Frontonasal Dysplasia: A Rare Case Report and Review of Literature

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Abstract

Frontonasal dysplasia or Binder syndrome is a rare congenital developmental defect of the maxilla and nasal skeleton. Synonyms are maxillo-nasal dysplasia, maxillo-nasal dysostosis, maxillofacial hypoplasia, or naso-maxillo-vertebral syndrome. Clinical feature includes a hypoplastic or absent anterior nasal spine, a short and flat nose with short columella, an acute nasolabial angle, a convex upper lip and class III malocclusion. We hereby report a rare case of Binder’s syndrome with clinical, radiographic features with cephalometric analysis, review of literature and discussed the treatment options.

Keywords: Frontonasal dysplasia; Binder syndrome; Hypoplastic maxilla; Hypertelorism; Maxillonasal dysplasia

Introduction

Binder’s syndrome [5-7] or maxillonasal dysplasia is a congenital malformation. The first case was described by Noyes. In 1962 Binder reported a comprehensive case of three unrelated children and recognized its syndromic features and called as dysostosis maxillonasalis [1,2]. Binder described 6 characteristic features of BS that includes, enlarged nasal angle (arhinoid face), abnormal position of nasal bones, intermaxillary hypoplasia, reduced or absent nasal spine, atrophy of nasal mucosa, absence of frontal sinus and malocclusion.

Several authors proposed different theories on etiology of Binder’s syndrome.

- Gorlin et al. suggested that maxillonasal dysplasia is a nonspecific abnormality of the nasomaxillary complex [3,4].
- During pregnancy exposure to exogenous factors like, vitamin K deficiency and vitamin K antagonist drugs (warfarin, acenocoumarin), anticonvulsives drugs, retinoid acid or birth trauma, leading to a disturbance of the prosencephalic induction center during embryonic growth. The process of nose formation takes place during the third month of pregnancy [5-7].
- According to Holmstrom et al. inhibition of the ossification center at the lateral and inferior border of the pyriform fossa is responsible for the growth retardation and that leads to Binder syndrome [8,9].
- Olow-Norderam reported the positive family history in 36% of his patients inherited in an autosomal recessive manner [4].
- Noguchi et al. investigated the correlation between transthyretin gene mutation and facial growth retardation in an animal studies. The mutation leads to delayed transthyretin production that causes massive cell death in the nasal placode leading to hypoplasia of the frontonasal region. Transthyretin is a transport protein that carries the thyroxine and retinol binding protein bound to retinol. Excess amount of retinol and its deprivation may lead to abnormal fetal development [5].

Case Report

A 15-year old male patient reported with a chief complaint of malaligned teeth and poor esthetics. The patient’s parents had consanguineous marriage and patient’s sister had undergone cleft palate surgery. There was no relevant history of pre-natal, natal disorders or long term maternal drug intake.

On extra oral examination, facial asymmetry was detected. Concave profile was seen due to midface deficiency. Flattening of right and left cheek was noticed. Hypoplastic was noticed in thealar basal region. Premaxilla was hypoplastic with flattening of maxillary base. Nose was hypoplastic, with flattened alae and the columella was short. Nasal bridge was flattened. Fronto nasal angle was absent. Sense of smell was normal. Hypertelorism was noticed. Philtral crests were poorly developed; bow shaped and rose vertically without convergence (Figure 1).

On intra-oral examination patient presented with a class III malocclusion. The tooth eruption pattern appeared normal. Crowding was seen in relation to upper anteriors. Lower first molars were decayed. Upper central incisors attrited. Upper right lateral incisor shows peglaterals. Upper left lateral incisor was congenitally missing (Figure 2a).
Figure 1: Water’s view shows facial asymmetry, deviated nasal septum.

Figure 2: (a) Intraoral photos showing crossbite and missing left lateral incisor and right peg lateral. (b) Orthopantomograph.

Radiological examinations reveal Orthopantomograph, Lateral Skull view and water’s views. Orthopantomograph reveals congenitally missing upper left maxillary incisor (Figure 2b) water’s view shows facial asymmetry, and deviated nasal septum (Figure 3).

Figure 3: Frontal view of the face showing facial asymmetry and Hypertelorism. Lateral view of the face showing flat nose, maxillary hypoplasia, and acute nasolabial angle.


Lateral cephalogram revealed hypoplastic anterior nasal spine and thinning of labial plate of the alveolar bone over upper incisors. Cephalometric studies revealed increased gonial angle (145°) and proclination of incisors. The nasolabial angle, measured between the tip of the nose and upper lip, is acute (65°). There is reduced sella nasion distance (35 mm) Decreased
anterior cranial base measurements, smaller maxilla vertically and antero-posteriorly were also noticed (Figure 4). Based on clinical and radiographic features, a diagnosis of Binder’s syndrome was arrived.

**Discussion**

Binder’s syndrome patients show characteristic appearance which are easily recognizable [10] such as,

- **Face:** Maxillary hypoplasia, flat and vertical nose, short columella, semilunar shaped nostrils. The upper lip is convex with an acute nasolabial angle. Angle type III malocclusion, true prognathism. Hypoplasia of the nasal floor and the adjacent part of the maxilla leads to Dish-face anomaly [1,11].
- **Teeth:** Small central upper incisors, amelogenesis imperfecta [1], crow in the upper jaw, protrusion of the maxillary incisors, anterior crossbite and open bite [2]. Amelogenesis imperfect 7
- **Palate:** Cleft palate, high arched palate.
- **Intelligence:** Occasionally mental retardation [1].
- **Vertebral:** Separate odontoid process, a short posterior arch, spina bifida occulta and blocked vertebrae [11].
- **Caries & Periodontium:** Due to the physical limitations and intellectual deficit in some patient’s leads to accumulation of biofilm develops gingivitis and carious lesions. In pedo patients caries progression is faster, due to lower mineral content leads to early loss of tooth structure which affects the occlusion [12].
- **Other features:** Strabismus, Deafness, or autonomic neuropathy may be seen occasionally congenital heart diseases [1,11].

Kepler-Noreuil et al. did a study on 24 patients in which 13 patients had trisomy 21, two with trisomy 18, one with mosaic trisomy 21, one with vitamin K epoxide reductase deficiency, one patient had Stickler syndrome, and another had fetal warfarin syndrome; and one case of Robinow syndrome was detected [5].

Olow-Nordenram et al. stated that 44% of a group of 43 patients presenting BS had malformations of cervical vertebrae, the atlas and axis being the most frequently affected [13]. Another study revealed that 15 of 28 patients (53.5%) suffered from cervicospinal and craniospinal anomalies [14]. Antoneli et al. reported a case with BS had mild conductive hearing loss and a cleft palate [15].

Cephalometric findings showed the following significant differences:

- Congenital absence of the anterior nasal spine,
- Hypoplasia of the middle third of the face,
- Midfacial retrusion and dishface,
- Hypoplastic frontal sinuses,
- Short anterior cranial base,
- Small cranial base angle,
- Reduced sagittal development of the nose,
- Straight profile,
- Retrogнатhic and short maxilla,
- Reduced sagittal depth of the nasopharynx,
- Reduced vertical growth of the maxilla.
- Vertebral anomalies includes malformations affected Cl or C2 and hypoplastic arches or abnormal ossification patterns,
- Reduced sella nasion distance,
- Length of the maxilla measured from the anterior surface of the maxilla to the posterior nasal spine is reduced [1,2,13,16].

According to Holmstroem et al., in Binder’s syndrome nasolabial angle has a value of 76–88° instead of the normal 103–117° (Segner and Hasund’s cephalometric analysis). The nasolabial angle, measured between the tip of the nose and upper lip. The acute nasolabial angle is mainly a result of the convexity of the upper lip, a deep fold or fossa between the nose and the upper lip, and a flat philtrum [7].

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Facial features</th>
<th>Other features</th>
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<tbody>
<tr>
<td>Chondroplasia punctata (CDP)</td>
<td>flat nose, flat face</td>
<td>scoliosis, shortening of ribs - usually asymetrical talipes</td>
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<tr>
<td>Robinow syndrome</td>
<td>flat nose, hypertelorism</td>
<td>short arms, macrocephaly, clinodactyly</td>
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<tr>
<td>Aarksog syndrome</td>
<td>flat nose, hypertelorism</td>
<td>clinodactyly, brachydactyly</td>
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<tr>
<td>Crouzon syndrome</td>
<td>hypoplastic maxilla, hypertelorism</td>
<td>short occipital-frontal diameter, craniosynostosis</td>
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<tr>
<td>Apert syndrome</td>
<td>flat nose, hypertelorism, hypoplastic maxilla</td>
<td>craniosynostosis, flat occiput, short occipital-frontal diameter, syndactyly, ventriculomegaly</td>
</tr>
<tr>
<td>Achondroplasia</td>
<td>low nasal bridge, mid-facial hypoplasia</td>
<td>megalcephaly, short tubular bones</td>
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<td>Fetal warfarin syndrome</td>
<td>flat face</td>
<td>Stipping</td>
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<td>Rudiger syndrome</td>
<td>flat nose</td>
<td>short digits</td>
</tr>
<tr>
<td>Stickler syndrome</td>
<td>flat nose, face, micrornathia, cleft palate</td>
<td>talipes, chondroplasia, osteo</td>
</tr>
<tr>
<td>Down syndrome</td>
<td>flat nose, flat face</td>
<td>macroglossia, congenital heart diseases, short stature, brushfield spots, microgenia, dermatoglyphs</td>
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<tr>
<td>Keutel syndrome</td>
<td>flat nose</td>
<td>abnormal cartilage calcification, neural hearing loss, peripheral pulmonary stenoses, brachytelphalangism</td>
</tr>
<tr>
<td>Desbuquois dysplasia</td>
<td>flat nose, flat face, monkey-like appearance</td>
<td>advanced bone maturation, vertebral changes [7].</td>
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Binder’s syndrome can be easily diagnosed with the help of 2D and 3D ultrasounds, beginning with the 21st week of pregnancy. It should be possible to distinguish each of these
syndromes on the basis of additional historical and clinical features (Table 1) [1,7].

Treatment includes Orthognathic and cosmetic surgery. Holmström and Kahnberg recommend a two-stage surgical procedure, first is maxillary osteotomy followed by the nasal improvement, both independent of the patient’s age [17]. Malalignment of teeth needs orthodontic treatment Maxillary retrognathia can be corrected by a Le Fort one maxillary advancement. Nasal septum, nasal bones and alteration of the glabellar depression are done in a Le Fort two osteotomy.

The onlay grafting technique like costal cartilage grafts for dorsal nasal augmentation, columnar lengthening, and premaxillary augmentation helps in facial growth [4].

Conclusion

Binder’s syndrome is a facial anomaly with hypoplasia of the nasomaxillary structures due to disturbance of growth in the prosencephalic induction centre. The patient must be kept under observation by the orthodontist as well as plastic surgeon since birth. Depending upon the severity of the malformation surgical intervention, nasal reconstruction, orthognathic as well as orthodontic treatment is necessary.

References