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Ameloblastoma of the jaws: A critical reappraisal based on a 40-years single institution experience

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SUMMARY

The 40-years of experience in a single institution with the treatment of previously untreated ameloblastoma have been reported, followed by a management protocol. Retrospectively, 25 consecutive patients treated between 1969 and 2009 have been analyzed. In 11 patients, a preoperative diagnosis of ameloblastoma was available. In the remaining 14 patients the diagnosis of ameloblastoma was a postoperative one. For the recurrence rate a minimum follow-up period of 5 years was observed; 20 patients met these criteria.

After primary radical surgery in five patients, no recurrences were observed. In case of conservative surgical treatment, performed in 15 patients, a recurrence was observed in eight (53%) of them. Six of these patients were then treated successfully by radical surgery, while two patients refused such surgical approach. In one of the patients with a recurrence a cervical lymph node metastasis was detected at the same time, resulting in a diagnosis of metastasizing ameloblastoma.

Introduction

In the World Health Organization Classification of Odontogenic Tumours a distinction is made between benign ameloblastoma and malignant ameloblastoma.[1] Within the group of benign ameloblastomas four subtypes are recognized, being the solid/multicystic, the desmoplastic, the unicystic and the extraosseous/peripheral type, respectively.[1] By far the majority of ameloblastomas are located within the jaw bones. The peripheral ameloblastoma is extremely rare, indeed. This type will not be discussed here any further.

Malignant ameloblastomas are extremely rare; in the WHO classification two main subtypes are recognized, being (1) the metastasizing ameloblastoma and (2) ameloblastic carcinoma.

The estimated incidence of ameloblastomas is approximately 0.5 per million population per year. There is no distinct gender predilection. Most cases are diagnosed between 30 and 60 years of age. Unicystic ameloblastomas are more common under the age of 20 years.[2] There are no well established etiologic factors. The posterior region of the mandible is the site of predilection. In approximately 40% of the cases there is an associated unerupted tooth, often the mandibular third molar. Ameloblastomas may remain asymptomatic before a facial swelling develops.

Ameloblastomas may present on conventional radiographs as a unilobular or multilobular corticated radiolucency resembling a cyst. Bony septae may result in a honeycomb appearance. Buccal and lingual expansion is more common in ameloblastoma than in keratocystic odontogenic tumours.[3] Resorption of roots may or may not be present. The radiographic differential diagnosis includes a variety of odontogenic cysts, a keratocystic odontogenic tumour, an odontogenic myxoma, as well as non-odontogenic tumours and cysts, such as a central giant cell granuloma and a simple bone cyst, respectively. Computed tomography and MRI may be helpful in establishing the extent of the lesion, particularly when located in the maxilla.

The preferred treatment of ameloblastoma is wide surgical removal with the possible exception for unicystic ameloblastoma, as will be discussed later.

In this study the 40-years single institution experience with the management of primary ameloblastoma will be reported and discussed in relation to the present concepts of this odontogenic tumour.

Patients and methods

Twenty-five consecutive patients, 14 men and 11 women, diagnosed with a histologically benign, previously untreated ameloblastoma at the Department of Oral and Maxillofacial Surgery between September 1969 and September 2009 have been included. The information retrieved from the files included age, gender, localization, preoperative diagnosis, histopathological subtype,

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type of surgery and follow-up information (Table 1). In 11 patients, a preoperative diagnosis of ameloblastoma was available. They all were advised to have radical surgery; three of them refused such treatment and have been treated by enucleation, instead. In the remaining 14 patients the diagnosis of ameloblastoma became apparent only after conservative surgical removal.

Routine follow-up consisted of annual clinical and radiographic examination, occasionally extended with CT-scans or MRI in cases of maxillary involvement, up to a period of at least 10 years. One patient primarily treated by radical surgery was lost to follow-up after 3 months due to refusal of the patient. With regard to treatment results only patients with a minimum follow-up period of 5 years were included; 20 patients fulfilled these criteria.

In the same period, 11 patients with a recurrent ameloblastoma have been observed who had been treated elsewhere previously. Furthermore, one patient with a primary malignant ameloblastoma has been recorded; one other patient was noticed with a peripheral ameloblastoma. These latter two patients will not be discussed here any further.

In view of the small number of patients no statistical analysis has been performed.

Results

These results are summarized in Table 2. Of the 20 patients who had been followed-up for at least 5 years, local recurrence was observed in eight (53%) of the 15 patients who had been treated by enucleation, seven of them within 5 years and one after 8.3 years. Six of these eight patients have additionally been treated by radical surgery; no second recurrences were observed in this group of patients during a mean follow-up period of 6.3 years. However, in one of these six patients a metastatic lymph node was detected during the reconstruction of the mandible, resulting in a diagnosis of metastasizing ameloblastoma. This patient has been discussed in detail elsewhere.[4] Two patients with a recurrence after enucleation refused radical surgery and were again treated by conservative surgery. Both patients experienced multiple recurrences thereafter.

In the five patients who had been initially treated by radical surgery and being followed for at least 5 years, no recurrences were observed.

Discussion

The demographic data of the presently reported patients are more or less in accordance with those obtained from the literature. Three patients with a unicystic ameloblastoma were observed; in only one of these patients the cystic lesion could be removed as an entirely intact specimen without any fragmentation during removal. In one patient, a desmoplastic ameloblastoma was observed. In one other patient a metastasizing ameloblastoma was diagnosed; the metastatic cervical lymph node was detected as an incidental finding during reconstructive surgery of the mandible because of a local recurrence 8 years after enucleation of a histologically benign ameloblastoma.

Table 1
Characteristics of 25 patients.

Gender	Male	14
	Female	9
Age (in years)		34.4 (range 12–70)
Localization	Mandible	20
	Maxilla	5
Diagnosis	Preoperative	11
	Postoperative	14
Histological subtype	Follicular	10
	Plexiform	7
	Follicular/plexiform	4
	Desmoplastic	1
	Unicystic	3
Type of surgery	Conservative	17
	Radical	8

Table 2
Recurrence in patients with a minimum follow-up period of five years ($n = 20$).

Type of treatment	Number of patients	Recurrence (%)
Conservative surgical removal (includes enucleation and/or curettage)	15	8* (53%)
Radical surgical removal	5	– (–)

* In one patient a cervical lymph node was encountered, resulting in a diagnosis of metastasizing ameloblastoma.

Treatment

In the present series a preoperative diagnosis of ameloblastoma was obtained in 11 of 25 patients. Of these 11 patients three refused to undergo radical surgery. In the remaining 14 patients the diagnosis of ameloblastoma became available only after enucleation of a unicystic or multicystic/solid lesion. In none of these 14 patients immediate additional radical surgery has been performed.

There are no randomized trials of *solid/multicystic* ameloblastoma with regard to the type of surgical treatment, varying from simple enucleation, marsupialisation followed by resection or curettage,[5] to aggressive removal, including a margin of one tissue plane in soft tissues when cortical perforation is present, clinically or radiographically.[6] In general, resection with a 1.5–2 cm margin beyond the radiological limit seems a safe procedure for the solid/multicystic type.[7] Nevertheless, the debate on the preferred treatment is ongoing.[8,9] In our institution, we have adopted a more aggressive surgical approach in the last decade, probably influenced by the excellent results of immediate reconstruction of the jaws and the strongly improved possibilities of prosthetic rehabilitation by the use of dental implants.

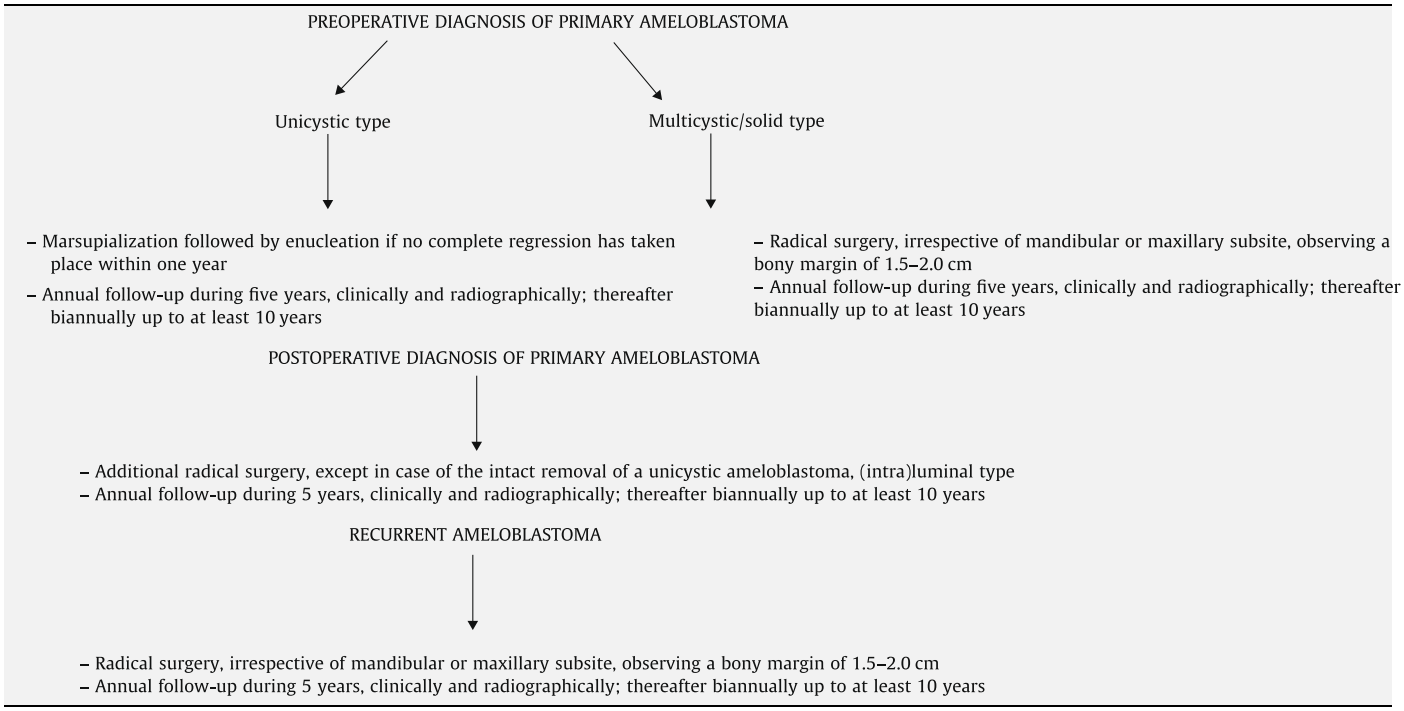
For the reporting on ameloblastomas it seems acceptable to group the treatment regimens into three modalities, being (1) conservative (includes enucleation and curettage), (2) marsupialization, and (3) radical surgery (includes resection with or without continuity defect).

In case of a preoperative diagnosis of solid/multicystic ameloblastoma in *children*, it is tempting, mainly for psychosocial reasons, to perform less aggressive surgery. Furthermore, some authors have suggested that ameloblastomas in children may behave less aggressively.[10] At the same time, one may advocate not to be less aggressive in view of the high recurrence rate after conservative surgery and the long life expectancy at childhood.

In case of a *unicystic* ameloblastoma, based on an incisional biopsy, some authors recommend decompression followed by enucleation within 3–4 months.[11–13] However, a preoperative biopsy in unicystic ameloblastoma is not always representative[14]; in fact, Ackerman et al. recommend not to perform such an incisional biopsy in these circumstances.[15] In the past, unicystic ameloblastomas have been reported to behave less aggressively than the solid/multicystic type.[16] However, in several recent studies this view has been challenged.[6,17,18]

Little is known about the role of postoperative radiotherapy in case of incomplete removal.[19] There are no reliable data about the possible value of additional treatment modalities such as the use of liquid nitrogen or tissue fixatives such as Carnoy's solution,[6] although such practice is recommended by some authors.[11,12]

Table 3
Management of benign ameloblastoma.



Comments:
1. The limited amount of tissue obtained during marsupialization may histopathologically not be representative for the entire lesion.
2. The use of fresh frozen sections for establishing a possible diagnosis of ameloblastoma is discouraged.
3. There is insufficient evidence that ameloblastomas in children can be treated less aggressively than in adults.
4. There is insufficient evidence to advice the use of adjunctive treatment, such as liquid nitrogen or Carnoy's solution, after surgical removal of an ameloblastoma
5. A "wait-and-see" policy after conservative removal of a multicystic/solid ameloblastoma in the body/symphysis region of the mandible or the anterior maxilla (between the first molars) is discouraged, since this policy carries the risk of uncontrollable disease.
6. The remote risk of regional or distant metastases (e.g. lungs) does not warrant routine studies for such events at follow-up.
7. Radiotherapy should only be considered in uncontrolled disease.

Recurrence

Eight of the 15 (53%) patients, primarily treated by enucleation and followed-up for at least 5 years, developed a recurrence of whom six were additionally treated by radical surgery; no second recurrences were observed in these six patients during a mean follow-up period of 6.3 years.

In a large series reported from South Korea, the follicular, granular cell and acanthomatous types had a relatively high likelihood of recurrence, while the desmoplastic, plexiform and unicystic types showed a relatively low potential for recurrences.[10] However, in the meta-analysis from Pogrel and Montes the histopathological subtypes, including unicystic ameloblastomas, did not seem to be relevant in this respect.[6] Partly based on our own experience, but also based on a number of large studies performed by others, the chance of recurrence seems to be more dependent on the method of surgical treatment rather than the histological subtype.[20]

Follow-up regimen

In our series, most recurrences were observed within a follow-up period of 4 years. In general, annual follow-up for at least 10 years is recommended. Others recommend annual follow-up until 5 years and every 2 years thereafter for at least 25 years.[11,21]

For the mandible the use of a panoramic view seems adequate. Unfortunately, the panoramic view is not very suitable for the detection of a possible recurrence in the maxilla or the maxillary sinus. In fact, in this location CT-scans are by far superior. In order to eliminate ionizing radiation exposure MRI may be an ever better imaging tool for maxillary lesions.

Management protocol

Based on the experiences reported in the literature and also on our own experience a treatment protocol has been proposed in Table 3.

Conflict of Interest Statement

None declared.

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