nordenram M. Masillonasal dysplasia (Binder's syndrome). A study of craniofacial morphology, associated malformations and family relations. *Swted Dent J [Suppl]* 1987;47:1-38.
9. Munro IR, Sinclair WJ, Rudd NL. Maxillonasal dysplasia (Binder's syndrome). *Plast Reconstr Surg* 1979;63:657-663.
10. Quarrell OWJ, Koch M, Hughes HE. Maxillonasal dysplasia (Binder's syndrome). *Med Genet* 1990;27:384-387.
11. Rintala A, Ranta R. Nasomaxillary hypoplasia-Binder's syndrome. *Scand J Plast Reconstr Swrg* 1984;19:127-134.
12. Holstrom H. Surgical correction of the nose and midface in maxillonasal dysplasia (Binder's syndrome). *Plast Reconstr Surg* 1986;78:568-580.
13. Bhatt YC, Vyas KA, Tandale MS, Panse NS, Bakshi HS, Srivastava RK. Maxillonasal dysplasia (Binder's syndrome) and its treatment with costal cartilage graft: A follow-up study. *Indian J Plast Surg*. 2008 Jul;41(2):151-159.