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Case Report

Binder's Syndrome: The Need of an Ortho-Surgical Approach

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Abstract

Despite the frequent presence of all characteristic orofacial features and profound dental malocclusions there is very little to be found in the literature of maxillofacial surgeons to diagnose a case of Binder syndrome or also known as maxillo-nasal dysplasia or maxillo-nasal dysostosis. Nordenram., et al. recommended to delay any definitive orthodontic treatment in individuals with this syndrome up to the time when growth has ceased. For severe cases they also advise for grafting in the nasal area along with Le-Fort I and II osteotomy. Thus, in such cases the orthodontic treatment should go hand in hand with the surgical team hence opting for an ortho-surgical approach. The aim of the orthodontist in the post-surgical phase should be improvement of facial beauty, relieve crowding of the arches and their co-ordination and to obtain a Class I molar relationship.

Keywords: Binders Syndrome; Maxillo-nasal Dysplasia; Orthodontic Management of Binders Syndrome

Introduction

Despite the frequent presence of all characteristic orofacial features and profound dental malocclusions there is very little to be found in the literature of maxillofacial surgeons to diagnose a case of Binder syndrome or also known as maxillo-nasal dysplasia or maxillo-nasal dysostosis [1]. This is a rare congenital malformation characterized by an abnormal development of the maxilla and the nasal complex. The extent of deformity in patients suffering with this syndrome is highly debatable.

According to some, it has its involvement only in the nasal region whereas for others, it affects the entire midface but actually it is a triad formed by a nasal flattening in the mid-sagittal profile, a verticalization of the nasal bones and a maxilla's retroposition. It has been suggested by some authors that the disorder cannot be represented as a single nosologic entity and that the use of the word 'syndrome' or 'dysplasia' is inappropriate. They suggest the use of Binder 'phenotype' [1] or 'association' or a 'symptom' as a non-specific abnormality of the naso-maxillary complex.

Etiology

The syndrome carries a rather obscure etiology. Binder had stated an archinecephalic origin with disturbance of prosencephalic induction centre during embryonic growth [2,3] for the defect. Although birth trauma was suggested as a causative factors by Noyes [4], Hopkin [5] denied it in his five cases reported. Ferguson and Thompson [6] and few others were in support of a genetic etiology and concluded that the inheritance of the syndrome could be autosomal recessive with reduced penetrance. Gorlin., *et al.* [7] finally concluded that the syndrome is a non-specific abnormality of naso-maxillary complex and the familial examples that occur are a result of complex genetic factors.

Clinical features

Initially described by Zuckerland [8] in 1882, it was Binder [9] who, in 1962, reported three cases and described the six most remarkable characters of the syndrome:

- Facial dysmorphism secondary to a naso-maxillary hypoplasia,
- Absence or decreased nasal bridge,
- Short nasal columella,
- Convex upper lip with associated dental angle class III malocclusion,
- Nasal mucosal atrophy,
- Absence of the frontal sinus (not obligatory).

Intra-oral dental features

- Proclination of upper anterior.
- Absence of anterior teeth (lateral and central incisors).
- Pseudo prognathism of the mandible.
- Open bite.
- Crowding.

Cephalometric evaluation

Patients with Binder syndrome have a typical facial appearance, including midfacial hypoplasia with verticalized nasal bones. Most patients have some or all of the features described, depending on the severity of the syndrome.

- Flattened tip and alar wings leading to acute naso-labial angle that is less than 90°.
- A short anterior cranial base length [3].
- Presence of a naso-frontal angle measuring between 150° and 160° (compared to the normal value of 135°).
- A concave midfacial profile caused by the hypoplasia of the upper maxilla and its retroposition.

Case Report and DIscussion

This case is of a 9-year-old male patient who reported to our clinic with a chief complaint of depressed mid face. On extra-oral examination (Figure 1) he presented with evident mid-facial hypoplasia. There is depressed nasal bridge with deficiency of malar prominence and a reduced fronto-nasal angle with concave profile. The lips remained competent at rest with absence of any obvious facial or functional asymmetry.

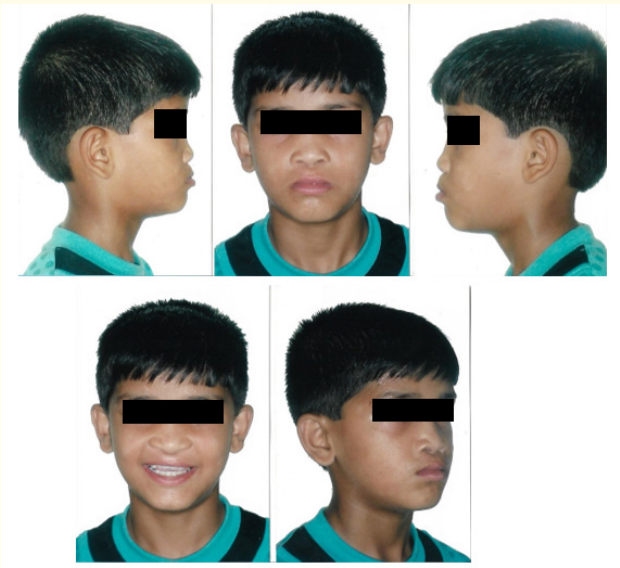

Figure 1: Extra-oral pictures.

Upon Intra-oral examination, the presence of mixed dentition was seen (Figure 2). Mild crowding was present in both arches while the maxillary arch exhibited displaced canines and left lateral incisor. In addition, reverse overjet of 1 mm was seen.

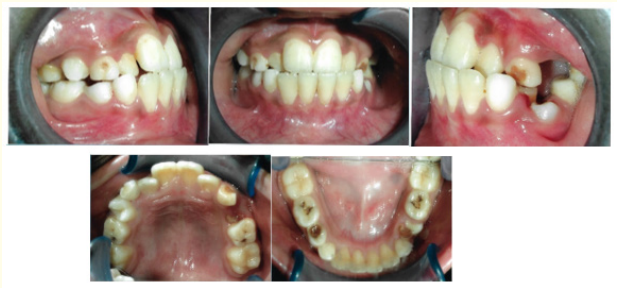

Figure 2: Intraoral pictures.

Lateral cephalogram (Figure 3) and Orthopantomogram (Figure 4) of the patient was obtained and studied. The cephalometric values (Table 1) of the case studied were as under.

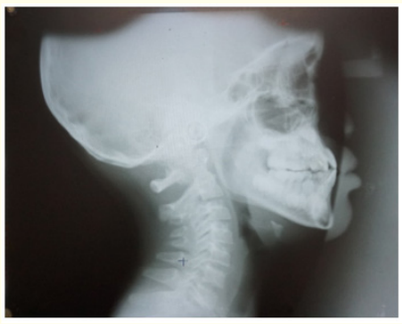

Figure 3: Lateral cephalogram.

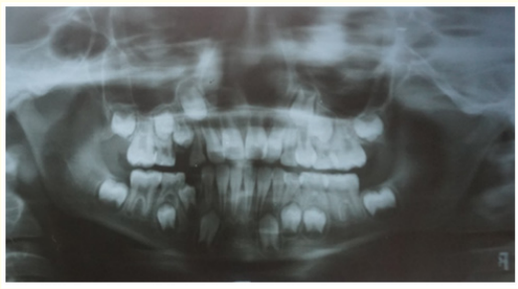

Figure 4: Orthopantomogram.

Cephalometric analysis

Tweed's	**Pre**	**Normal**	**Bjork's**	**Pre**	**Normal**
FMA	20	25	N.-S.-Ar.	131	123 ± 5
IMPA	97	90	S.-Ar.-Go.	137	143 ± 6
FMIA	63	65	Ar.-Go.-Me.	119	128 ± 7
Down's	**Pre**	**Normal**	**Soft-tissue analysis**	**Pre**	**Normal**
Facial Angle	89	87.8 (82-95)	Steiner Steiner's Upper lip	3	0
Angle of convexity	-16	0 (-8.5-10)	H9h8b8 Steiner's Lower lip	6	-2
AB Plane angle	8	-4.6 (0- -9)	Rickett's "E"line(upper)	1.5	-4
Mandibular. Plane Angle	23	21.9 (17-28)	Rickett's "E"line(lower)	5	-2
"Y" Axis	62	59.4 (53-66)	"H" Angle	13	10 (7-15)
Cant of Occ. Plane	3	9.3 (1.5-14)	"Z" Angle	68	80 (± 9)
Mx.1 to Md.1	117	135.4 (130-150.4)	S.T. Facial Angle	92	91 ± 7
Md.1 to O.P.	24	14.5 (3.5-20)	Nose Prominence	10	14-24
Md.1 to M.P.	3	1.4 (-8.5-7)	Sup. Sul. Depth	6	3 (1-4)
Mx.1 to A Pog.	13	2.7(-1 - 5) mm	S.T .Sub N to "H" line	10	5 (3-7)
			A to N- Pog	-7	0 ± 2
Steiner's	**Pre**	**Normal**	**Basic U.L. Thickness**	**13**	**15**
SNA	74	82	Mx1 to V.B of U. lip	14	13-14
SNB	81	80	Lip- Strain	0	1
ANB	-7	2	L. lip to "H" line	4	1-2
SND	78	76	Inf. Sul to "H" line	3	5
Mx.1 to NA	19/48	4/22	S.T. chin thickness	10	10-12
Md.1 to NB	4/23	4/25	U. L. length	21	24
Pog. to NB	3.5	0	L. L. chin length	45	40-45
Pog.-Md.1 to NB	3.5:4	1:1	Naso-labial Angle	81	94 -110
O.P. to SN	12	14.5	JARABAK RATIO		
Go-Gn to SN	26	32			
Mx.1 to Md.1	117	131	S-Go/N-Me×100	71.4%	62-65%
1 to SN	123	104 ± 7	U/L Ant. Facial height		
1 to FH	128	107	N-ANS	47	45%
WIT'S	-9	0 - 1	ANS-Me	65	55%

Table 1: Cephalometric evaluation.

Treatment options

Looking at the cephalometric variables we can now understand why the treatment of this complex maxilla nasal deformity would mainly require both orthodontic and surgical intervention.

A SNA angle of 72°, ANB of -7°, WIT's of -9°, Angle of Convexity of -16°and AB Plane angle of 8° is suggestive of maxillary hypoplasia presenting with skeletal class III jaw bases. Although nature compensates itself which is evident from the dental and soft tissue compensation. The dental compensation has occurred by excessive proclination of the maxillary anteriors which is clear as seen in the values of upper 1 to NA as 19mm ahead of NA and 48° to NA line rather than the normal 4mm and 22°. Also, both the upper 1 to SN and FH shows increased values of 123° and 128° respectively rather than their normal of 104° and 107°.

The soft tissue compensation can be seen in the values of S-line and E-line to the upper and the lower lip linear measurements. A decreased nasolabial angle as also suggested as a classic feature for the syndrome by Binder [9] can be seen.

Nordenram., *et al*. [3] recommended to delay any definitive orthodontic treatment in individuals with this syndrome up to the time when growth has ceased. For severe cases they also advise for grafting in the nasal area along with Le-Fort I and II osteotomy. Thus, in such cases the orthodontic treatment should go hand in hand with the surgical team hence opting for an ortho-surgical approach. The aim of the orthodontist in the post-surgical phase should be improvement of facial beauty, relieve crowding of the arches and their co-ordination and to obtain a Class I molar relationship [10].

Conclusion

Even though the malocclusion is closely related to maxillo-nasal dysplasia, the condition used to be treated by plastic surgeons alone traditionally. However, the combination of various facial and dental deformities creates the necessity for an inter-disciplinary managemental approach with formulation of a proper treatment plan. This is very essential since milder cases of Binder's syndrome can be treated by combination of minor surgical procedures followed by orthodontic treatment unlike the more complex cases.

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