

Unicystic Ameloblastoma in a Child Treated with a Combination of Conservative Surgery and Orthodontic Treatment: A Case Report

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Unicystic ameloblastoma (UAM) is a variant of intraosseous ameloblastoma that occurs as a single cystic cavity. This report describes a case of UAM of the mandible in a seven-year-old girl. The lesion radiographically mimicked a dentigerous cyst. Under the primary diagnosis of a dentigerous cyst, marsupialization was performed to erupt the first molar involved in the cystic lesion and to obtain a definitive diagnosis. The biopsy specimen revealed ameloblastoma. During careful observation, orthodontic treatment, which was performed to upright and promote the eruption of the first molar involved in the tumor, maintained the space needed for enucleation of the tumor. Finally, the second primary molar was extracted, and the lesion was enucleated at 3 years and 4 months after marsupialization. The results of the histological examination revealed UAM. Conclusively, the treatment course not only avoids a resection of the mandible but also induces eruption of the teeth involved in the tumor. Thus, the combination of conservative surgery and orthodontic treatment was effective in the management of UAM that mimics a dentigerous cyst.

Keywords: Unicystic ameloblastoma, Orthodontic treatment, Marsupialization, Enucleation

INTRODUCTION

Four types of ameloblastoma can presently be distinguished: ameloblastoma (formerly solid/multicystic ameloblastoma¹), unicystic ameloblastoma (UAM), extraosseous/peripheral ameloblastoma, and metastasizing ameloblastoma.² UAM is defined as a variant of intraosseous ameloblastoma that occurs as a single cystic cavity² and is considered to be a less aggressive form compared with the solid or multicystic type.³ Currently, two main histological variants of UAM are considered to exist: luminal and mural.¹ The luminal variant displays a cystic pattern lined by ameloblastomatous epithelium that protrudes into the lumen. The mural variant displays tumor cells within the cystic wall. Given that the mural variant frequently results in recurrence compared with

the luminal variant, tumor invasion into the underlying connective tissue should be considered when planning the treatment for UAM.^{4,5} It has been suggested that mural variants should be treated radically.^{6,7} However, the majority of UAMs occur in children,⁸ and the resection of the mandible in pediatric patients leads to complications, such as dysfunction and deformity. Treatment with simple enucleation is considered adequate for the majority of UAM cases.^{9,10} Even in adult cases, enucleation and curettage were the most common techniques for the management of UAM with and without mural invasion of the tumor.^{4,11,12} According to some authors, recurrence is likely not an important consideration and should not be considered as equivalent to failure because a second surgery can be successful.^{12,13}

This report describes a case of UAM of the mandible in a 7-year-old girl that mimics a dentigerous cyst given its association with an unerupted permanent mandibular first molar. Orthodontic treatment and marsupialization were useful given that it enabled the successful enucleation of the tumor without the need for tooth removal.

Case report

A 7 years and 9 months year old girl who was referred to the Department of Pediatric Dentistry at Tsurumi University Dental Hospital with a chief complaint of an unerupted left mandibular first molar. A panoramic radiograph revealed a unilocular, well-defined, radiolucent lesion surrounding the crown of the left mandibular first molar with delayed eruption (Figure 1). The radiograph demonstrated that the first molar was located near the mandibular border. A normal trabecular bone structure was not evident in the area above the cystic lesion of the first molar. Intra- and extra-oral

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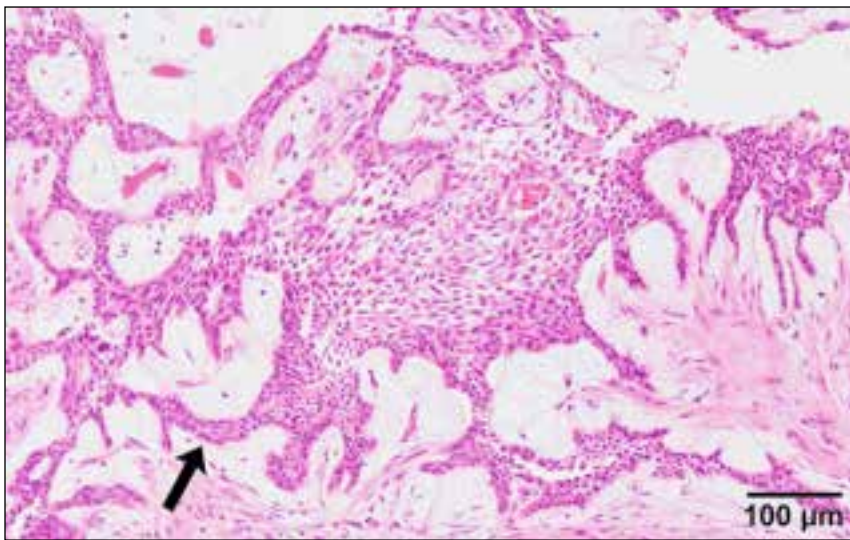
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Figure 1. A panoramic radiograph obtained at the first visit revealing a unilocular, well-defined, radiolucent lesion surrounding the crown of the unerupted left mandibular first molar. Image obtained at 7 years and 9 months of age.



Figure 2. The histological findings of the specimen obtained at the marsupialization of the unerupted left mandibular first molar revealing a plexiform ameloblastoma. Arrow indicates ameloblastomatous tumor cells. Specimen obtained at 7 years and 10 months of age.



examinations revealed no abnormalities other than the delayed eruption of the first molar. Cone beam computed tomography (CBCT) was not performed because the extent of the lesion was apparent on the panoramic radiograph. Based on clinical and radiographic findings, a dentigerous cyst was considered as the primary diagnosis. Differential diagnoses included a cystic ameloblastoma and an adenomatoid odontogenic tumor. An excisional biopsy was planned to obtain a definitive diagnosis.

The cystic lesion of the crown of the molar was treated by marsupialization (age: 7 years and 10 months), which was performed in an attempt to allow the tooth to erupt. The soft tissue surrounding the first molar was removed and served as a histological specimen for definitive diagnosis. A histological examination revealed a plexiform ameloblastoma with anastomosing strands and cords of tumor cells (Figure 2). The stroma consisted of myxomatous connective tissue and fibrous tissue with dense collagen fibers.

CBCT was used to follow-up the lesion given that information about the location of the lesion and the relationship between the lesions and their adjacent anatomical structures is useful for minor oral surgery.^{14, 15} The radiograph demonstrated that the left first molar had begun to erupt at two months postoperatively (age: 8 years; Figure 3). However, the direction of the eruption was mesioangular, and only a small space was present between the left second primary molar and the first molar. Thus, it was difficult to enucleate the remaining tumor at the mesial area of the crown of the first molar. Four months after marsupialization (age: 8 years and 2 months), a Halterman appliance was placed on the mandibular dentition by cementing metal bands on the left second primary molar and the right first molar to upright the left first molar (Figure 4A). An orthodontic button was bonded to the first molar, and distal traction was applied to the tooth using an elastic power chain (Figure 4B).

Three months after the placement of the orthodontic appliance (7 months after marsupialization; age: 8 years and 5 months), surgical curettage was performed around the first molar crown. The tissue specimen exhibited evidence of remaining tumor tissue. Radiographic observation was continued. Given that the radiolucent lesion was evident at 3 years and 3 months after the marsupialization (age: 11 years and 1 month; Figure 5), enucleation was performed. The second primary molar was extracted, and

Figure 3. The radiographic findings during follow-up. Image obtained at 8 years of age (2 months after marsupialization).

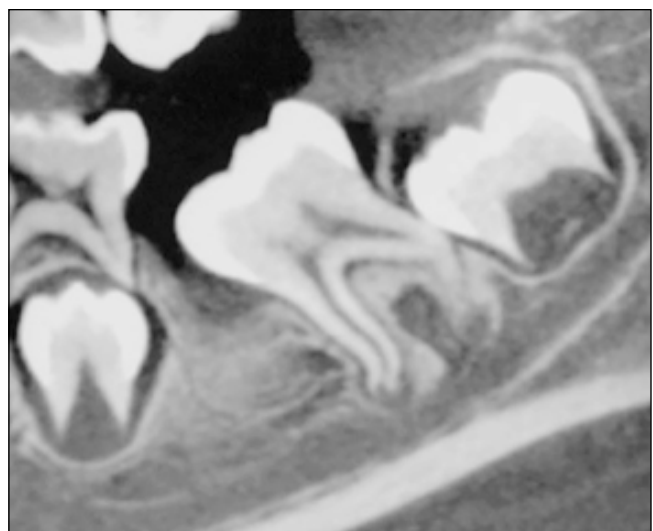


Figure 4. Orthodontic treatment at 8 years and 2 months of age (4 months after marsupialization). A Halterman appliance was placed on the mandibular dentition by cementing metal bands on the left second primary molar (A), and an orthodontic button was bonded to the left first molar to upright the tooth using an elastic power chain (B).

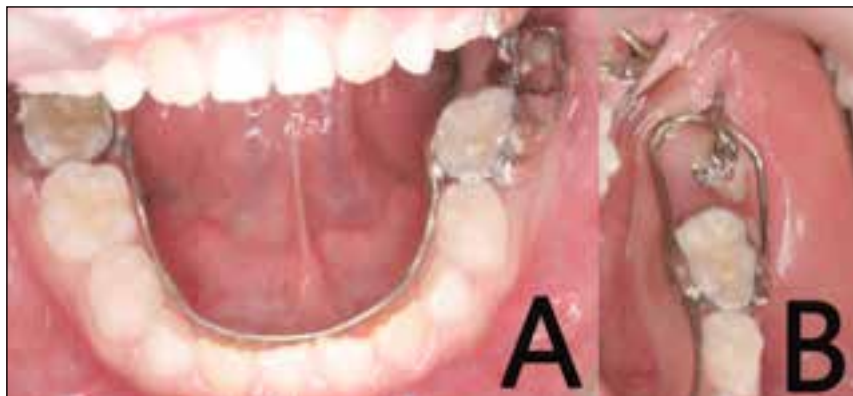


Figure 5. The radiographic findings at 11 years and 1 month of age (3 years and 3 months after marsupialization).

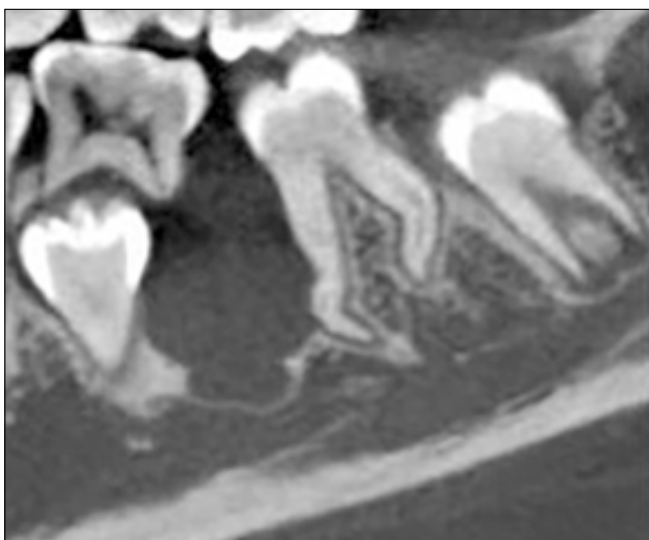


Figure 6. Enucleation of tumor. The second primary molar was extracted, and the lesion was enucleated at 11 years and 2 months of age (3 years and 4 months after marsupialization).



the lesion was enucleated at 3 years and 4 months after marsupialization (age: 11 years and 2 months; Figure 6). Histological examination revealed a UAM (Figure 7A) lined by ameloblastomatous epithelial cells with both intraluminal plexiform proliferation of tumor cells (Figure 7A arrow) and mural invasive tumor islands in the fibrous connective tissue (Figure 7B, arrow head).

The radiolucent area was significantly reduced after the enucleation. One year after the enucleation, the first molar erupted, and the alveolar bone formation was noted around the root although a panoramic radiograph revealed the small bony defect on the mesial aspect of the first permanent molar. Careful clinical and radiographic follow-up was performed. To date, after 5 years of follow-up, no evidence of tumor recurrence was noted (Figure 8; 7 years and 7 months after the marsupialization; age: 15 years and 5 months). The shape and size of the defect on the mesial of the first molar remain unchanged.

Figure 7. The histological findings of the specimen obtained at the enucleation of the lesion. **A,** Image of entire unicyclic ameloblastoma with intraluminal plexiform proliferation of tumor (arrow); **B,** Mural invasive islands of the tumor (arrowhead).

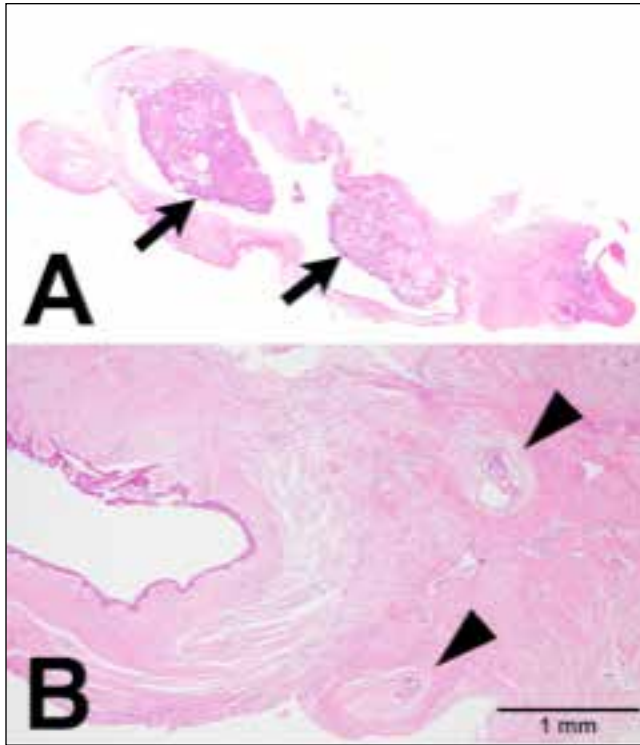
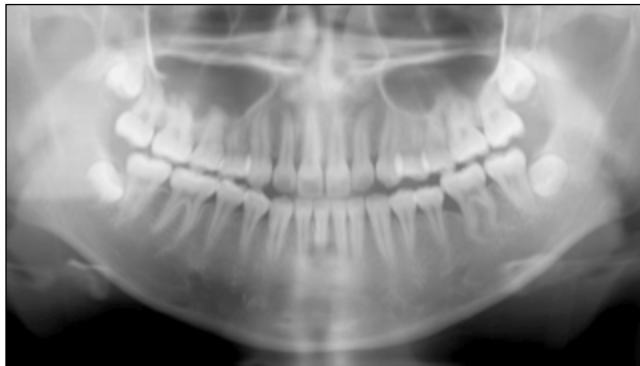


Figure 8. A panoramic radiograph obtained at 15 years and 5 months of age (7 years and 7 months after marsupialization; 4 years and 3 months after enucleation of the lesion).



DISCUSSION

The treatments for UAM include resection, enucleation, and marsupialization with or without other second-phase treatments.¹¹ In the present case, marsupialization was performed because it was assumed that the lesion was a dentigerous cyst. Biopsy was performed to establish a definitive diagnosis. Although UAM and adenomatoid odontogenic tumor were suspected based on the radiographic findings, it was not possible to distinguish between cyst and tumor. UAM is frequently associated with unerupted teeth and is often misdiagnosed as a dentigerous cyst.^{6, 8, 16} This misdiagnosis can lead to initial treatment with marsupialization.⁸ A microscopic examination of the excised specimen obtained at the time of

marsupialization in our case revealed plexiform ameloblastoma. It was not possible to distinguish between solid/multicyclic ameloblastoma and UAM with the specimen obtained during marsupialization; however, UAM was strongly suspected. Given that the resection of the mandible in pediatric patients may lead to complications, such as dysfunction and deformity, we decided to choose a conservative surgical approach for future treatment plan.

Marsupialization is considered for the exteriorization or decompression of the cyst.¹¹ Marsupialization followed by enucleation is one therapeutic approach for ameloblastoma.^{11, 12, 17} There are several reports of cystic ameloblastoma cases in which the tumor completely disappeared after marsupialization alone, which suggests that marsupialization is useful for avoiding wide resection of the mandible in patients with UAM.¹⁷ The reason for this phenomenon remains unknown because exteriorization should not cause a tumor to regress.¹¹ Marsupialization and enucleation were applied not only to the unilocular UAM but also the UAM that mimicked a dentigerous cyst; thus, the impacted teeth within ameloblastomas can be preserved and maintain function.^{16, 18} A similar effect has been reported in adenomatoid odontogenic tumors.^{19, 20} In the present case, the lesion was histologically diagnosed as ameloblastoma at the first biopsy, and we assumed that the tumor was not completely enucleated by marsupialization and remained around the crown of the first molar. Accordingly, we performed careful follow-up as the next step and planned to perform enucleation when the residual tumor increased in size. To enable the future performance of enucleation and to allow the teeth to erupt, orthodontic treatment was applied in addition to marsupialization. Similar to our case, orthodontic treatment was successfully applied in some reported cases of tumors involving the crown of the unerupted teeth.^{16, 19, 20}

In summary, we performed marsupialization first to enable eruption of the first molar and obtain a definitive diagnosis of the lesion. Secondly, orthodontic treatment, which was performed to upright and promote eruption of the first molar, maintained space for the enucleation of the tumor. Finally, enucleation of the lesion was accomplished. The enucleated specimen determined the final diagnosis as UAM. Conclusively, the treatment course not only avoids a resection of the mandible but also induces eruption of the teeth involved in the tumor. Thus, the combination of conservative surgery and orthodontic treatment was effective in the management of a UAM that mimics a dentigerous cyst.

CONCLUSION

The present case demonstrates that the combination of conservative surgery and orthodontic treatment is an effective management options for UAM that mimic a dentigerous cyst.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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