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Case Report

Binder's syndrome: a rare disease

Chandana Athmananda, Kowsthuba Narayan, Praveen Unki*

Department of Paediatrics, Adhichunchanagiri Institute of Medical Sciences, Mandya, Karnataka, India

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***Correspondence:**
Dr. Praveen Unki,
E-mail: praveenu3@gmail.com

ABSTRACT

Von Binder described a rare congenital developmental disorder in 1962. It is a developmental disorder which is affecting the maxillary anterior region and nasal complex (nose and jaw) hence he gave the name maxillonasal dysplasia. Binder gave six classical characteristic features: arhinoid face, maxillary hypoplasia, reduced or absent nasal bridge, malformation of nasal bones, hypertrophy of turbinates with atrophy of nasal mucosa, absent frontal sinus (not compulsory). We reported a 8 years old female child presented with complaints of history of bilateral hearing loss and stuttering noticed from 2 years of age with history of delayed language and milestone development with below average academic performance and examination findings showed crescent shaped face, hypertelorism, flattened nasal bridge and ear examination findings showed low set ears. Oropharynx examination showed high arched palate, palatal scar and bifid uvula. Diagnostic nasal endoscopy showed bilateral hypertrophy of inferior turbinates and atrophy of nasal mucosa, pure tone audiometry done which shows B/L sensorineural hearing loss. Binder's syndrome is a rare congenital malformation that mainly affects facial features. Treatment planning requires nose reconstruction first of all, but sometimes other medical care (such as orthodontic and orthognatic treatment) is also necessary. 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.

Keywords: Binder's syndrome, Nasomaxillary dysplasia, Congenital anomalies, Arhinoid face

INTRODUCTION

Von Binder described a rare congenital developmental disorder in 1962. It is a developmental disorder which is affecting the maxillary anterior region and nasal complex (nose and jaw) hence he gave the name maxillonasal dysplasia.

Binder gave six classical characteristic features: arhinoid face, maxillary hypoplasia, reduced or absent nasal bridge, malformation of nasal bones, hypertrophy of turbinates with atrophy of nasal mucosa; absent frontal sinus (not compulsory).

Individuals with these classical facial features are easily identifiable. They might also have flat short nose, by Algarswamy 2016 upper jaw, short columella, convex upper lip, broad philtrum, malocclusion/crowding of teeth, pseudoprogathism by Mehrotra 2019.[1,2] Apart from facial features child can also have decreased hearing and decreased intelligence with misalignment of eye-strabismus.

CASE REPORT

An 8-year-old female presented with history of decreased hearing capacity in bilateral ears and stuttering noticed from 2 years of age. The child was born to a 3rd degree consanguineous married couple, with cleft palate for which she was operated 6 years back. As the child had deficiency of bilateral alar cartilage for which silicone implantation-dorsal augmentation of nose was done 4

years back. There was no other relative antenatal/prenatal exposure to long term drug intake history. History was suggestive of delayed language and social milestone development with below average academic performance. Child also had history of social anxiety.

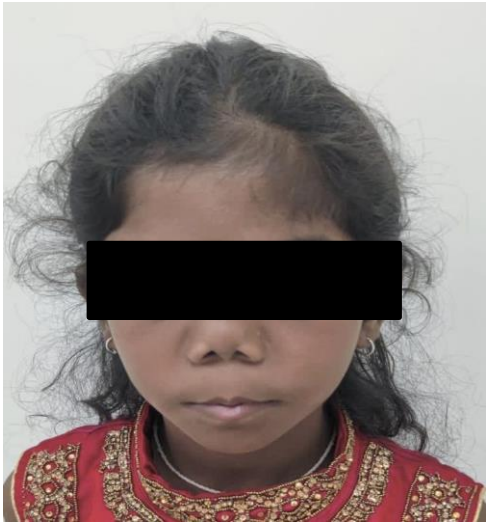

Figure 1: Arhinoid face, maxillary hypoplasia, reduced or absent nasal bridge, malformation of nasal bones, hypertrophy of turbinates with atrophy of nasal mucosa, and absent frontal sinus (not compulsory).

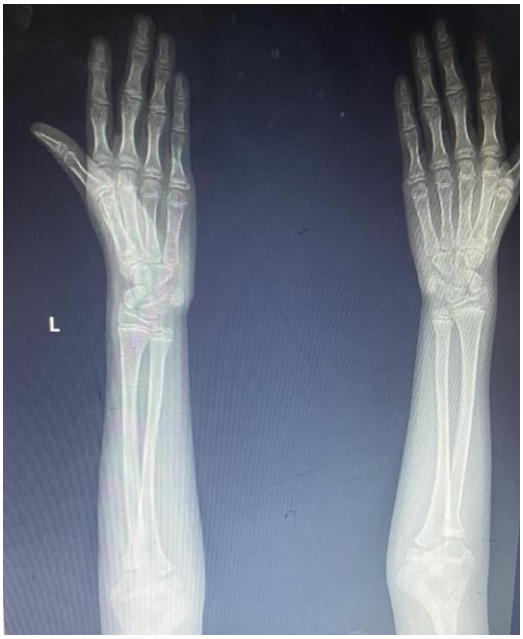

Figure 2: X-ray of forearm and hand, increased distance between the radial and ulnar bones and left palm shows overlapping of the carpal bone and proximal phalanx.

On examination child was inspected and mid face showed hypertelorism, flattened nasal bridge with long philtrum, crescent shaped nostrils, with deficiency of bilateral alar cartilage. Ear examination showed low set ears with double tragus. Oropharynx examination showed crowding of teeth, high arched palate with palatal scar and bifid uvula. Further head to toe examination showed pectus excavatum and abnormal carrying angle (which was increased). Extremities showed atrophy of the thenar eminence, flat foot with valgus deformity.

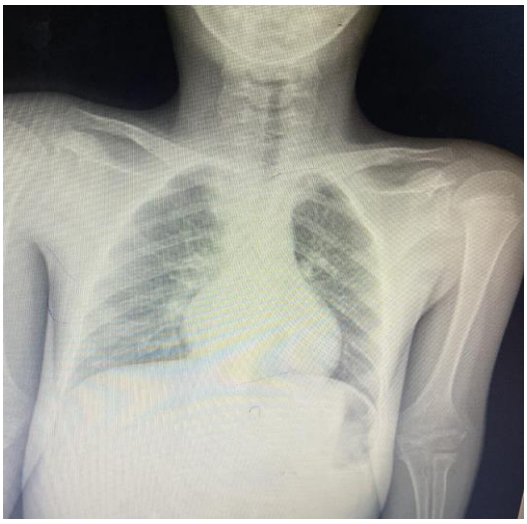

Figure 3: Radiographs showing normal X-ray skeleton.

Systemic examination showed no abnormalities. The child was evaluated further to rule out any other congenital anomalies. Following clinical examination diagnostic nasal endoscopy was done and found to have bilateral hypertrophy of inferior turbinates and atrophy of nasal mucosa. Pure tone audiometry was done and found to have bilateral moderately severe sensory neural hearing loss with right ear: 60 dBHL and left ear: 65 dBHL. Echocardiogram was within normal limits. Skeletal radiographs were also normal.

In view of delayed milestones, child was further evaluated and found to have borderline intelligence with mild to moderate intellectual disability. Hearing aid was given for hearing loss. Psychiatric counselling was done in regular follow ups.

Differential diagnosis

The differential diagnosis for Binder's syndrome was based on similar facial features in other syndromes. This included warfarin embryopathy, Down's syndrome, Apert syndrome, Stickler syndrome, Keutel syndrome and acrodysostosis. These syndromes can frequently be misdiagnosed during fetal ultrasonogram (USG) examinations. Binder's syndrome can be easily diagnosed by using 2D and 3D ultrasounds, beginning with the 21st week of pregnancy. Early diagnosis was important, as nasal malformations are present in 1:1600 fetuses and may play an essential role in the early diagnostics of congenital diseases and syndromes.

DISCUSSION

Binder's syndrome is a under development of mid-facial skeleton which is involving nasomaxillary complex. Von Binder reported 3 cases in 1962, also Hopkins reported 5 cases in 1963 and gave classical facial features. This was seen in less than 1 per 10,000 live births, exact incidence was not known as many patients may be undiagnosed or misdiagnosed.

The root cause which was given by Binder was disturbance of prosencephalic induction of centre, but this may be seen because of birth trauma, vitamin K deficiency during embryonic growth. Few studies also said that it might be genetic-autosomal recessive trait with incomplete penetration, but both the parents should carry the trait to pass it down. In few cases there was a history of pregnancy exposure to lithium, ethanol, phenytoin and warfarin.

Prenatal diagnosis of Binder's syndrome can be done with two dimensional and three-dimensional ultrasound scan at 21 weeks of gestation. First indication of any abnormality was flattened fetal nose demonstrated in sagittal plane. Binder's syndrome can also be associated with other congenital anomalies like congenital heart diseases, mental retardation, hearing loss. Association with chondrodysplasia punctata was noted in five cases. Naryctal in 1987 said that it might be associated with esophageal achlasia. Binder's syndrome should be differentiated from Robinow, Aarskog, Rudiger syndrome. Binder's syndrome was usually diagnosed clinically as it had characteristic facial features and correlated with further radiographs and investigations. This syndrome involved hypoplasia/dysplasia of nasomaxillary complex, which led to various degrees of deformity. Patients may present with characteristic features like hypoplasia of nasomaxillary complex associated with hypertelorism, flattened nasal bridge, abnormal positioning of nasal bones, nasal mucosa atrophy, convex upper lip, malalignment of teeth, short columella, reduced naso-labial angle. There can also be absent or hypoplasia of frontal sinus but can be only seen in 44-50% of the cases. Some cases of Binder's can be associated with other congenital anomalies like Down's syndrome. Some children might also have mental retardation or developmental delay. In our case report, Binder's syndrome was diagnosed based on the symptoms which suggestive of prosencephalic centre deformity such as cleft palate, absent nasal bridge, high arched palate (midline structural abnormalities). Surgical correction included corrections of cleft palate, Lefort I and II osteotomy with nasal grafting, maxillonasal hypoplasia can be treated with graft replacement. Orthodontic treatment for cosmetic reason would be supportive. Rhinoplasty is planned in near future.

Compared to the study of Mehrotra et al our study had involved the clinical management by the combination of pediatrician, plastic surgeon and pediatric surgeon for the better outcome both surgically and medically and was going to be having better results compare to the study which was conducted at 2016.[1]

Compared to the study conducted by Algarswamy et al our study had involved even the pediatric surgeon and ENT surgery team involved in looking after the surgical prognosis along with plastic surgeon which gave early and better results.[2]

Compared to the Jain et al our study had better prognosis because of involving pediatricians who had a regular follow up of the case and for a better prognosis level.[3]

Compared to Roa et al my team was able to diagnose the defect early and treat both medically and surgically and gives a better outcome.[5]

CONCLUSION

Binder's syndrome is a rare congenital malformation that mainly affects facial features. Treatment planning requires nose reconstruction first of all, but sometimes other medical care (such as orthodontic and orthognatic treatment) is also necessary. From the day the child is born he or she should be under the care of a plastic surgeon and orthodontist. 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.

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