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Management of large pediatric ameloblastoma: Conservative approach with 4-years follow up

Emtenan Abdulrahmman Almajid [a], Alia Khalid Alfadhel [b,*]

[a] Department of Oral & Maxillofacial Surgery, Prince Sultan Military Medical City, Riyadh, Saudi Arabia
[b] Al-Farabi College for Dentistry and Nursing, Riyadh, Saudi Arabia

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ABSTRACT

Ameloblastoma is a rare, benign, slowly growing, locally invasive, odontogenic tumor of epithelial origin with high recurrence rate. We present a case of large unicystic ameloblastoma in a 13-year old girl. The tumor involved the right mandibular body, ramus and condyle and surrounded the crown of an unerupted second molar. Conservative surgery was chosen as a first choice treatment in order to preserve the structural and functional integrity of the mandible. No recurrence was observed during a 4-year follow-up.

1. Introduction

Ameloblastoma is a rare benign odontogenic tumor of epithelial origin that is slowly growing, locally aggressive, with a high recurrence rate. It has a peak incidence in the third and fourth decades of life. It accounts for 11% of all odontogenic tumors, but it constitutes only about 1% of all tumors of the jaws [1,2].

Ameloblastoma commonly arises from the posterior region of mandible, but it may occur also in the maxilla; the incidence of mandibular to maxillary ameloblastomas is four to one [3]. It has a tendency to grow in a buccolingual direction and is often associated with unerupted third molar. Ameloblastoma usually presents as a slowly growing, painless swelling. However, other manifestations (such as facial deformity, dental displacement or impaction, malocclusion, soft tissue invasion, and loosening of teeth) may be found depending on the size of the tumor [4,5].

There are three types of ameloblastoma: the intraosseous solid or multicystic lesion, the unicystic type, and the peripheral (extraosseous) ameloblastoma. It is important to differentiate between the three types as each type is treated differently [3].

Imaging studies may show ameloblastoma as a corticated unilocular or, more commonly, multilocular, well-demarcated radiolucency, surrounding the crown of an unerupted tooth. Histopathological examination is essential for diagnosis because it may appear like an odontogenic cyst on clinical or radiologic examination [6]. Moreover, histopathological examination helps to identify the subtype of unicystic tumors as the prognosis differs according to the subtype, a factor that may influence the choice of treatment. [7]

The treatment of ameloblastoma is controversial. Several surgical modalities, including both conservative and radical approaches, have been performed with varying results. Long term follow up is essential because recurrence may occur many years after the primary surgery [6,8].

In this case report, we present a 13-year old female patient with a large unicystic ameloblastoma and discuss our approach to treatment. This case had uncommon features that deserved to be reported including the young age of the patient and the absence of recurrence after conservative surgery despite being a mural variant of unicystic ameloblastoma.

* Corresponding author. Al-Farabi College for Dentistry and Nursing, Riyadh, Saudi Arabia.
E-mail address: 200911289@alfarabi.edu.sa (A.K. Alfadhel).

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1.1. Case description

In 2014, a Saudi 13-year old girl, not known to have any medical problems, was referred with a right mandibular swelling to the Oral and Maxillofacial Surgery Department, Prince Sultan Military Medical City, Riyadh, Saudi Arabia. The patient complained from progressive swelling and discomfort with no pain or numbness. Past history was negative for intake of medication or surgical intervention.

Clinical examination of the patient revealed a large swelling that involved the right ramus and body with buccolingual extension. No fistula, discoloration, or palpable lymph nodes were found. The patient had normal mouth opening with slight deviation to the right side. Intra oral findings included missing second and third molars and shifting of lower mandibular dental midline to the left side.

A panoramic radiograph showed an extensive radiolucent area involving the right mandibular body, ramus, and condyle and surrounding the crown of unerupted second molar. The lower third molars were congenitally missing (Fig. 1). The CT scan of the head and neck region with contrast dye and 3D reconstruction showed large right mandibular expansile cystic lesion with cortical bony dehiscence that measured about 3.7×8.2 cm (Fig. 2).

The differential diagnosis of this mass was either ameloblastoma or odontogenic keratocyst tumor. The diagnosis was confirmed with histopathological examination, which showed unicystic ameloblastoma with mural invasion (Fig. 3).

The treatment plan was discussed with the patient and her family. The treatment options included either to do hemi-mandibularectomy with safety margins to avoid recurrence as definite treatment or to perform conservative surgical excision with extraction and close follow up for lifelong. The patient and her family chose the second plan, and an informed consent was obtained from her parent. Under general anesthesia the patient underwent surgical excision of the tumor, extraction of second molar, and osteoplasty with inferior alveolar nerve preservation. She was prescribed only antibiotics and analgesics after the operation and she had a smooth postoperative period. After two days, the patient was discharged from the hospital. Both the patient and her family were instructed about the routine follow up, both clinically and radiographically, every 6 months for 5 years after the surgery, then annual for life long. Now four years have passed since the surgery and the patient is showing complete bone remodeling with no signs of recurrence (Fig. 4).

2. Discussion

Ameloblastoma is a rare, slowly growing, benign, epithelial odontogenic tumor of the jaws. We presented in this case a unicystic ameloblastoma in a 13 year old girl. One of the special features in this case was the young age of the patient, as the tumor is reported to be uncommon in children and usually presents in the third and fourth decades of life [9]. Pediatric ameloblastoma accounted for 8.7%–15% of all ameloblastomas in western countries [10,11] and accounted for 14.6%–25% in Asian and African countries [12,13].

Because of the relative rarity of this tumor, its aggressive behavior, and the high recurrence rate, its surgical treatment is controversial with no satisfactory evidence proving which treatment modality is the most effective [9,14].

Conservative surgery for unicystic ameloblastoma usually consists of Enucleation [15,16] and curettage [15]. Some surgeons used marsupialization [17–19] to reduce the size of the lesion, followed by second stage surgery. More aggressive treatment (radical resection) involves resection of the tumor with a safety margin of 1 cm or even hemi-mandibularectomy [20].

Unlike the radical approach, conservative surgery is usually associated with a high recurrence rate. Some studies have argued that unicystic ameloblastomas are better to be treated by resection with a safety margin, to avoid recurrence as some subtypes may invade surrounding tissues [20]. Nevertheless, another point of view regards that recurrence is not the most important consideration in case of children as even recurrent cases were shown to require less aggressive treatment than that would have been performed for initial lesion [18,21,22].

The choice of the surgical modality should take many factors into consideration, including: the clinical subtype, its location in the jaws, its size, the patient's age and the patient's availability for follow up examinations [23–25]. We will proceed to discuss these factors and how we reached the treatment choice in our patient.

The histological type of the tumor is a decisive risk factor for its recurrence and hence the effectiveness of the recommended treatment. Higher recurrence rates have been reported for certain types of ameloblastoma regardless of the nature of treatment [17,26]. Unicystic ameloblastoma is believed to have a better prognosis because it is less aggressive and has a more favorable response to conservative surgery than the multicystic or solid types [27].

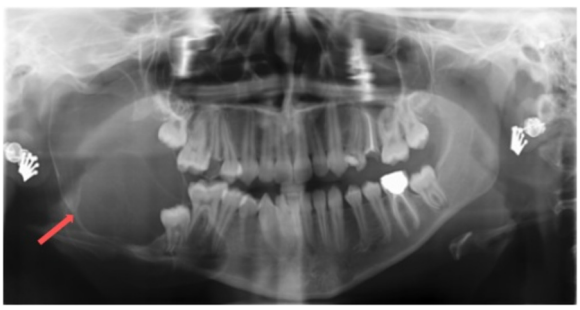

Fig. 1. A panoramic radiograph showing the presence of an extensive radiolucent area involving the right mandibular body, ramus, and condyle, suggestive of a cystic lesion.

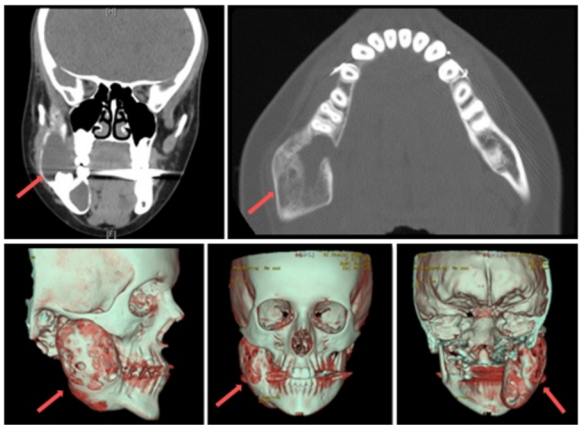

Fig. 2. Computerized tomography & three-dimensional images showed a large expansile cystic lesion replacing the ramus of the right mandible with cortical bony dehiscence.

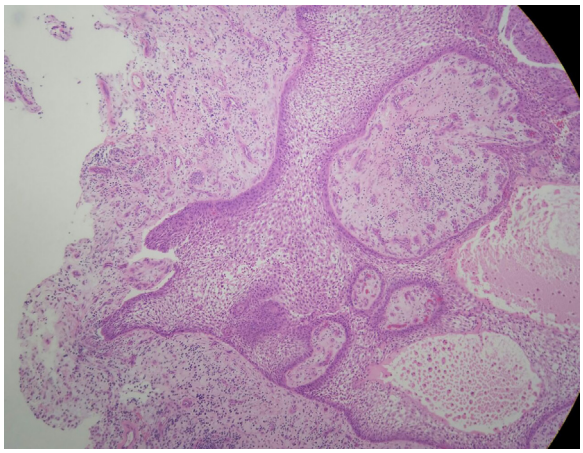

Fig. 3. Histopathological image showed unicystic ameloblastoma with mural invasion, which confirms the diagnosis (hematoxylin and eosin stain, 200×).

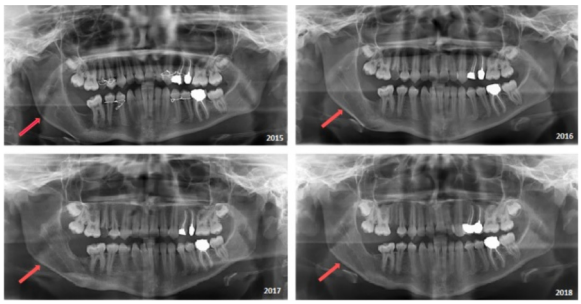

Fig. 4. Advanced bone remodeling at 4 years follow-up showing no signs of recurrence; remodeling is visible.

Unicystic ameloblastomas exhibit four subtypes. The luminal and intraluminal varieties are treated surgically by enucleation and has a good prognosis because the tumor is confined by the fibrous connective tissue wall of the cyst. However, mural and invasive unicystic ameloblastomas extend to involve the surrounding connective tissue wall of the cyst and, therefore, they can invade the surrounding bone. Hence, some studies recommended treatment of the mural and invasive varieties by resection [7]. However, the tumor in our case was diagnosed as the mural subtype and the patient underwent conservative surgery, with no recurrence until the time of this report (four years after surgery).

The tumor in this patient aroused on the right side in the posterior region (involving the right body, ramus and condyle), which is the most commonly affected site [5]. Ameloblastomas that arise in the posterior maxilla require aggressive surgical treatment as they are

near to vital structures and treatment of future recurrences is difficult in this site [23].

The size of the tumor in our patient was relatively large, but corresponds to sizes reported by other case series [18]. The painless nature of the tumor allows it to attain large size before being detected. This adds to the challenges of treatment as it becomes difficult to detect the tumor early enough to provide a treatment modality that preserves the continuity of the mandible [10].

Ameloblastomas that present during childhood poses some special problems for surgical treatment. The growth of the jaw, behavior and prognosis of the tumor are different than adults. In children, it is very important to preserve the facial bone structure in order to maintain the aesthetic appearance among other children, allow continued mandibular growth and to ensure proper functioning. Therefore, minimal surgical manipulations should be considered for pediatric oral and maxillofacial benign tumors [9,18,19,28,29]. Radical resection could result in facial deformity, masticatory dysfunction, irregular jaw movement and negative psychological impact. Therefore, despite the high success rate for radical resection of the tumor, the conservative approach was favored by our team, and it resulted in no recurrence (despite being a mural variant), besides an excellent postoperative function and aesthetics.

Similarly, some previous studies have reported the use of conservative surgery for children with unicystic ameloblastoma with no recurrence during the follow up for 2–15 years after the surgery [9,18]. On the other hand, some studies have reported varying rates of recurrence after conservative surgery for the unicystic tumor [10,11,30,31]. These controversial reports are likely the results of differences between the studies as regards the subtype of the unicystic tumor, the site and the exact surgical procedure used. Resection for unicystic ameloblastoma resulted in the lowest recurrence rate (3.6%). Enucleation followed by the application of Carnoy's solution has resulted in a recurrence rate of 16%, which was the best, except for resection. Marsupialization followed by second procedure resulted in an 18% recurrence rate. The use of enucleation alone resulted in the highest recurrence rate (30.5%) [14].

Owing to the tumor's liability to recur after surgical treatment, follow up for a long time is recommended. Follow up depends on annual clinical examination and performance of imaging studies. The imaging modalities used are OPG, CT scan, MRI. CT scan can detect small tumors and show the extension of the tumor into the surrounding organs [32,33]. Magnetic resonance imaging is comparable to CT scan [34] However, the use of CT or MRI for follow up for a long period may be difficult due to issues related to availability and cost. Orthopantomograms are less likely to detect recurrences early and they cannot distinguish cystic expansion from erosion of adjacent bone [35]. Both OPG and CT scan have the hazard of increased risk of radiation – and possibly of tumors of the head and neck [36–38]. The readiness of the patients or the family to comply with follow up should be ensured to determine the treatment modality.

Conclusion: In children, the conservative surgical approach is the best treatment option despite the potential of recurrence. Radical resection can cause significant negative functional and aesthetic outcomes. The aim of treatment is always to preserve the structural and functional integrity whenever possible.

Limitation: This case report is limited by the period of follow up (four years in our case) as the tumor may recur much later in life.

Declaration of interest

The authors declare no conflict of interest.

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This work received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Ethical approval

This work has been approved by the Ethical Committee of Prince Sultan Military Medical City, Riyadh, Saudi Arabia (ethics approval no. 1030).

Patient consent

Written consent was obtained from the patient.

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