Binder's syndrome and aesthetic improvement: A case report

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Abstract
Binder's syndrome is a rare congenital anomaly affecting the facial skeleton. It is also known as nasomaxillary hypoplasia affecting anterior maxilla and the nasal complex. The etiology of this syndrome is not well defined. The diagnosis is mainly done through analyzing the morphological characteristics and the radiological findings. Here we present a case of Binder's syndrome with its morphological features, which has been corrected with surgical augmentation of both maxilla as well as the nasal complex using costochondral cartilage grafts.

Keywords: Binder's syndrome, Hypoplastic maxilla, Nasolabial angle, Costochondral graft.

Introduction
Binder's syndrome is a rare syndrome involving the facial skeleton. The syndrome is characterized by the nasomaxillary hypoplasia resulting in a hypoplastic maxilla and depressed nose. The exact facial features are the maxillary retrusion, saddle nose, small columella and triangular nostril with an acute nasolabial angle[1]. There will be secondary mandibular prognanthism in severe case of Binder's syndrome resulting in Angle class lll malocclusion. The exact etiology of this condition is not known with postulated hereditary as well as vitamin K deficiency during the embryonic growth as postulated factors for its occurrence[2]. The disturbance of the prosencephalic growth centre during embryonic life is suggested as the root cause by Binber. The main problem with this syndrome is the unacceptable aesthesis rather than functional disability. Thus the management banks on surgical augmentation of the maxilla and the depressed nose along with correction of dental mal-alignment. The augmentation can be done with autogenous or alloplastic materials. The dental alignment correction varies from orthodontic procedure to orthognathic surgical correction depending on the severity of Binder's syndrome.

Case report
Our patient is a 17 year old girl presented to us with worries about the appearance of her mid face and nose. On clinical examination she had mid face hypoplasia with mildly retruded maxilla. The nasal complex showed depressed dorsum of nose with saddle nose deformity. The nasolabial angle was acute (45 degree) with a short columella and a convex upper lip (Fig. 1). She had already undergone orthodontic treatment for her malocclusion with good results. On clinical evaluation the dental occlusion was Angle class l. Lateral cephalogram showed maxillary hypoplasia with retrusion of maxilla around the pyriform area with class l occlusion (Fig 2). The SNA was 85 degree and SNB was 86 degree. Soft tissue marking on the lateral cephalogram also showed the depressed nasal dorsum with saddle nose deformity and acute nasolabial angle.

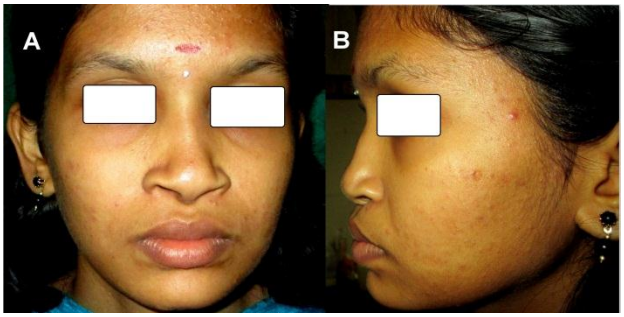

Fig. 1: A-Pre-operative frontal profile view. B- Lateral profile view showing saddle nose deformity, acute nasolabial angle and retruded maxilla.

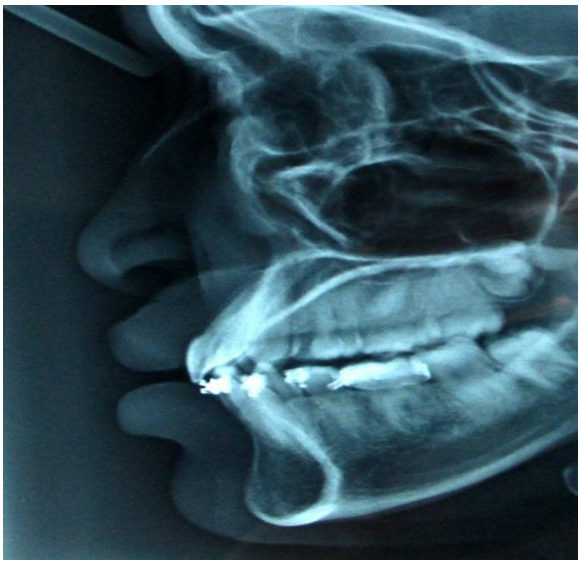

Fig. 2: Lateral cephalogram showing the soft tissue as well as the bony profile of our patient with Binder's syndrome.

Surgery

The preoperative planning of the augmentation of the anterior maxilla and the rhinoplasty with an L strut was made. The augmentation was done with costochondral cartilage graft taken from the right side chest wall. The cartilage was harvested through a small sub mammary incision. The cartilage graft was planned in a fashion that a block of cartilage for the hypoplastic maxilla to give a platform over which open augmentation rhinoplasty was performed. The approach to the maxilla was through the upper gingivobuccal sulcus in subperiosteal plane. The cartilage was fixed with screws into the maxilla (Fig 3). The alar bases were augmented with cartilage chips placed in the same plane below the nasal tripod. The columella was strengthened with a columellar strut and the dorsum of nose was augmented with a costal cartilage graft (Fig. 4). Both theses cartilages were carved from the centre part of the costal cartilage to prevent warping. The columellar strut was placed in slot made in the costal cartilage block placed in the anterior maxilla and secured with sutures. The closure of upper gingivobuccal sulcus and the skin incisions were done in layers. The donor site closure was done in layers after putting negative suction drains. The external nasal splintage was done.

The post operative profile of the patient showed improvement in the midface hypoplasia with projection of the nasal base area, a normal appearing nose and normal nasolabial angle of 84 degree. The saddle nose deformity was completely corrected (Fig. 5).

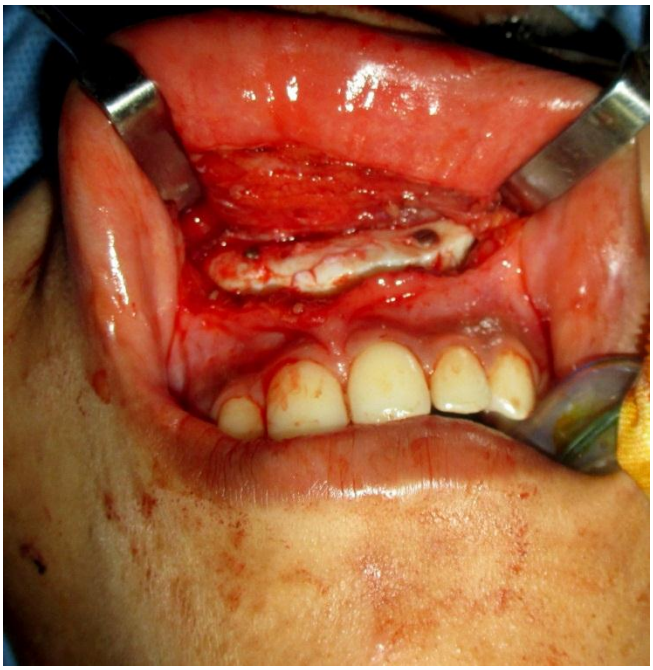

Fig. 3: Intra-operative picture showing, the screw fixation of the costochondral cartilage graft to the premaxilla.

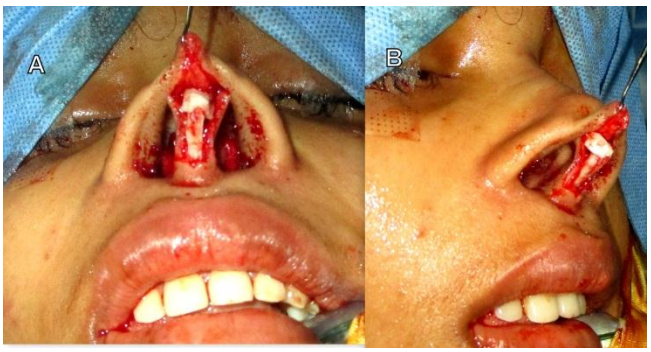

Fig. 4: Intra-operative picture of columellar strut (A) and dorsal graft (AandB) in the form of an L strut.

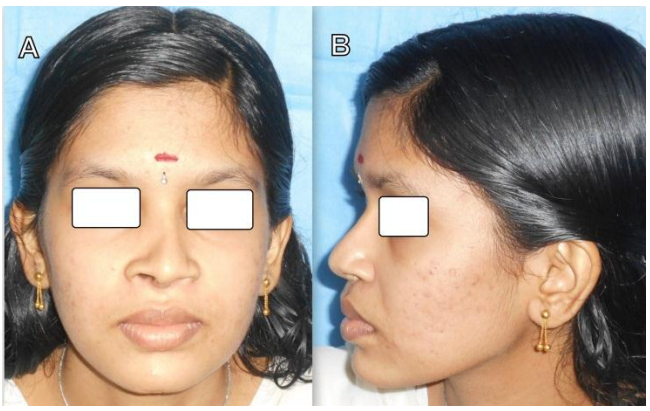

Fig. 5: Post-operative picture A. Frontal profile showing well defined dorsum B. Showing good nasolabial angle and projected tip of nose with absent saddle nose deformity.

Discussion

Binder's syndrome is characterized by the dish mouth appearance of the face due to the nasomaxillary hypoplasia. Von Binder in 1962 described this syndrome with short nose with flat bridge, absent frontonasal angle, absent anterior nasal spine, limited nasal mucosa, short columella, acute nasolabial angle, perialar flatness and convex upper lips. They often have a tendency to go for class lll malocclusion. Binder considered these deformities due to rhinocephalic dysplasia later termed as nasomaxillary dysplasia. Holmstrom H[3] described these anomalies in the form of crescent shaped nostril without a sill, low set flat nasal tip, stretched out and shallow cupid's bow, retracted columella lip junction, lack of normal triangular flare at the nasal base, convex upper nasal tip with a wide shallow phitrum and a perpendicular alar cheek junction. He found out a possible heridetary inheritance in the form of autosomal recessive trait with incomplete penetrance. His experience with Binder's syndrome showed use of cancellous bone graft along with anterior advancement of the nasal septum for projection of the nasal tip[4].

Jain U et al in their report of Binder's syndrome described the clinical findings in detail with a management schedule of the condition according to the severity and time of presentation. Goh RC et al in their study evaluated 24

patients treated over a period of 27 years and used both silastic as well as autologous tissue for the reconstruction. According to them cartilaginous graft had an extra edge over bone graft due to less occurrence of resorption of the graft. They also used silastic material only for dorsal augmentation along with cartilage graft[5]. Gewalli F et al evaluated nasal and maxillary augmentation done with bone and cartilage graft, in centers in Mexico and Sweden to look for the merits of each method. Even though the cartilage graft showed stability in nasal tip projection they had relapse in both long term and short term follow ups. The nose tip length ratio and normalization of anthropometric variables were noticed in both cases in long term follow ups[6]. Chummun S et al reviewed 107 patients treated with costochondral cartilage grafts for nasal dorsal augmentation out of which 46% patients were of Binder's syndrome. They came to a conclusion that cantilevered nasal costochondral grafts are excellent option for nasal dorsal augmentation[7]. Deshpande SN[8] in his patients with Binder's syndrome managed deformity correction according to the involvement of bone as well as soft tissue. In case of class lll malocclusion Lefort ll osteotomy and advancement was done and calvareal bone graft and silastic materials were used for nasal augmentation. Bhatt CY et al tried costochondral grafts for both maxillary as well as nasal augmentation and came out with acceptable results[9]. In our case we used costal cartilage for the augmentation of both maxilla as well as the nose.

Holmes et al[10] had a treatment plan in nasomaxillary dysplasia for prepubertal and post pubertal individuals differently. In prepubertal patients he applied silicone tissue expansion prior to graft insertion. In postpubertal individuals he directly went for graft placement in initial sitting itself. He managed maxillary deficiency with carlilage graft along with doral augmentation with cantilever technique with grafts dwelled in to the frontal sinus wall. He concludes his study stating that the tissue expansion technique is an excellent option for the pliability and stretching of the contracted nasal tissue prior to graft placement. He also states this can also be used in post pubertal individuals with contracted soft tissue envelopeMonasterio et al studied Binder's syndrome patients in long term. They started the nasal and the pyriform area early prepubertal age along with tissue expansion. The role of sequential lengthening in dorsum and the columella had advantages. The Lefort osteotomy was reserved for the patients with class lll malocclusion once the cross the teenage period[11]. In our patient the soft tissue envelope was yielding and there was no secondary class lll malocclusion, so we were able to come out with good result.

Conclusion

The diagnosis of Binder syndrome is straight forward with its clinical and radiological characteristics. Costochonral graft augmentation of nose and maxilla gives satisfactory result in Binder syndrome with class l dental occlusion.

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