Case Report

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BINDER'S MAXILLO-NASAL DYSPLASIA WITH PHALANGEAL HYPOPLASIA

Anshul Mahajan[1], Sujoy Ghosh[2], Mahesh Verma[3], Sunita Gupta[4]

1.Post-graduate Student, Department of Oral Medicine & Radiology Maulana Azad Institute of Dental Sciences New Delhi, India.
2.Associate Professor, Department of Oral Medicine & Radiology Maulana Azad Institute of Dental Sciences New Delhi, India
3.Director, Principal & Professor and Head, Department of Prosthodontics Maulana Azad Institute of Dental Sciences New Delhi, India
4.Professor & Head, Department of Oral Medicine & Radiology, Maulana Azad Institute of Dental Sciences, New Delhi, India.

ABSTRACT:

Binder's syndrome is a rare developmental disorder that affects the anterior maxillary and nasal region. The epidemiological and etiologic status of this syndrome is still poorly understood owing to the lack of frequency data. Characteristic features of Binder's syndrome include a concave profile with hypoplastic nose and maxilla, short columella and acute nasolabial angle. In addition, vertebral anomalies and phalangeal deformities can also be associated with this syndrome. We report a case of Binder's syndrome with terminal phalangeal hypoplasia highlighting the radiographic features and discussing its differential diagnosis.

Keywords: Binder, Maxillo-nasal Dysplasia, Phalangeal, Hypoplasia

INTRODUCTION

Binder's syndrome (BS) is an uncommon developmental anomaly affecting the midface region predominantly the anterior maxilla and the nasal complex. [1] Although first reported by Noyes in 1939, it was described as a distinct clinical syndrome by Binder in 1962.[1,2] A characteristic facial appearance comprising of abnormally short nose and flat nasal bridge, underdeveloped upper jaw, and a relatively protruding lower jaw (class III malocclusion) is seen in individuals with Binders syndrome.[3] Additionally, vertebral anomalies along with deformities of phalanges have been found to be associated with BS.[1,3] We report such a case of Binder's syndrome with terminal phalangeal hypoplasia discussing the clinical and radiologic features differentiating it from other possible conditions

CASE DETAIL:

A 10 year old male patient reported to the department of Oral Medicine & Radiology with a complaint of decayed teeth. Physical examination of the patient showed short stature, short and stubby fingers and toes, speech disability and mental retardation. Extra oral examination revealed a deficient midface profile with a broad forehead, saddle nose with crescent shaped nostrils, short columella, convex upper lip, broad philtrum and acute nasolabial angle (Fig 1A-1E). Intra oral examination revealed a high arched palate with proclined maxillary and

*Corresponding Author Address: Dr. Anshul Mahajan Email: anshulmahajan@hotmail.com

mandibular anterior teeth and anterior open bite. All the four permanent first molars were carious and the permanent maxillary central incisors showed evidence of enamel hypoplasia at the incisal edges (Fig 1F). A lateral skull radiograph revealed absence of anterior nasal spine and anterior clinoid process with hypoplasia of nasal bone and maxilla (Fig 2). Radiographs of the hands and feet showed hypoplasia of the distal phalanges (Fig 3, 4). Owing to the clinical and radiologic features a diagnosis of BS was made.

DISCUSSION:

Binder's syndrome or maxillonasal dysplasia is described as an uncommon anomaly mainly affecting the middle third of the face.[4] The etiopathogeneis of BS still remains under a cloud of doubt owing to the paucity of population frequency data and various theories have been put forward to explain it. Noyes suggested birth trauma as a possible cause but could not explain the absence of anterior nasal spine as a result of it.[2,4] Binder suggested that there was a disturbance of the prosencephalic induction centre during embryonic growth. [5] Hopkin on the other hand advocated developmental injury as an etiologic factor. [2,4] Another theory suggests the role of drugs causing vitamin K deficiency during pregnancy which induces maxillonasal hypoplasia and abnormal cartilage calcification.[4] Warfarin, a coumarin anticoagulant can lead to agenesis of corpus callosum and midline facial deformity.[4] Striker suggested an early vomeral premaxillary synostosis as the cause of midface hypoplasia whereas Narcy suggested abnormal neural crest migration during embryonic development as the probable etiologic factor.[2,4]

An individual with BS has a characteristic facial appearance comprising of a flattened nose with crescent shaped nostrils, short and retracted columella, convex upper lip with flat philtrum and acute nasolabial angle.[2,3] The skeletal changes include hypoplastic anterior nasal spine resulting in flattening of the skeletal nasal prominence with a hypoplastic maxilla. These features were also consistent with our case. The anterior cranial base is short with a posteriorly positioned maxilla and prognathic mandible producing a Class III malocclusion. Additionally, anomalies in the vertebral structures such as separate odontoid process, short posterior arch, spina bifida occulta and blocked vertebrae can also be seen none of which were noticed in our case.[1,3] Terminal phalangeal hypoplasia is another skeletal deformity that maybe associated with BS which was also seen in the digits of both hands and feet in our case.[1] Other associated anomalies include orofacial clefting, strabismus, mental retardation, disturbed speech and sense of smell, hearing loss and non-specific congenital heart defects. [1] In our case mental retardation with disturbed speech were evident. These facial and skeletal features necessitate a clear understanding of the differential diagnosis of BS which includes chondrodysplasia punctata, fetal warfarin embryopathy,

Down's syndrome, Stickler syndrome and Keutel syndrome (Table 1).[2,3,6-10]

Management of patients with BS should be an interdisciplinary approach including orthognathic and plastic surgery along with orthodontics to correct the facial, skeletal and dental anomalies. The surgical modalities involve nasal dorsum reconstruction elevating the tip of the nose and lengthening the nasal dorsum.[3] Severe skeletal class III malocclusion can be corrected by Le Fort I osteotomy with orthodontic therapy whereas mild cases of malocclusion can be corrected by orthodontic therapy alone. [2,3] Along with esthetic and functional management psychological and speech therapy should also be instituted if the need arises.

CONCLUSION:

Binder's syndrome is a rare congenital anomaly affecting the nasomaxillary structures. A precise clinical and radiologic examination can aid in diagnosing the condition accurately and in a timely manner. Management of patients with BS should include a multi-disciplinary approach with the goal of providing esthetic and functional quality of life.

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TABLE:

Table 1. Differential Diagnosis Of Binder Syndrome [6-10]

Syndrome	Features similar to Binder's syndrome	Differentiating features
Chondrodysplasia punctata	Flat face, saddle nose, short extremities, brachytelephalangism, vertebral anomalies.	Joint contractures, congenital cataracts, ichthyotic skin lesions, epiphyseal stippling, tracheal & laryngeal calcifications.
Fetal warfarin embryopathy	Flat face, nasal hypoplasia, hypoplasia of distal phalanges, short extremities, mental retardation.	Epiphyseal stippling of vertebrae and long bones, optic atrophy, blindness, corneal opacity, deafness.
Down syndrome	Flat face, flat nose, prognathism, high arched palate, mental retardation, disturbed speech.	Macroglossia, cardiovascular, hematopoietic and neuromuscular anomalies.
Keutel syndrome	Flat face, brachytelephalangism, short stature, mental retardation.	Abnormal cartilage calcification, hearing loss, peripheral pulmonary stenosis, respiratory problems.
Stickler syndrome	Midface hypoplasia, flat nose, anteverted nares, high arched palate and vertebral anomalies.	Prominent eyes with myopia, retinal degeneration, sensorineural deafness

FIGURES:

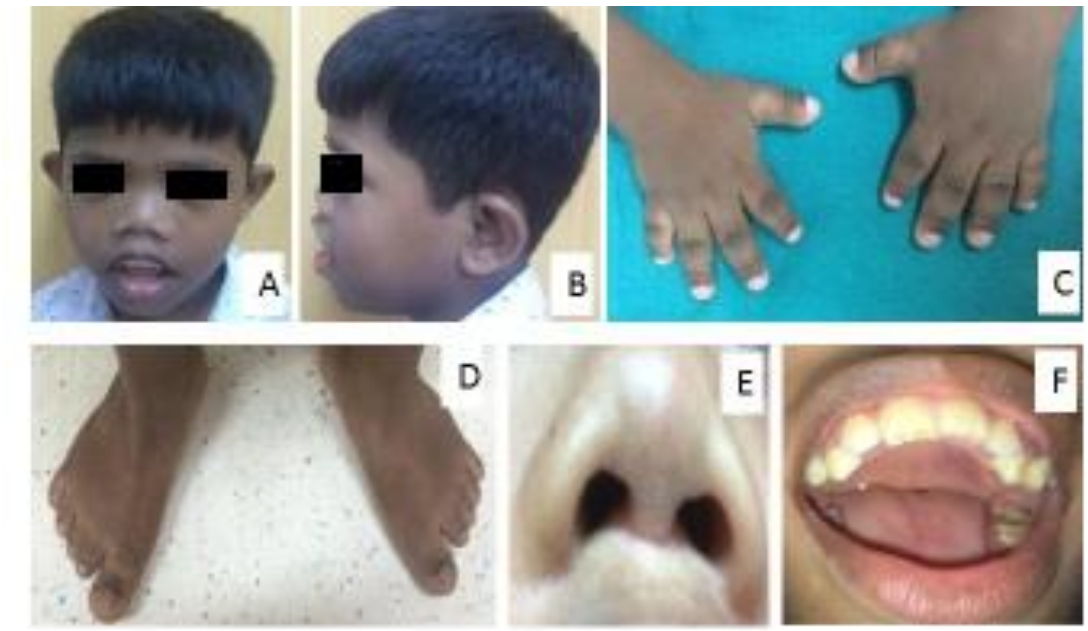

Figure 1: A), B) Extra oral photograph depicting a deficient midface profile, saddle nose, short columella, convex upper lip, broad philtrum and acute nasolabial angle C), D) short and stubby fingers and toes E) crescent shaped nostrils F) proclined maxillary anterior teeth with hypoplastic enamel at the incisal edges.

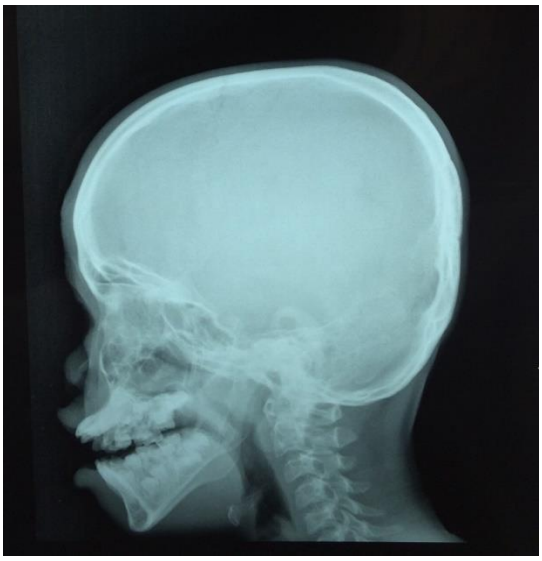

Figure 2: Lateral skull radiograph showing hypoplasia of nasal bone and maxilla with absence of anterior nasal spine and anterior clinoid process

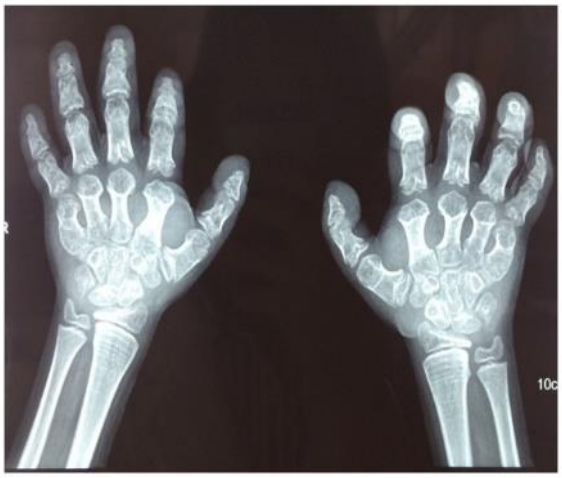

Figure 3: Radiograph of the hands showing hypoplasia of the distal phalanges

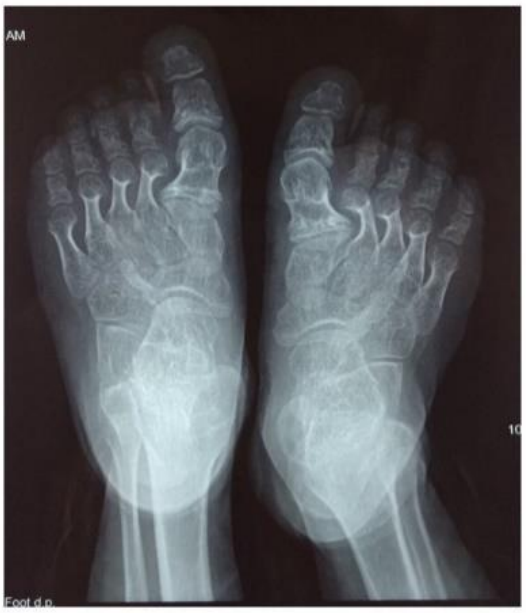

Figure 4: Radiograph of the feet showing hypoplasia of the distal phalanges