Rare disease

Binder's syndrome

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Binder syndrome or maxillonasal dysplasia is an uncommon developmental anomaly affecting primarily the anterior part of the maxilla and nasal complex. The characteristic findings are a failure of development in the premaxillary area with associated deformities of the nasal skeleton and the overlying soft tissues. Affected individuals typically have an unusually flat, underdeveloped midface (midfacial hypoplasia), with an abnormally short nose and flat nasal bridge, underdeveloped upper jaw, relatively protruding lower jaw and/or a 'reverse overbite' (or class III malocclusion). A 16-year-old female reported to clinic with chief complaint of facial deformity, on examination patient had classical feature of maxillonasal dysplasia. A brief review of the condition and the proposed multi-disciplinary treatment plan is discussed.

BACKGROUND

Effects on the facial features are: arhinoid face, intermaxillary hypoplasia (associated with malocclusion), abnormal position of the nasal bones, nasal mucosa atrophy, anterior nasal spine agenesis and (in most cases) a lack of frontal sinuses. Other deformities, as well as mental retardation, are also possible. Due to the clinical appearance, patients require surgical and orthodontic treatment. The main surgery performed in these patients is nose reconstruction with bone or cartilage grafts. Usually patients require more than one surgical procedure due to graft resorption and an unsatisfactory appearance. Orthodontic treatment is based on class III treatment (pseudo-mesio-occlusion) and relieving dental crowding. The treatment of malocclusion may require combined orthodontic and surgical treatment. In younger patients, maxillary protraction with rapid palatal expansion could be an adequate approach.

CASE PRESENTATION

A 16-year-old female reported to the clinic with chief complaint of facial deformity. There was no family history of similar complaint as well medical history was non-contributory. On extra oral examination, the patient had mesoprosopic face with hypoplasia of middle third of the face, a broad, flat nose, horizontal nostrils, a short columella and broad philtrum and a marked groove at the nasolabial junction (figure 1). On profile examination, the patient presented with a concave profile with straight divergence, depressed nose, decreased nasolabial angle and protruded both upper and lower lips; lower third of the face appeared to be normal (figure 2). Intraoral examination reveals class I incisors in relation with Angle's class I molars and canine. There was positive overjet and overbite with lower midline shifted to the left by 1 mm (figure 3). On cephalometric examination, the patient had classical features of skeletal class III malocclusion on account of point A-Nasion-Point B angle of -4.5° and Witwatersand appraisal of -15 mm. This is due to the reduced size (unit maxillary length of 39 mm, normal value 52 mm) as well as posterior position of the maxilla

(N $\perp$ to Pt A -5 mm) whereas the size and position of mandible was normal. The maxillo-mandibular differential was 8 mm as suggested by position of A-B point on Frankfurt horizontal plane value of -4 mm as compare to normal value of $+4$ mm. Maxillary incisor were proclined as suggested by one (maxillary central incisor) to relationship of point A to Nasion perpendicular value of 44° and 17 mm, Sella-Nasion to one value of 121° (normal value 102°), one to palatal plane value 133° (normal value 110°). Position of mandibular incisors was normal. This may be

Figure 1 Frontal view.

Figure 2 Lateral view.

due to the dentoalveolar compensation of the maxillary deficiency (figures 4 and 5).

INVESTIGATIONS

- ▶ Lateral cephalogram with cephalometric analysis
- ▶ Photographs
- ▶ Study model.

DIFFERENTIAL DIAGNOSIS

- ▶ Warfarin embryopathy
- ▶ Down's syndrome
- ▶ Apert syndrome
- ▶ Stickler syndrome
- ▶ Keutel syndrome
- ▶ Acrodysostosis.

TREATMENT

The aims of the treatment were:

- ▶ correction of facial deformity
- ▶ correction of nasal depression
- ▶ to correct proclination of maxillary incisors
- ▶ correct the lower centreline
- ▶ to obtain a good class I canine and molar occlusion.

An interdisciplinary treatment was planned in collaboration with orthodontist and oral and maxillofacial surgeon.

The treatment plan was as follows:

- ▶ oral prophylaxis to improve oral hygiene.
- ▶ orthodontic treatment to decompensate the malocclusion—extraction of maxillary first bicuspid to retract maxillary incisors, orthodontic micro implants will be

Figure 3 Occlusal relationship.

use to maximise the incisors retraction and to achieve the maximum reverse overjet as possible so as to facilitate the maximum surgical advancement of the maxilla.

- ▶ orthognathic surgery (Le Fort I maxillary advancement of 8 mm) to correct maxillary deficiency.
- ▶ post surgical orthodontic treatment to achieve finishing and detailing for proper intercuspation.
- ▶ Rhinoplasty by costal cartilage graft.

DISCUSSION

Zuckermandl in 1882 described an anomaly in the anterior nasal floor in which the normal crest that separates the nasal floor from the anterior surface of the maxilla was absent, and instead, a small pit (the fossa prenasalis) constituted the inferior margin of the piriform aperture.¹ In 1939, Noyes² described a patient with a flat nasal tip sitting on a retruded maxillonasal base, and in 1962, von Binder³ 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. von Binder postulated that these defects were caused by rhinocephalic dysplasia, which he called 'maxillonasal dysostosis'. Since that time, the condition has been known as maxillonasal dysplasia or Binder syndrome.⁴ Posnick and Thompson⁵ noted that the physical findings of Binder syndrome are the result of hypoplasia (depression) of the anterior nasal floor (fossa prenasalis) and localised symmetric maxillary hypoplasia of the alar rim regions. From the basal view, typical variations from normal include a retracted columella-lip junction; lack of normal triangular flare at the nasal base, a perpendicular alar-cheek junction; convex upper nasal tip with a wide, shallow philtrum; crescent-shaped nostrils without a sill; low-set and flat nasal tip; and stretched and shallow Cupid's bow.⁶ The most striking characteristics of the nose are vertical shortening, lack of tip projection, perialar flattening and an acute nasolabial angle.

Holmstrom found a hereditary connection in 16 per cent of 50 patients with Binder syndrome, and inheritance may be as an autosomal recessive trait with incomplete penetrance.⁶⁻⁸

This feature is caused by a disturbance of growth in the prosencephalic induction centre. The process of nose

Figure 4 Lateral cephalogram.

formation normally takes place during the third month of pregnancy.^{9 10} Males and females are equally affected by Binder's syndrome.

Beside hypoplasia of the nose, patients may present bilateral loss of hearing and mental retardation (although there is no proof that Binder's syndrome is associated with lower intelligence).¹¹ The majority of cases are isolated, but there are some reports of family recurrence of the syndrome (among siblings or parents and children); a positive family history was reported in 36% of the subjects of one study.^{12 13} This suggests that the inheritance pattern is either autosomal recessive with reduced penetrance or multifactorial.^{9 11–13} There are reports that Binder's syndrome has a direct association with vitamin K deficiency.^{9 14} In some cases, other congenital diseases and abnormalities (such as Down's syndrome, autonomic neuropathy and strabismus) are observed.¹¹ According to Nedev, 5% of patients are found to present hearing loss and the same number of patients present congenital heart diseases.⁹

The nasolabial angle, measured between the tip of the nose and upper lip, is acute. According to Holmstroem *et al.*, in Binder's syndrome it has a value of 76°–88° instead of the normal 103°–117° (according to Segner and Hasund's cephalometric analysis). 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.^{3 9–11 15}

Delaire *et al.* observed that most Binder's syndrome patients suffer from microdontia of the central upper incisors; a lack of lateral incisors has also been observed, but rarely.^{11 16}

In addition to skeletal changes in the skull, about 44.2% of Binder's syndrome patients present irregularities in the cervical spine, which arise during those structures' development in the third month of pregnancy – the same period

Figure 5 Cephalogram tracing.

as nose formation. Most frequently the C1 and C2 vertebrae are affected. Their hypoplastic arches may present abnormal patterns of ossification.^{8 9 11} The most frequent anomalies in vertebral structures are separate odontoid process, a short posterior arch, spina bifida occulta and blocked vertebrae.⁹

The management of these patients depends on the level of complexity because of variations in the midface discrepancy and the occlusal relationship. Some lesser-affected patients can be managed by using orthodontic camouflage alone, whereas others require multi-disciplinary input. The effects of posteroanterior traction, used to improve the facial aesthetics of an 11-year-old boy with maxillo-nasal dysplasia, were analysed; improvement in maxillary retrusion vis-à-vis the mandible only was accomplished, and the recorded advancement of the maxillary bones was slight (0.6 mm).¹⁷

Osteotomy and grafts (bone or cartilage) are frequently used in the management of Binder syndrome. Grafts, when limited to the nasal sill, do not address the 'dish face' deformity characteristic of the syndrome. LeFort II osteotomy, although correcting the midface deficiency, can overcorrect the occlusal relationship if good dental camouflage has occurred, making a LeFort I osteotomy or orthodontic therapy necessary. It has been suggested that mild cases of Binder syndrome, without severe malocclusion, can be managed by using a cartilage graft around the nostril sill, dorsum, and the pyriform aperture, correcting the paranasal and infraorbital deformity without osteotomy.¹⁸ In more severe cases, however, a LeFort II osteotomy can correct the midface deficiency and the class III occlusion seen in 15% of patients with Binder syndrome, but it does not always fully address the nasal flatness. Twenty-seven patients with maxillonasal dysplasia of various degrees were examined on both short- and long-term bases.¹⁹ For some patients, surgical treatment began at age 3, but others were treated as teenagers or young adults. The overall goals were to augment the skeletal deficiencies of the mid-face and begin the soft-tissue expansion process as early as possible.

Learning points

- ▶ Binder's syndrome is a rare congenital malformation that mainly affects facial features.
- ▶ Ultrasound screening and non-invasive genetic tests are performed to look for congenital anomalies as well as assessment of gestational age.
- ▶ The timing and types of procedures involved in the treatment of patients with maxillonasal dysplasia depend on the severity of the malformation and are planned individually.
- ▶ The optimum care of these patients requires an interdisciplinary approach; therefore all patients should be subjected to a thorough treatment planning in collaboration between orthodontists and oral and maxillofacial surgeons and/or plastic surgeon.

Competing interests None.

Patient consent Obtained.

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