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 1975¹⁾, is characterized by multiple synostoses, including craniosynostosis,

choanal atresia, and humeral synostosis⁴⁾. 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

Fig. 1 Facial photographs
Midface hypoplasia, frontal bossing, and deep nasal bridge are recognized.

dental situation of such patients, but there have been only few reports of the dental findings in cases of Antley-Bixler syndrome⁶⁾. 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 Center⁸⁾. 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, midface 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

Fig. 2 Oral photographs
Upper and lower arches are crowded, and lower incisors are large.

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

Fig. 3 Oral photographs
Total cross bite and open bite are recognized.

Fig. 4 Panoramic radiographs
Two mandibular incisors are missing.

Fig. 5 Carpal bones' radiograph
Retardation of carpal bones are recognized.

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 toward 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.

Table 1 Roentgen cephalometric analysis

	Mean	S. D.	M.V.	
1. Facial angle	86.1	3.3	(80.0)	
2. Convexity	6.4	3.0	(-3.9)	
3. A-B plane	-5.2	2.5	(6.0)	
4. Mandibular plane	24.8	5.9	(49.0)	
5. Y-axis	64.0	3.1	(74.0)	
6. Occlusal plane	8.4	4.2	(21.0)	
7. Interincisal	131.6	5.6	(166.0)	
8. L-1 to Occlusal	21.3	5.3	(6.0)	
9. L-1 to Mandibular	97.1	4.9	(57.0)	
10. U-1 to A-P plane	7.8	2.5	(-3.0)	
11. FH to SN plane	5.4	2.4	(3.0)	
12. S N A	83.4	2.6	(73.0)	
13. S N B	80.0	2.5	(78.0)	
14. SNA-SNB diff.	3.4	1.7	(-5.0)	
15. U-1 to FH plane	110.8	5.6	(88.0)	
16. L-1 to FH plane	61.6	5.8	(76.0)	
17. Gonial angle	117.5	8.1	(145.0)	
18. Ramus angle	88.5	4.5	(102.0)	

Restriction of maxilla and hyper-divergent pattern of mandible are recognized.

DISCUSSION

Antley-Bixler syndrome is primarily characterized by craniosynostosis, short skull, mid-face hypoplasia, choanal atresia, radiohumeral synostosis, and multiple arthrogyriposis

(Table 2). In addition, hydrocephalus, contracted pelvis, and contracted thorax may also be observed clinically⁴⁾. In girls, external genital anomalies such as absence of the vagina and hypoplasia of the labia majora are commonly noted⁷⁾. Severe secondary

Fig. 6 Cephalometric radiographs
Restriction of the anterior base of the skull, orbits and maxilla are recognized.

Fig. 7 Frontal cephalometric measurements
A narrow upper arch is recognized.

Fig. 8 Lateral cephalometric measurements
Mandibular vertical growth with a short ramus of the mandible and skeletal open bite 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 relatives³⁾ and autosomally recessive inheritance

Fig. 9 Profilogram by roentgen-cephalometry
 Restriction of maxilla and mandibular are recognized.

has been suggested. In other types of cranio-synostosis 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 anti-mycotic 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 syndrome³⁾. None of these factors were identified in our patient.

The severity of the radiohumeral synostosis has been reported to vary widely among patients, and humeral synostosis is not believed to be an important criterion for diagnosing this disease⁵⁾. 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 established⁵⁾.

Koshiba *et al.*⁶⁾ observed missing maxillary second premolar, narrowing of the maxillary arch width, tendency toward Skeletal Class III

Table 2 Clinical synopsis

	Our Case
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	
Radiohumeral synostosis	*
Femoral bowing	+/-
Neonatal femoral fractures	—
Ulnar bowing	+/-
Joint contractures	○
Hands	
Arachnodactyly	—
Camptodactyly	○
Neurologic:	
Central nervous system	
Variable mental retardation	○
Hydrocephalus	○

*: Chondral radioulnar synostosis

○; present

—; absent

+/-; not definite

modified from Escobar *et al.*²⁾

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.*⁹⁾. 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.

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