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Research Paper

Binder's syndrome: a case report and literature review

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ABSTRACT : Binder's syndrome is an uncommon entity characterized by midfacial hypoplasia. The individuals with this syndrome are easily recognizable by a maxillary hypoplasia and flat vertical nose. The current work presents a case of this rare syndrome and describes its general features. **Keyswords** :Binder's syndrome, midfacial hypoplasia, congenital malformation, Orthodontic treatment

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I. INTRODUCTION :

Binder's syndrome or maxillonasal dysplasia is a rare congenital malformation and the causes are unclear. In 1939, Noyes[1] described a patient with a flat nasal tip sitting on a retrudedmaxillonasal base, and in 1962, von Binder[2] described a syndrome consisting of a short nose with a flat bridge, absent frontonasal angle, absent anterior nasal spine, limited nasal mucosa, short columella and acute nasolabial angle, perialar flatness, convex upper lip and a tendency to class III occlusion. Occasionally, there may be hypoplastic frontal sinuses. Binder's patients may present other congenital diseases and abnormalities (such as Down's syndrome, autonomic neuropathy, strabismus, congenital heart diseases, vertebral anomalies and mental retardation) [3]. The management of these patients depends on the level of complexity because of variations in the midface discrepancy and the occlusal relationship

II. CASE REPORT:

A 20-year-old female reported to the oral and maxillofacial surgery department with the chief complaint of facial deformity (Fig.1). The anamnesis of the patient did not show any abnormalities in the various health-related and cognitive parameters. There was no family history of similar complaint. Extraoral examination showed hypoplasia of the middle third of the face, a wide and flattened nose, short columella, horizontal nostrils, wide philtrum and convex upper lip with an acute nasolabial angle. Intraoral examination revelated class I incisors in relation with Angle's class I canine and molar.

Cephalometrique analysis showed classes Ш skeletal malocclusion with а pseudomandibularprognathism, hypoplastic maxilla and agenesis of the anterior nasal spine (Fig.2). Orthodontic treatment was not necessary because of compensatory effects in dental arches. The patient was planned for open rhinoplasty. Firstly a costal cartilage grafts were harvested from the right side of the chest through a small submammary incision (Fig.3), the cartilge was kept in 0.9% NaCl and gentamicin solution to prevent warping. To achieve an anterior projection of the nose, three cartilaginous splinters were placed: one on the dorsum, the second into the columella, another piece of cartilage was put in the right alar region to correct the alar deformity. We didn't have any post operative complication. The aesthetic result was well accepted by the patient (Fig.4).

III. DISCUSSION :

Although the majority of cases of this malformation are sporadic, familial recurrence has been noted by a number of authors [3,4]. Noyes [1] considered that his patient's abnormalities resulted from birth trauma but did not comment on how this could account for the absent anterior nasal spine.

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. The nasal deformity can be corrected with bone grafts, cartilage grafts or the alloplastic materials [5]. Similarly, paranasal only grafting or a Le Fort I or II osteotomy has been described for the correction of the midface hypoplasia and malocclusion. In our cases with class I occlusion, we have used only cartilage grafts to correct the nasal deformity

IV. CONCLUSION:

The case report presents the craniofacial characteristics compatible with binder's syndrome, as supported by literature. The knowledge of ideal proportion of the face helped us to achieve the correct diagnosis of the syndrome and its proper treatment.

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Figure 1 : Lateral view of the face showing flat nose, maxillary hypoplasia, and reduced Frontonasal angle



Figure 2: Lateral cephalogram showing absence of the anterior nasal spine,maxillary hypoplasia and mandibular pseudo prognathism.



Figure 3: submammary incision for rib cartilage



Figure 4: Afterdorsal augmentation.

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