Orthodontic Treatment for a Child with Melanotic Neuroectodermal Tumor during Infancy: A Case Report

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Abstract: Background: Melanotic neuroectodermal tumor of infancy (MNTI) is a very rare tumor, and case reports of orthodontic treatment for patients with MNTI may be previously unreported. This article describes the orthodontic treatment for a 7-year-old girl with MNTI in the mandible. Case: Her chief complaint was anterior crossbite. Although she had an MNTI diagnosis at the age of 8 months, it remained subclinical. Therefore, she has been regularly followed-up by computed tomography (CT) and magnetic resonance (MR) imaging without aggressive treatment. We had worried about the stimulation of MNTI on the mandible by changing her occlusal position with orthodontic treatment. Therefore, we sufficiently explained to her and her family that orthodontic treatment was at risk for worsening MNTI. However, they desired treatment that consisted of maxillary protraction and slow expansion to correct anterior crossbite and encourage the permanent tooth eruption. After 19 months of active orthodontic treatment, the anterior crossbite was improved, and the eruption of permanent teeth made good progress. No evidence of progression and exacerbation of MNTI has been found by both CT and MR imaging. As the observation period is still short, we need a careful and long-term follow-up of her occlusion and MNTI. Furthermore, when we encounter rare cases without previous experience and reports, informed consent was of particular importance.

Keywords: orthodontic treatment; melanotic neuroectodermal tumor of infancy (MNTI); mandibular protrusion; expansion appliance; maxillary protraction

1. Introduction

Anterior crossbite is defined as a condition characterized by the anterior negative overjet [1]. Some causes of anterior crossbite have been reported, including genetic factors and functional or dental imbalances [2]. This condition could be easily found by patients or their families because it affects their appearance and masticatory function. Therefore, many patients hope to initiate early orthodontic treatment in deciduous or mixed dentition stages. On the plus side of the treatment during growth period, we might be able to prevent them from developing skeletal or dental Class III malocclusion, chronic trauma, and periodontal problems [3,4]. The common treatment options for crossbite therapy in children include expansion appliances, facial masks, and chin cup appliances [5–7].

We can find many articles written about crossbite therapy in children [3,5–11]. However, it is rarely possible for us to find the case reports of orthodontic patients with melanotic neuroectodermal tumor of infancy (MNTI). The reason is that MNTI is a very rare tumor, and only 500 papers have been reported so far [12]. Although MNTI is generally considered a benign tumor in newborns, it has a possibility of becoming malignant and metastatic [13,14]. This tumor often tends to be characterized by rapid growth and aggressive bone destruction without pain and exulceration [12]. Then, facial asymmetry and feeding difficulties occur [12,15]. The sites of predilection were the maxilla (68–80%),
followed by the skull (10.8%), mandible (5.8%), and brain (4.3%) [16–18]. A gender predilection ratio reported that male patients had slightly higher incidence for MNTI compared to female patients at a median age of 4.5 months. On the other hand, female patients showed a higher prevalence at 3 years of age and older [17]. The typical treatment for MNTI is surgical resection, but many studies indicate high recurrence rates [19–22]. Therefore, careful follow-up is very important in cases of MNTI.

No article has previously described the orthodontic treatment of a patient with MNTI. Moreover, this patient has the unprecedented process of MNTI as the reduction in the size of the tumor without aggressive treatment. While the observation period is still short, her occlusion is well maintained and she does not have deteriorating findings of MNTI. Therefore, we present the early treatment of anterior crossbite for the child patient with MNTI in this report.

2. Case Report

A pediatric patient had a chief complaint of anterior crossbite. Her frontal view was almost symmetrical, and the facial profile was straight (Figure 1).

She had no family history of Class III malocclusion. Her overjet was −2.0 mm, and overbite was 3.0 mm. The terminal plane was a mesial step type on both sides. Severe crowding was observed in both arches and the left mandibular anterior teeth showed abnormal morphology.

She had an MNTI diagnosis at the age of 8 months in oral surgery, and surgical resection was considered and proposed. However, her parents did not approve of surgery because it was the most invasive and unimaginable option for them. Therefore, she was followed closely without aggressive treatment for a period of time. A month after the
MNTI diagnosis, the reduction in the size of the tumor could be confirmed by clinical examination, computed tomography (CT) scans, and magnetic resonance imaging (MRI), though she did not receive aggressive treatment. Her MNTI indicated a similar reduction tendency every year, and still no evidence of progression has been found.

Cephalometric analysis, when compared with the Japanese norm, showed a skeletal Class III jaw-base relationship (ANB, \(-2.0^\circ\)) because of mandibular overgrowth (SNB, 82.5\(^\circ\)). Wits value was \(-7.0\) mm. The mandibular plane angle was within the normal range (Mp-SN plane, 34.2\(^\circ\)). The upper and lower incisors were lingually inclined (U1-SN, 97.0\(^\circ\) and L1-Mand. pl., 80.6\(^\circ\)), and it resulted in an increase of the interincisal angle (148.2\(^\circ\)).

A panoramic radiograph showed the lack of eruption spaces for the permanent teeth (Figure 2).

![Figure 2. Pretreatment lateral cephalogram (A) and panoramic radiograph (B).](image)

Though we listed these problems as previously described, we were deeply concerned about orthodontic treatment for the patient with MNTI because of an unprecedented case. Furthermore, we had worried about the stimulation of MNTI on the mandible by changing her occlusal position with orthodontic treatment. Therefore, we sufficiently explained the risk to her and her parents that there was a possibility that her tumor progression would be induced by orthodontic treatment, and we would have to discontinue orthodontic treatment if any such symptoms appeared. However, they strongly preferred orthodontic treatment and expressed that they would be responsible for whatever occurred. Thus, we formed a treatment plan.

Our proposed therapy consisted of maxillary protraction and slow maxillary expansion. The mechanical device was a facemask combined with a removable expansion plate. The maxillary protraction could help to accelerate the maxillary growth. Moreover, activating cells in the maxillary sutures by the combination of maxillary expansion appliance would enhance an effect of maxillary protraction, and its device also helped to create eruption spaces for the permanent teeth. It was somewhat concerning that one of the supporting points of the facemask was her chin. Therefore, we explained sufficiently to her and her family that orthodontic treatment posed a risk of worsening MNTI once again. As it did not make them falter in their determination to the treatment, her orthodontic treatment was initiated. She was evaluated regularly for signs of advancing MNTI deterioration as before in oral surgery.

At the age of 7 years and 9 months, maxillary expansion was performed by a removable expansion plate firstly. The plate included the anterior springs that tipped upper incisors buccally and the hooks that were placed in the canine region for a facemask. She was instructed to use it every night and turn the expansion screws weekly on the specified day of week (1/4 turn: 0.25 mm). After 6 months, we ensured that she became accustomed to use this appliance properly, and maxillary protraction was also started by the facemask. The facemask was adjusted to fit her and used rubber bands attached to hooks for applying
force forward on her maxilla. A pulling force of 300 g by the rubber band was applied on each side.

About 5 months after the start of maxillary protraction, the overjet was corrected (Figure 3). However, we encouraged her to continue use of the facemask for an additional 3 months because the overjet should be overcorrected. A removable expansion plate continued to be used for the duration of checking for the eruption of permanent teeth. After 19 months of active treatment, maxillary expansion was also stopped. We placed a Hawley retainer on the maxillary arch for retention and monitored the eruption of permanent teeth and the maxillomandibular skeletal relationship (Figure 4).

At a 6-month follow-up, the overjet and overbite were well maintained (overjet, 3.5 mm and overbite, 3.0 mm). On the facial photographs, her facial profile remained...
straight and balanced. Moreover, the oral photographs and a panoramic radiograph showed that the eruption of permanent teeth made good progress (Figures 5 and 6).

![Figure 5](image_url1)

**Figure 5.** Six-month retention facial and intraoral photographs. (A) Lateral view; (B) frontal view; (C) right molar relationship; (D) centric occlusion; (E) left molar relationship; (F) overjet; (G) mirror view (maxillary); (H) mirror view (mandibular).

![Figure 6](image_url2)

**Figure 6.** Lateral cephalogram (A) and panoramic radiograph (B) at 6-month retention.

Posttreatment cephalometric evaluation showed a skeletal Class I jaw base relationship (ANB, 0.5°). Concomitantly, Wits value was increased by 2.8 mm. The mandibular plane angle was maintained within the normal range (Mp-SN plane, 34.4°). The upper and lower incisors were labially inclined as compared with pretreatment (U1-SN, 118.0° and L1-Mand. pl., 82.0°) and the interincisal angle was leaded to the normal range (125.6°) (Table 1).
Table 1. Cephalometric summary [23,24].

| Variables                        | Japanese Norm | SD | Pretreatment (7 Years 4 Months) | Posttreatment (9 Years 11 Months) |
|----------------------------------|---------------|----|---------------------------------|-----------------------------------|
| ANB                              | 2.8           | 2.4| −2.0                            | 0.5                               |
| SNA                              | 80.8          | 3.6| 80.5                            | 80.5                              |
| SNB                              | 77.9          | 4.5| 82.5                            | 80.0                              |
| Mand.pl.–SN pl.                  | 37.1          | 4.6| 34.2                            | 34.4                              |
| U1-SN pl.                        | 105.9         | 8.8| 97.0                            | 118.0                             |
| L1-Mand. pl.                     | 93.4          | 6.8| 80.6                            | 82.0                              |
| Interincisal angle               | 123.6         | 10.6| 148.2                           | 125.6                             |
| Linear (mm)                      |               |    |                                 |                                   |
| Wits value                       | −1.92         | 2.52| −7.0                            | −4.2                              |
| Overjet                          | 3.1           | 1.1| −2.0                            | 3.5                               |
| Overbite                         | 3.3           | 1.9| 3.0                             | 3.0                               |

As for MNTI, both CT and MRI found no evidence of progression and exacerbation until now.

3. Discussion

This article described the successful outcome of the early treatment for the child patient with anterior crossbite and MNTI, which is a very rare tumor.

We had another plan initially. The alternative was chin-cup therapy to improve anterior crossbite. The chin-cup was considered an effective treatment appliance to control mandibular growth in patients with anteriorly positioned mandibles [25–27]. However, this approach would have a significant influence over the tumor compared to other appliances because the chin-cup directly applied a force to her mandible with MNTI. Therefore, we did not recommend it and offered this alternative to the patient and her family.

Presently, her occlusion was corrected and stabilized by our selected therapy. We think that we should keep the skeletal relationship and the eruption of permanent teeth under observation until completion of growth. Several reports indicated the difficulties of the early treatment and retention of skeletal Class III malocclusion in children due to their inability to predict mandible growth [10,28,29]. Therefore, follow-up is necessary and important to retain her collected skeletal or dental occlusion. Furthermore, this girl and her parents hope the additional orthodontic treatment of the mandibular anterior crowding in permanent dentition stage. For the treatment of this malocclusion, we will have to extract the malposed tooth or another one in MNTI. Therefore, she is cautiously determined whether it can be treated at the timing of second phase treatment using the multiple bracket method.

In the present case, symptoms produced by MNTI were almost same ones as previous reports, but the subsequent process was not completely different. Triggers that her parents noticed were swelling of her alveolar ridge in the left mandibular region and following facial asymmetry. At the age of 8 months, she was referred to our department. On clinical examination, smooth swelling extended from the right mandibular primary canine to the left mandibular molar region (Figure 7). It was firm without ulceration or discharge and caused her neither pain nor feeding difficulties.

MRI and CT showed a large and widespread mass on the mandible (Figure 8). The definitive diagnosis of MNTI was made by biopsy. After we were provided it, we consulted with pediatricians, cranio-facial surgeons, and radiologists about treatment options, complications, and prognosis. Though surgical resection had high possibilities of serious complications and interfering with her life, we recommended surgery because we expected a complete cure. However, her parents did not approve of surgery, as was discussed above. In addition, the result of CT and MRI indicating the reduction in the size of the tumor after biopsy encouraged their decision (Figure 8).
Figure 7. Pictures of the patient at the age of 8 months. (A) Facial frontal view; (B) intraoral view showing swelling in the left mandibular region.

Figure 8. Sequential radiographic changes for MNTI by CT on the left side and MRI on the right side. (A) Before the start of treatment at the age of 8 months; (B) 1 month later, after the biopsy; (C) 4 years and 6 months later, after the biopsy; (D) 6 years and 6 months later, after the biopsy.
She has been regularly monitored MNTI over the past 9 years by clinical and radiograph examination, and their findings have not demonstrated a growth or progression of the tumor but a reduction (Figure 8). As far as we know, this is the first case report in which the patient of MNTI had a good outcome without aggressive treatment. The reasons for this satisfactory course remains unknown. We will continue to follow her until she is grown up because Lurie [30] and Jain et al. [31] reported the fairly rare cases of MNTI in older children and adults.

4. Conclusions

Our article about orthodontic treatment for the patient with MNTI may be the first case report. In addition, MNTI that we encountered in this report was a rare tumor, and it had an unusual site of onset and a marvelous course of treatment. It was very difficult for us to decide on the right treatment. Therefore, the importance of informed consent was reawakened. That we had enough time to explain and discuss treatment with the patient, her family, and several doctors was a major contributing factor to our success. We would also like to achieve future treatment by close liaison with them and report further studies with long-term follow-up in this patient.

Author Contributions: Conceptualization, N.H. and Y.M.; methodology, N.H. and T.N.; software, N.H.; validation, N.H., T.N., K.S. and Y.M.; formal analysis, N.H. and T.N.; investigation, N.H., T.N., K.S. and Y.M.; data curation, N.H.; writing—original draft preparation, N.H.; writing—review and editing, T.N., K.S. and Y.M.; visualization, N.H.; supervision, T.N. and K.S.; project administration, Y.M. All authors have read and agreed to the published version of the manuscript.

Funding: This research received no external funding.

Institutional Review Board Statement: Not applicable.

Informed Consent Statement: Informed consent was obtained from all subjects involved in the study.

Data Availability Statement: Data are contained within the article.

Conflicts of Interest: The authors declare no conflict of interest.

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