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Case Report

A CASE OF ANTLEY-BIXLER SYNDROME WITH SEVERE SKELETAL CL. III MALOCCLUSION

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Abstract

Antley-Bixler syndrome is a disorder characterized by craniosynostosis, midface hypoplasia, choana blockade, and radiohumeral synostosis. However, the features of occlusion remain unclear.

In this paper, we report a case of Antley-Bixler syndrome, a 7-year-old boy, from the viewpoint of orthodontics. From lateral cephalometric head film analysis, remarkable retardation of the anterior subcranial base, infraorbitale, and maxilla were notable, as was vertical growth restriction of the maxilla. The choana blockade tendency was also recognized.

Moreover, although reverse occlusion was present, a mandibular retrognathic tendency was also present, and a short ramus mandible, remarkable mandibular vertical growth pattern, and skeletal open bite were present. In the dentition, two of the lower incisors were missing, and the present lower incisors were large. Maxillary and mandibular first molars were delayed in eruption.

For treatment, the solutions to such remarkable skeletal problems were limited by the insufficiency of recovery of cranial formation after the operation. We planned a non-surgical treatment to expand the maxilla.

It will be necessary to continually consider the treatment of his malocclusion as he continues to grow.

Key words: Antley-Bixler syndrome—Craniosynostosis—Midface hypoplasia—Radiohumeral synostosis—Skeletal problems

INTRODUCTION

Antley-Bixler syndrome, first reported by Antley and Bixler in 1975\(^7\), is characterized by multiple synostoses, including craniosynostosis, choanal atresia, and humeral synostosis\(^5\). The patients with craniofacial deformities usually have severe complex malocclusions, so the importance of orthodontics for such patients is increased. We need to understand the
dental situation of such patients, but there have been only few reports of the dental findings in cases of Antley-Bixler syndrome. Recently, we encountered a case with this syndrome, and the dental findings in that patient are reported here.

CASE PRESENTATION

This boy has undergone orthodontic treatment and dental management at the Department of Orthodontics, Suidobashi Hospital of Tokyo Dental College, since he was 7 years and 1 month old. He had been diagnosed to have Antley-Bixler syndrome at Saitama Children’s Medical Center. His height was 113 cm, his body weight was 15 kg, and developmental arrest was clear. He had been operated on for hydrocephalus at the age of 4 months, for plagiocephaly at the age of 18 months, and for inguinal hernia at the age of 3 years. The major findings on physical examination included frontal bossing, mid-face hypoplasia, a slight protrusion of the eyes, and a depressed nasal bridge (Fig. 1). The oral fissure was small; narrow and crowded upper and lower dentition, total cross bite, and open bite were noted. In the dentition, the two mandibular incisors were missing bilaterally, the present lower incisors were large, and mesioversion of the maxillary first bilateral molar was noted (Figs. 2, 3, and 4). In the deciduous dentition, fusion of
the left mandibular deciduous central incisor and the lateral incisor was observed. The bone age corresponded almost to the chronologic age of seven years old. However, the osteoepiphysis formation of the digitus was accelerated, and the growth of the carpus was delayed (Fig. 5).

Cephalometric roentgenography revealed marked restriction of the anterior base of the skull, orbits, and maxilla, and restriction of vertical growth of the maxilla. A tendency toward choanal atresia was also observed. The mandible exhibited reverse bite, and a tendency forward restriction was noted. Vertically, a pattern of marked mandibular vertical growth with the short ramus of the mandible was noted, and skeletal open bite was observed (Table 1, Figs. 6, 7, 8 and 9).

For surgical orthodontic treatment in such a case with marked skeletal deformity, distraction osteogenesis of mid-face bones was considered, but discontinuity was noted after cranioplasty. Therefore, bone induction could not be done, and attachment of a device to the cranial bone was considered difficult. Single-stage of anteroinferior improvement by Le Fort III-type osteotomy was proposed, but informed consent could not be obtained from the patient and his parents, in view of the bone transplantation and surgical intervention involved. Only orthodontic lateral expansion of the maxillary arch is in progress.
### DISCUSSION

Antley-Bixler syndrome is primarily characterized by craniosynostosis, short skull, midface hypoplasia, choanal atresia, radiohumeral synostosis, and multiple arthrogryposis (Table 2). In addition, hydrocephalus, contracted pelvis, and contracted thorax may also be observed clinically\(^5\). In girls, external genital anomalies such as absence of the vagina and hypoplasia of the labia majora are commonly noted\(^7\). Severe secondary

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**Table 1 Roentgen cephalometric analysis**

<table>
<thead>
<tr>
<th>Mean</th>
<th>S. D.</th>
<th>M.V.</th>
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<tbody>
<tr>
<td>1. Facial angle</td>
<td>86.1</td>
<td>3.3</td>
</tr>
<tr>
<td>2. Convexity</td>
<td>6.4</td>
<td>3.0</td>
</tr>
<tr>
<td>3. A-B plane</td>
<td>-5.2</td>
<td>2.5</td>
</tr>
<tr>
<td>4. Mandibular plane</td>
<td>24.8</td>
<td>5.9</td>
</tr>
<tr>
<td>5. Y-axis</td>
<td>64.0</td>
<td>3.1</td>
</tr>
<tr>
<td>6. Occlusal plane</td>
<td>8.4</td>
<td>4.2</td>
</tr>
<tr>
<td>7. Interincisal</td>
<td>131.6</td>
<td>5.6</td>
</tr>
<tr>
<td>8. L-1 to Occlusal</td>
<td>21.3</td>
<td>5.3</td>
</tr>
<tr>
<td>9. L-1 to Mandibular</td>
<td>97.1</td>
<td>4.9</td>
</tr>
<tr>
<td>10. U-1 to AP plane</td>
<td>7.8</td>
<td>2.5</td>
</tr>
<tr>
<td>11. FH to SN plane</td>
<td>5.4</td>
<td>2.4</td>
</tr>
<tr>
<td>12. SN A</td>
<td>83.4</td>
<td>2.6</td>
</tr>
<tr>
<td>13. SN B</td>
<td>80.0</td>
<td>2.5</td>
</tr>
<tr>
<td>14. SNA-SNB diff.</td>
<td>3.4</td>
<td>1.7</td>
</tr>
<tr>
<td>15. U-1 to FH plane</td>
<td>110.8</td>
<td>5.6</td>
</tr>
<tr>
<td>16. L-1 to FH plane</td>
<td>61.6</td>
<td>5.8</td>
</tr>
<tr>
<td>17. Gonial angle</td>
<td>117.5</td>
<td>8.1</td>
</tr>
<tr>
<td>18. Ramus angle</td>
<td>88.5</td>
<td>4.5</td>
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</table>

Restriction of maxilla and hyper-divergent pattern of mandible are recognized.
respiratory tract disorders may be associated with the airway closure. Since apnea is associated with a poor prognosis, early countermeasures are necessary. Systemic management is expected to improve the prognosis for mental development.

No congenital anomalies, including cleft palate, were seen in any of the patient’s family members or siblings. The onset of this syndrome has been reported to be associated with marriage between siblings or blood relatives and autosomally recessive inheritance.
Table 2 Clinical synopsis

<table>
<thead>
<tr>
<th>Group (II)</th>
<th>Our Case</th>
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<tr>
<td>I. 5y2m (4y3m—5y11m)</td>
<td>0</td>
</tr>
<tr>
<td>II. 7y8m (6y2m—8y11m)</td>
<td>0</td>
</tr>
<tr>
<td>III. 10y1m (9y0m—11y10m)</td>
<td>0</td>
</tr>
<tr>
<td>IV. 12y11m (12y3m—14y4m)</td>
<td>0</td>
</tr>
<tr>
<td>V. 23y7m (19y11m—28y11m)</td>
<td>0</td>
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Head and Neck:

- **Head**
  - Brachycephaly
  - Large anterior fontanelle

- **Face**
  - Frontal bossing
  - Midface hypoplasia
  - Long philtrum

- **Ears**
  - Dysplastic ears
  - Stenotic external auditory canals

- **Eyes**
  - Proptosis

- **Nose**
  - Depressed nasal bridge
  - Choanal atresia or stenosis

- **Tooth**
  - missing
  - narrow dental arch
  - reverse bite

Skeletal:

- **Skull**
  - Craniosynostosis, coronal and lambdoidal

- **Limbs**
  - Radioulnar synostosis
  - Femoral bowing
  - Neonatal femoral fractures
  - Ulnar bowing
  - Joint contractures

- **Hands**
  - Arachnodactyly
  - Camptodactyly

Neurologic:

- **Central nervous system**
  - Variable mental retardation
  - Hydrocephalus

*: Chondral radioulnar synostosis
0: present
—: absent
+/-: not definite
modified from Escobar et al.2

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has been suggested. In other types of craniosynostosis such as Crouzon’s syndrome, mutation of the FGFR2 gene has been reported. Abnormality of the FGFR2 gene has also been noted in some cases of the Antley-Bixler syndrome. On the other hand, the possible involvement of some abnormality of steroid metabolism in the onset of this syndrome has also been suggested. Intake of the antifungal drug fluconazole by the mother during pregnancy has been suggested to lead to an increased incidence of the birth of babies with features of this syndrome. None of these factors were identified in our patient.

The severity of the radioulnar synostosis has been reported to vary widely among patients, and humeral synostosis is not believed to be an important criterion for diagnosing this disease. Synostosis of the elbow joint was not observed in our patient; nevertheless the range of motion of this joint was restricted. Radioulnar synostosis was suspected, but a definite diagnosis could not be established.

Koshiba et al. observed missing maxillary second premolar, narrowing of the maxillary arch width, tendency toward Skeletal Class III enlargement of the mandibular angle, and retraction of the chin in a patient of Antley-Bixler syndrome. Our patient also exhibited similar features; narrow maxillary arch, Class III malocclusion, and missing teeth. Adhesion of the mandibular deciduous incisors and missing the permanent successors in normal children has been reported by Tsujino et al.

According to their report, in the 152 children who had fused deciduous teeth, the cases with fusion of mandibular central and lateral
deciduous incisors showed a relatively high percentage (39.6%) and 11.1% of them were congenitally missing their permanent successors. Congenital missing of permanent teeth was also noted in our patient.

There have been no reports regarding the treatment of malocclusion in these cases. Lateral expansion and anterior traction of the maxillary arrangement of teeth were considered as non-surgical orthodontic treatment approaches. However, a maxillary anterior traction device inevitably becomes a burden on the calvarium, and such a load must be avoided. Therefore, for the time being, only lateral expansion of the maxillary arch is in progress. We think that there is need to establish the treatment planes for marked restriction and vertical suppression of the maxilla while paying attention to the response to the lateral expansion.

ACKNOWLEDGEMENTS

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REFERENCES


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