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Case Report

Conservative Management of Mural Subtype Unicystic Ameloblastoma in Young Patient: A Case Report with One Year of Follow Up

Genç Hastada Mural Alt Tip Unikistik Ameloblastomun Konservatif Tedavisi: Bir Yıllık Takipli Bir Olgu Sunumu

Aydın Onur Gerçek[1], Selen Adiloğlu[1], Alper Aktaş[1]

ABSTRACT

Introduction: Ameloblastoma is a benign tumor primarily found in the jawbone. Unicystic ameloblastoma, first categorized in 1977, comprises four distinct types and is considered a less aggressive variant of ameloblastoma. Marsupialization, a conservative surgical approach, involves creating an open pocket by suturing the edges of a lesion to facilitate fluid drainage and promote healing.

Case Report: This case report details the surgical management of a 13-year-old boy diagnosed with unicystic ameloblastoma and an impacted left mandibular third molar, who was referred to the Oral and Maxillofacial Surgery Department at Hacettepe University during the COVID-19 pandemic due to a radiolucent lesion surrounding the impacted tooth, which caused facial asymmetry. The treatment plan included the extraction of the second molar, a biopsy, and marsupialization. During the one-year follow-up, no recurrence was noted.

Conclusion: Marsupialization, which involves creating an open pocket by suturing the edges of a lesion to the oral cavity or sinuses for fluid drainage, is a conservative treatment approach particularly suitable for young patients. This method preserves the growth potential of the bone, improves aesthetic outcomes, and protects vital structures. In this case, due to the COVID-19 pandemic, conservative treatment under local anesthesia was preferred, with enucleation performed after marsupialization. Such approaches address clinical needs effectively and support patients' psychological well-being during challenging periods like pandemics.

Keywords: Ameloblastoma; COVID-19; Jaw neoplasms

ÖZET

Giriş: Ameloblastoma, çene kemiğinde sıklıkla görülen benign bir tümördür. İlk olarak 1977 yılında tanımlanan unikistik ameloblastoma, dört farklı alt tipe sahip olup ameloblastomanın daha az agresif bir varyantı olarak kabul edilmektedir. Marsupyalizasyon, kist epitelinin bir bölümünün cerrahi olarak çıkarılması ve kistin ağız mukozasına açılması yoluyla drenajı kolaylaştırarak iyileşmeyi teşvik eden konservatif bir cerrahi yaklaşımdır.

Vaka raporu: 13 yaşında bir erkek çocuk, yüz asimetrisine neden olan gömülü alt çene üçüncü molarını çevreleyen radyolusent bir lezyonla Hacettepe Üniversitesi Ağız, Diş ve Çene Cerrahisi Anabilim Dalı'na başvurdu. Uygulanan tedavi protokolü, ikinci molar dişin çekilmesi, biyopsi yapılması ve marsupyalizasyon prosedürünü içermiştir. Bir yıllık takip sürecinde herhangi bir rekürrens tespit edilmemiştir.

Sonuç: Marsupyalizasyon, kistik yapıların drenajı için lezyonun kenarlarının ağız boşluğuna veya sinüslere dikilmesiyle sıvı drenajı için açık bir cep oluşturulmasını içeren konservatif bir tedavi yaklaşımıdır ve özellikle genç hastalar için uygundur. Bu yöntem, kemiğin büyüme potansiyelini korurken estetik sonuçları iyileştirir ve hayati yapıların korunmasını sağlar. Bu vakada, COVID-19 pandemisi nedeniyle lokal anestezi altında konservatif tedavi tercih edilmiş ve marsupializasyonun ardından enükleasyon uygulanmıştır. Bu tür yaklaşımlar, klinik gereksinimleri etkin bir şekilde karşılarken, pandemiler gibi zorlu dönemlerde hastaların psikolojik olarak kendilerini idame etmeleri daha kolay olacaktır.

Anahtar Kelimeler: Ameloblastom; COVID-19; Çene neoplazileri

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İletişim: Dr. Aydın Onur Gerçek
Hacettepe University, Faculty of Dentistry, Department of Oral and Maxillofacial Surgery, Ankara, Turkey
E-mail: aogercek@gmail.com
[1] Hacettepe University, Faculty of Dentistry, Department of Oral and Maxillofacial Surgery, Ankara, Turkey,

INTRODUCTION

Unicystic ameloblastoma (UA), first described in 1977, is characterized by a single well-defined cavity and is classified into subtypes based on histopathological characteristics. The 5th edition of the World Health Organization (WHO) classification of ameloblastoma, published in 2022, delineates five distinct categories of this odontogenic tumor: conventional ameloblastoma, unicystic ameloblastoma, extraosseous/peripheral ameloblastoma, adenoid ameloblastoma, and metastasizing ameloblastoma.[1] It is generally less aggressive compared to conventional ameloblastoma.[2] UA constitutes approximately 5-15% of all ameloblastoma cases and predominantly occurs in younger individuals, typically during the second to third decades of life, in contrast to the solid form, which is more commonly observed in the fourth decade. Clinically, UA are often characterized by well-defined, unilocular or multilocular radiolucent lesions, frequently associated with impacted teeth, particularly in the posterior mandible.[3] The mandibular to maxillary UA ratio is reported to be 13:1.[4] The classification of unicystic ameloblastomas includes three histological types: luminal, intraluminal, and mural, which have implications for their biological behavior and treatment outcomes.[5]

Many experts consider radical surgery, which involves the removal of solid or multicystic ameloblastoma with 1 cm safe margins and resection of adjacent soft tissue, to be the most effective treatment option, followed by subsequent reconstruction.[6] Some authors propose an alternative approach that combines conservative surgery with long-term follow-up, particularly in cases where patient compliance is favorable and there is a low risk of adjacent structure involvement.[7] Conservative surgery encompasses techniques such as marsupialization and enucleation or curettage, with or without additional methods like cryotherapy, thermal or chemical cauterization, radiotherapy, or chemotherapy.[8,9] Historically, recurrence rates after enucleation and curettage of UA have been reported to be between 10% and 20%.[2] However, there remains insufficient evidence and a lack of consensus regarding the most effective treatment strategies.

Marsupialization can mitigate the adverse effects associated with definitive treatment. It aids in preserving the inferior alveolar nerve, lowers the risk of pathological fractures, and eliminates the necessity for a bone graft. Nevertheless, its effect on mandibular unerupted anomalies remains uncertain, with recurrence rates varying from 4.5% to over 50%.[10]

During the COVID-19 pandemic, the management of benign lesions in the maxillofacial region underwent significant changes due to the need for heightened infection control measures and the prioritization of urgent cases. The pandemic led to a re-evaluation of surgical practices, particularly for benign lesions, which are often elective procedures.[11] Despite healthcare restrictions during the COVID-19 pandemic, head and neck tumor surgery has remained one of the most actively pursued surgical treatment areas.[12] In this context, marsupialization performed under local anesthesia appeared to be a practical option, reducing exposure risks while effectively managing the lesion. This case report discusses the surgical management of ameloblastoma during the COVID-19 pandemic. A 13-year-old boy underwent marsupialization, a conservative surgery for an unerupted left mandibular third molar.

CASE REPORT

A 13-year-old male patient with facial asymmetry in the left mandibular angular region, attributed to an impacted left mandibular third molar, was referred to the Oral and Maxillofacial Surgery Department at Hacettepe University. Written informed consent for the surgical procedures and the publication of this case report was obtained from the patient and their legal guardians. The patient reported no systemic diseases. Upon examination, expansion was observed in the left mandibular angle area, extending into the retromolar region. An unerupted left mandibular third molar was identified, with no accompanying pain or ipsilateral paresthesia.

A panoramic radiograph revealed a well-defined, solitary radiolucent lesion in the region of the left lower third molar. This lesion extends from the distal root of the second molar to the left ascending ramus and is associated with an unerupted third molar. Cone beam computed tomography (CBCT) scans indicated a cystic lesion encircling the impacted lower left wisdom tooth, displacing the inferior alveolar nerve (Figure 1).

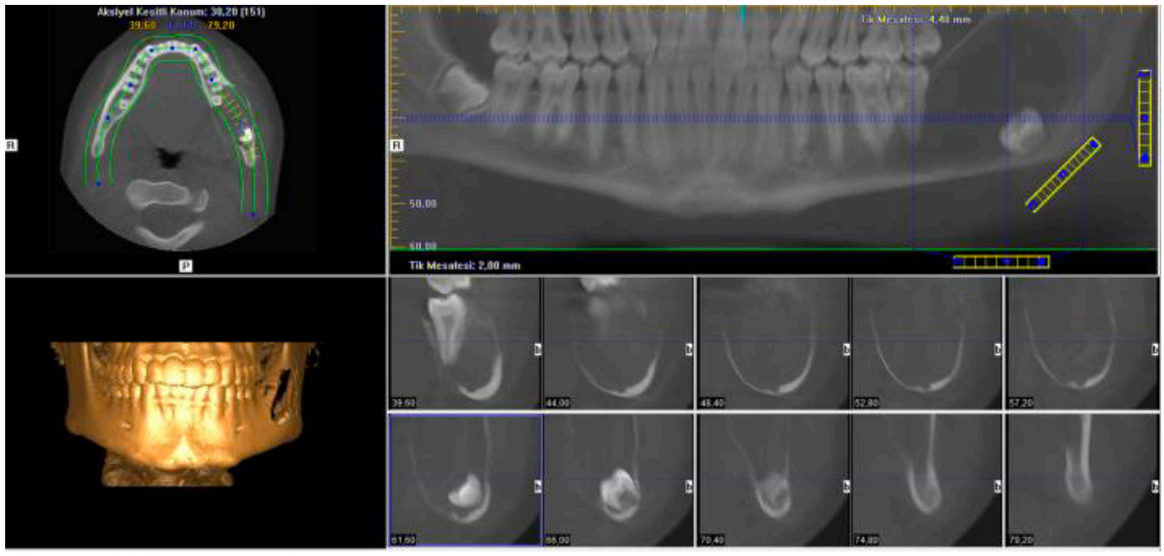

Figure 1. CBCT radiograph of the patient with thin buccal and lingual bone wall.

The lower left second molar exhibited incomplete root formation and lacked sufficient bone support. The lower left third molar was positioned at the jaw angle. An incisional biopsy was conducted, leading to a diagnosis of mural type UA following histopathological examination. The initial step of marsupialization involved surgically removing a portion of the cystic wall to alleviate the intracystic pressure on the overlying bone and mucoperiosteum, thereby creating a clear bone outline of the lesion (Figure 2). The second molar and the impacted third molar teeth were extracted at this stage to minimize postoperative complications and reduce the risk of fracture formation. A medicated gauze dressing, infused with petrolatum and 3% Bismuth Tribromophenate, was used to pack the large lesional cavity, ensuring continuity between the marsupialized lesion and the oral environment (Figure 3). After confirming the epithelialization of the cyst wall, the drain was removed.

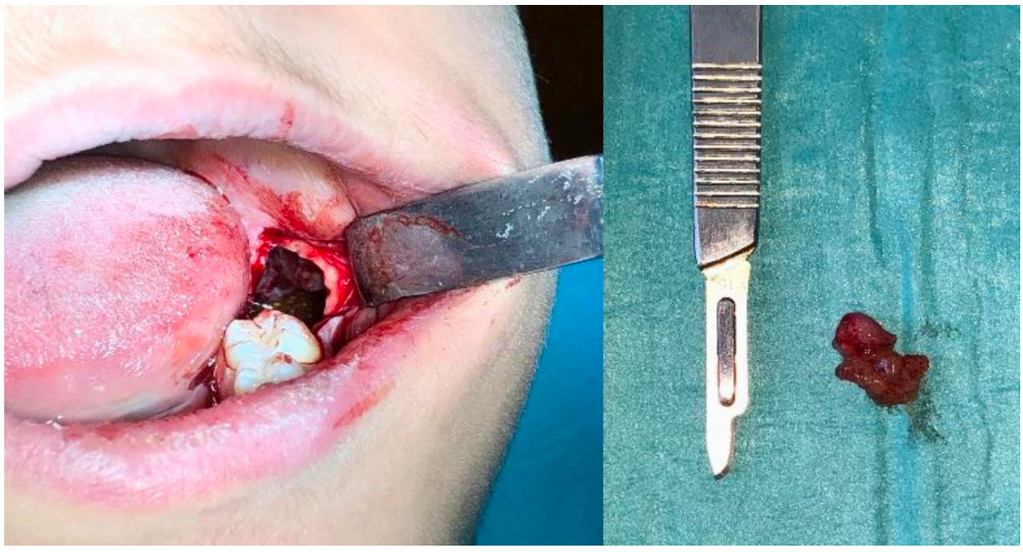

Figure 2. Removal of part of the cystic wall.

Figure 3. Dressing with fine-mesh gauze containing petrolatum and 3% Bismuth Tribromophenate.

Figure 4. The lesion diminished in size and new bone formation was seen.

Post-operatively, the patient was prescribed a course of antibiotics (amoxicillin-clavulanate, 1000 mg twice daily) for one week to prevent infection and nonsteroidal anti-inflammatory drugs (deksketoprofen, 25 mg two times daily) for five days to manage pain and inflammation. Antiseptic mouth rinses (chlorhexidine gluconate 0.12%) were recommended twice daily to maintain oral hygiene. The patient was scheduled for a month of follow-up appointments and daily management following the surgery, after which the frequency would shift to weekly check-ups. The parents were advised to ensure proper hygiene of the oral cavity by regularly irrigating the lesion until the definitive operation could be done.

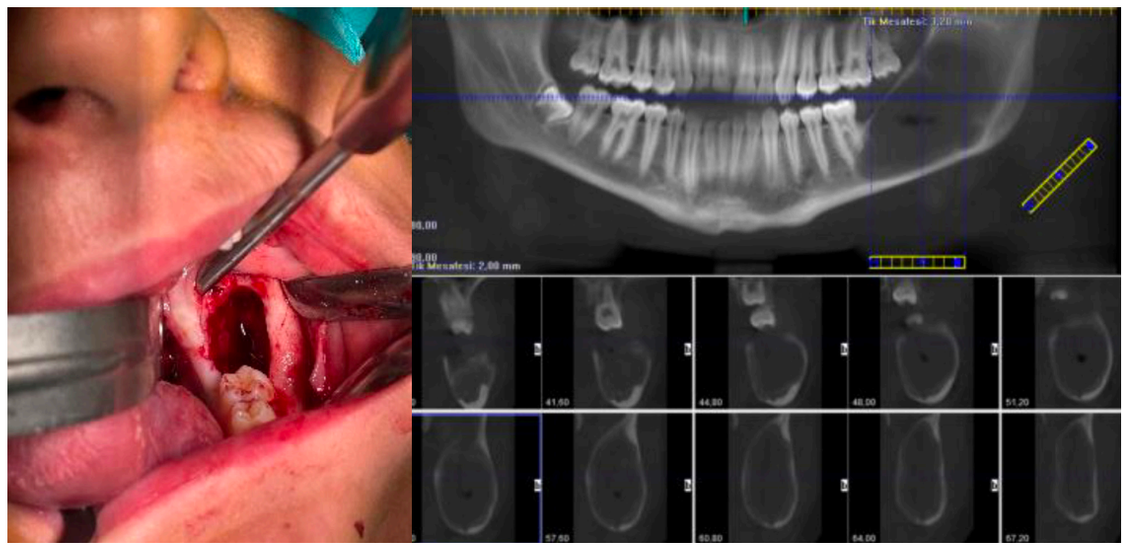

Figure 5. Six months of follow-up with a clear bony outline.

Cone-beam computed tomography was performed six months after marsupialization. The nerve was located within the lesion cavity, surrounded by a radiopaque border, with no paresthesia or complaints reported. The lesion showed a reduction in size on radiographs, and new bone formation was observed (Figure 4). The patient's enucleation was performed after six months of follow-up, achieving a clear bony outline. The final biopsy supported the first biopsy results (Figure 5). During the assessment, we evaluated wound healing, ipsilateral paresthesia of the lower lip, bone formation, and restoration of normal bone contour, along with monitoring for any potential lesion recurrence. Enucleation was performed for six months postoperatively, and no recurrence was observed during the one-year follow-up. By the final follow-up stage, a normal mandibular bony contour was achieved, accompanied by complete remodeling (Figure 6).

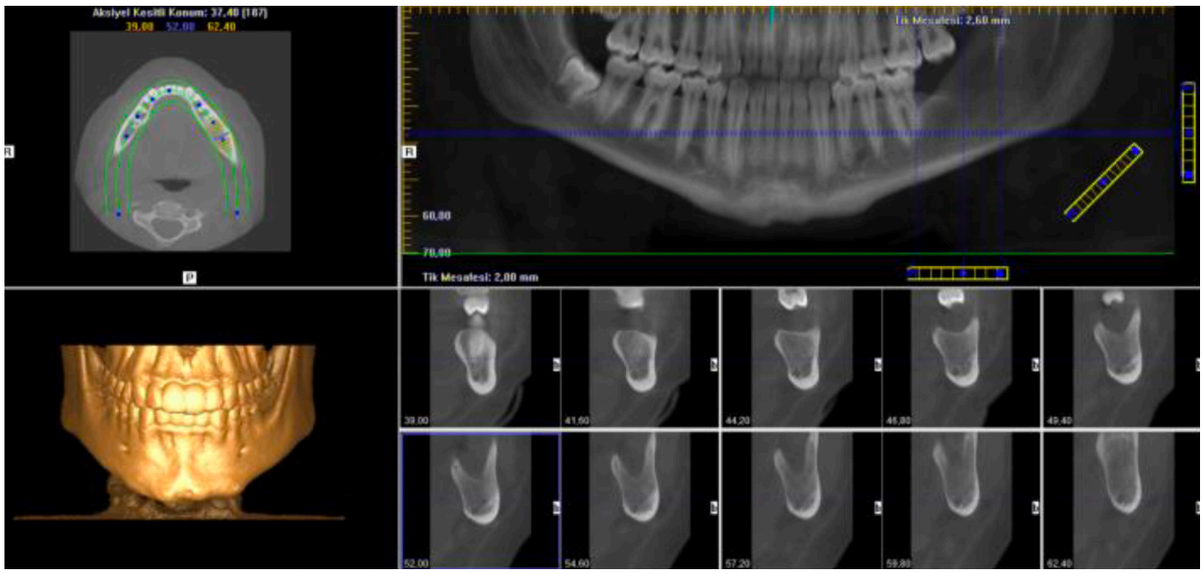

Figure 6. Improvement was achieved after one year of follow-up.

DISCUSSION

Unicystic ameloblastomas are benign but potentially aggressive tumors that usually occur in the mandibular region and expand within the bone. Marsupialization is a widely accepted procedure for the treatment of large mandibular cystic lesions.[13] It is a good alternative treatment for cases that cannot be treated under general anesthesia.

The treatment of UA by marsupialization is a subject of controversy; some consider it to be too radical, while others feel it is too conservative. Various treatment options have been employed for UA, including segmental or marginal resection, though more conservative treatments have also been commonly reported.[8] While enucleation and curettage are the most common procedures, some clinicians prefer marsupialization performed in a two-stage surgery to preserve vital structures.[13,14] Studies indicate that marsupialization can be an effective initial treatment, especially in cases where complete excision may pose a higher risk of complications or where the cyst is large enough to warrant a more conservative approach.[15,16] In the present case, our treatment of choice was marsupialization due to the patient's young age, aesthetic expectations, and desire to protect vital structures. One of the issues that should be considered in such patients is the possibility of recurrence. Marsupialization was preferred because it provided advantages regarding lesion control during the period when the operating room was closed due to COVID-19.

Many studies have demonstrated that the possibility of recurrence after resection is minimal.[8,17] A study comparing the recurrence rates of four different treatment options for unicystic ameloblastoma yielded the following results: 3.6% for jaw resection, 30.5%

for enucleation followed by the application of Carnoy's solution, and 16% for marsupialization, with or without additional second-line treatments. The findings indicated that jaw resection had the lowest recurrence rate, followed by enucleation with Carnoy's solution. In contrast, enucleation alone demonstrated the highest recurrence rate. Additionally, the effectiveness of marsupialization could not be fully evaluated since most cases necessitated a second stage of surgery.[6,8] Luminal UA typically indicates a less aggressive tumor type and has a better prognosis. In contrast, mural types often result in higher recurrence rates in children.[18] It is important to remember that significant complications can arise after resection, leading to disfigurement, dysfunction, and psychological distress, particularly in young patients.[17] When aggressive surgery is considered, especially in young patients, the possible psychological effects should be taken into consideration. Our treatment decision was made based off the patient's age, the specific type of ameloblastoma (mural), the high risk of complications associated with the size of anatomical formations, and the restricted access to general operating rooms during the pandemic.

Limited by deficiencies and inaccuracies, previous case reports of young patients with UA treated with marsupialization call for an updated and more comprehensive approach.[19] It is advisable to consider the following points when making decisions for young patients: 1) ongoing facial growth, variations in bone physiology (such as a higher percentage of cancellous bone, increased bone turnover, and reactive periosteum); 2) challenges in initial diagnosis; and 3) the dominance of pre-erupted teeth in unicystic ameloblastoma cases. Given these factors, we opted for marsupialization combined with close monitoring, with the objective of initiating early intervention in the event of recurrence.

In younger patients, UA can be effectively treated through enucleation with bone curettage. However, some surgeons have used alternative adjuvants to reduce the recurrence rate in this treatment. Recurrence rates have been reported, especially in cysts with osseous perforation.[20] While Lau and Samman recommended[8] Carnoy's solution after enucleation, Khare *et al*.[21] tried an alternative variation that would allow faster ossification of the gap with an osteoperiosteal flap. Re-entry surgery with cryotherapy using liquid nitrogen following enucleation is a promising treatment option for large UA lesions. This method may aid in preventing tumor recurrence, as discussed by Ogunsalu *et al*.[22] In the case report by Paulo *et al*.[23] enucleation following marsupialization is considered an effective option for ameloblastomas located in the ramus, as this condition is a benign, slow-growing jaw neoplasm that originates from odontogenic epithelium. A conservative approach in children not only yields favorable aesthetic results but also helps preserve their craniofacial development. This treatment preference is consistent with the evidence found in the existing literature and takes the patient's age into account.

In some instances, adjunctive therapies may be considered post-surgery to minimize the risk of recurrence. These can include cryotherapy, thermal or chemical cauterization, and, in rare cases, radiotherapy or chemotherapy.[24] The identification of the BRAF-V600E driver mutation in mandibular ameloblastomas and other mutations has prompted the search for targeted therapies for jaw ameloblastomas.[25] Attempts to treat ameloblastomas with BRAF inhibitors (BRAFi), alone or in combination with mitogen-activated protein kinase (MEK) inhibitors (MEKi), have been reported as a last-line treatment.[26] BRAF-targeted therapy was effectively used for the first time in a neoadjuvant protocol for a small group of young patients with unicystic, mural-type ameloblastoma.[27] In the study conducted by Hirschhorn *et al*.[28] the first comprehensive histomorphologic findings on BRAF-targeted ameloblastomas were presented, highlighting the effect of BRAFi on the inflammatory response by activating tumor-infiltrating lymphocytes and cytotoxic lymphocytes while downregulating immunosuppressive factors to create an anti-tumor environment.

UA is seen in young patients with multiple impacted teeth. In these cases, these impacted teeth are usually removed during surgery along with the tumors to prevent their recurrence.[29] In order to reduce the number of operations during the COVID-19 pandemic, we chose to extract the second molar and wisdom tooth by performing an incisional biopsy during marsupialization.

During the challenging work conditions of the pandemic era, the conservative management of large cys-

tic jaw lesions through decompression followed by surgery has led to high levels of patient satisfaction and decreased the necessity for extensive surgical procedures. The average reduction rate observed was 80.8%, with no recorded instances of recurrence.[30] In consideration of the young patient's well-being and psychological welfare, we chose to perform enucleation followed by marsupialization to mitigate long-term effects.

CONCLUSION

Marsupialization is a highly effective conservative treatment option for unicystic ameloblastoma, particularly in young patients, where preserving vital anatomical structures and supporting craniofacial growth are critical. Based on the findings of this case report, patient selection criteria should emphasize age, lesion type, and the potential impact on facial aesthetics and function.

Long-term follow-up is essential for monitoring bone regeneration and detecting potential recurrences. Regular imaging, such as CBCT, and careful clinical evaluations are recommended to ensure successful outcomes. Additionally, integrating minimally invasive techniques like marsupialization during limited healthcare resources, such as the COVID-19 pandemic, can provide a practical and patient-centered approach. This case highlights the importance of tailoring treatment strategies to individual patient needs and balancing effective lesion management with the preservation of quality of life.

REFERENCES

1. Soluk-Tekkesin M, Wright JM. The World Health Organization Classification of Odontogenic Lesions: A Summary of the Changes of the 2022 (5th) Edition. Turk Patoloji Derg 2022;38:168-84.

2. Robinson L, Martinez MG. Unicystic ameloblastoma: a prognostically distinct entity. Cancer 1977;40:2278-85.

3. Poonja K, Galinde J, Dave PM. Unicystic Ameloblastoma. Journal of Contemporary Dentistry 2015;5:40-2.

4. Philipsen HP, Reichart PA. Unicystic ameloblastoma. A review of 193 cases from the literature. Oral Oncol 1998;34:317-25.

5. Garcia NG, Oliveira DT, Rodrigues MT. Unicystic Ameloblastoma with Mural Proliferation Managed by Conservative Treatment. Case Rep Pathol 2016;2016:3089540.

6. Ooi A, Feng J, Tan HK, Ong YS. Primary treatment of mandibular ameloblastoma with segmental resection and free fibula reconstruction: achieving satisfactory outcomes with low implant-prosthetic rehabilitation uptake. J Plast Reconstr Aesthet Surg 2014;67:498-505.

7. Hammarfjord O, Roslund J, Abrahamsson P, Nilsson P, Thor A, Magnusson M, *et al*. Surgical treatment of recurring ameloblastoma, are there options? Br J Oral Maxillofac Surg 2013;51:762-6.

8. Lau SL, Samman N. Recurrence related to treatment modalities of unicystic ameloblastoma: a systematic review. Int J Oral Maxillofac Surg 2006;35:681-90.

9. Nowair IM, Eid MK. A modified surgical approach for the treatment of mandibular unicystic ameloblastoma in young patients. J Craniomaxillofac Surg 2020;48:148-55.

10. Zheng CY, Cao R, Hong WS, Sheng MC, Hu YJ. Marsupialisation for the treatment of unicystic ameloblastoma of the mandible: a long-term follow up of 116 cases. Br J Oral Maxillofac Surg 2019; 57:655-62

11. Caldeira PC, Schuch LF, Tavares TS, Santos-Silva AR, Vargas PA, Pérez-de-Oliveira ME, *et al*. Impact of the COVID-19 pandemic on public University laboratories of oral and maxillofacial pathology: A Brazilian multicenter study. Oral Dis 2022;28:2423-31.

12. Maffia F, Fontanari M, Vellone V, Cascone P, Mercuri LG. Impact of COVID-19 on maxillofacial surgery practice: a worldwide survey. Int J Oral Maxillofac Surg 2020;49:827-35.

13. Zheng C, Cao R, Hong W, Sheng M, Hu Y. Marsupialisation for the treatment of unicystic ameloblastoma of the mandible: a long-term follow up of 116 cases. British Journal of Oral and Maxillofacial Surgery 2019;57:655-62.

14. Datarkar A, Rai A, Bhawalkar A, Jain A. Clinical outcome following conservative treatment of 58 mandibular ameloblastoma patients: a retrospective study. Oral Maxillofac Surg 2023;27:601-8.

15. Srikar M, Sangamesh N, Vidya K. Marsupialization for the Management of Unicystic Ameloblastoma-Retrospective Study. Indian Journal of Public Health 2020;11:593.

16. Zhang X, Liu L, Yang X, Wang L, Zhang C, Hu Y. Expression of TP53 and IL-1α in unicystic ameloblastoma predicts the efficacy of marsupialization treatment. Medicine (Baltimore) 2018;97:e9795.

17. Pogrel MA, Montes DM. Is there a role for enucleation in the management of ameloblastoma? Int J Oral Maxillofac Surg 2009;38:807-12.

18. Seintou A, Martinelli-Kläy CP, Lombardi T. Unicystic ameloblastoma in children: systematic review of clinicopathological features and treatment outcomes. Int J Oral Maxillofac Surg 2014;43:405-12.

19. Scariot R, da Silva RV, da Silva Felix W, Jr., da Costa DJ, Rebellato NL. Conservative treatment of ameloblastoma in child: a case report. Stomatologija 2012;14:33-6.

20. Eckardt AM, Kokemüller H, Flemming P, Schultze A. Recurrent ameloblastoma following osseous reconstruction--a review of twenty years. J Craniomaxillofac Surg 2009;37:36-41.

21. Khare G, Kumar S. Application of osteomucoperiosteal flap in large unicystic ameloblastoma to promote early bone healing: An alternative to primary bone grafting. Contemp Clin Dent 2011;2:283-6.

22. Ogunsalu C, West W, Lewis A, Williams N. Ameloblastoma in Jamaica--predominantly unicystic: analysis of 47 patients over a 16-year period and a case report on re-entry cryosurgery as a new modality of treatment for the prevention of recurrence. West Indian Med J 2011;60:240-6.

23. de Paulo LF, Oliveira MT, Rodrigues Á R, Zanetta-Barbosa D. Treatment of an extensive unicystic ameloblastoma in a 7-year-old child: the best approach? Br J Oral Maxillofac Surg 2015;53:292-4.

24. Peter J, Emmatty TB, Jose B, Methippara JJ, Sebastian R, Vasu S. Unicystic Ameloblastoma Associated with Primary Mandibular Second Molar: A Case Report. Int J Clin Pediatr Dent Oral Dis 2022;28:2423-31.

25. Brown NA, Rolland D, McHugh JB, Weigelin HC, Zhao L, Lim MS, *et al*. Activating FGFR2-RAS-BRAF mutations in ameloblastoma. Clin Cancer Res 2014;20:5517-26.

26. Kaye FJ, Ivey AM, Drane WE, Mendenhall WM, Allan RW. Clinical and radiographic response with combined BRAF-targeted therapy in stage 4 ameloblastoma. J Natl Cancer Inst 2015;107:378.

27. Hirschhorn A, Campino GA, Vered M, Greenberg G, Yacobi R, Yahalom R, *et al*. Upfront rational therapy in BRAF V600E mutated pediatric ameloblastoma promotes ad integrum mandibular regeneration. J Tissue Eng Regen Med 2021;15:1155-61.

28. Hirschhorn A, Grynberg S, Campino GA, Dobriyan A, Patel V, Greenberg G, *et al*. Histopathologic and Molecular Insights Following the Management of Ameloblastomas via Targeted Therapies - Pathological and Clinical Perspectives. Head Neck Pathol 2024;18:129.

29. Sano K, Yoshimura H, Tobita T, Kimura S, Imamura Y. Spontaneous eruption of involved second molar in unicystic ameloblastoma of the mandible after marsupialization followed by enucleation: a case report. J Oral Maxillofac Surg 2013;71:66-71.

30. Ozkan A, Erguven SS, Bayar GR, Sencimen M. The impact of COVID-19 pandemic on patient satisfaction and clinical outcomes after treatment of odontogenic cysts with decompression followed by surgery. Stomatologija 2021;23:101-5.