

Oral Health Division Ministry of Health Malaysia

MANAGEMENT OF AMELOBLASTOMA



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STATEMENT OF INTENT

The clinical practice guidelines are meant to be a guide for clinical practice, based on the best available evidence at the time of development. Adherence to these guidelines may not necessarily lead to the best clinical outcome in individual patient care. Every healthcare provider is responsible for the management of their patient based on the clinical presentation and management options available locally.

These guidelines were first issued in December 2015 and will be reviewed in 2019 or earlier if new evidence becomes available.

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GUIDELINES DEVELOPMENT AND OBJECTIVES

The Development Group for this Clinical Practice Guidelines (CPG) consisted of Oral Maxillofacial Surgeons, Oral Pathology and Oral Medicine Specialists, Paediatric Dental Specialists, Dental Public Health Specialists and Dental Officers. Clinical audit indicators have also been identified for the purpose of monitoring and evaluating outcomes.

Evidence was retrieved from publications from the 1972 onwards. Literature search was carried out using the following electronic databases: PUBMED/MEDLINE; Cochrane Database of Systemic Reviews (CDSR); Health Technology Assessment (HTA) and full text journal articles via OVID search engine. In addition, the reference lists of all relevant articles retrieved were searched to identify further studies. The search process was conducted between May 2012 and May 2013 and only literatures in English were included.

All articles retrieved were appraised by at least two members and graded according to the levels of evidence presented in the form of evidence tables and discussed during group meetings. The levels of evidence table were adopted from the modified version of the United States (U.S) / Canadian Preventive Services Task Force, while the grading of recommendations was based on the modified version of the Scottish Intercollegiate Guidelines Network (SIGN). The CPG was based on the findings of relevant published evidence.

The draft was reviewed by a team of internal / external reviewers and was available on the websites of the Ministry of Health, Malaysia and Academy of Medicine, Malaysia for comments and feedback. Recommendations were presented to the Technical Advisory Committee for CPGs, and finally to the Health Technology Assessment and CPG Council, Ministry of Health, Malaysia for approval.

OBJECTIVE

To provide evidence-based guidance in the management of ameloblastoma

SPECIFIC OBJECTIVES

- To disseminate and reinforce knowledge on the management of ameloblastoma among healthcare professionals
- To provide timely and appropriate management of ameloblastoma by healthcare professionals

CLINICAL QUESTIONS

The clinical questions addressed by the guidelines are:

- i. What is the aetiology of Ameloblastoma?
- ii. How is ameloblastoma classified according to WHO?
- iii. What are the clinical presentations of Ameloblastoma?
- iv. What are the investigations in diagnosing ameloblastoma?
- v. What are the factors determining mode of treatment?
- vi. Is there any difference in management based on specific group?
- vii. What method of treatment results in lowest recurrence rate?
- viii. What is the recommended protocol for follow up?

TARGET POPULATION

This guideline is applicable to all patients with ameloblastoma

TARGET GROUP

The guideline is applicable to all healthcare professionals involved in the management of ameloblastoma.

HEALTHCARE SETTINGS

Hospitals with adequate facilities and specialist support.

PROPOSED CLINICAL AUDIT INDICATORS FOR QUALITY MANAGEMENT

Proportion of patients with recurrence of ameloblastoma:

* Recurrence:

Reappearing of symptoms or lesions after an intermission or remission.

LEVELS OF EVIDENCE AND GRADES OF RECOMMENDATIONS

LEVEL	STUDY DESIGN			
1	Evidence obtained from at least one properly designed randomised controlled trial.			
II-1	Evidence obtained from well - designed controlled trials without randomization.			
II-2	Evidence obtained from well - designed cohort or casecontrol analytic studies, preferably from more than one centre or research group.			
II-3	Evidence obtained from multiple time series with or without the intervention. Dramatic results in uncontrolled experiments (such as the results of the introduction of penicillin treatment in the 1940s) could also be regarded as this type of evidence.			
Ш	Opinions or respected authorities, based on clinical experience; descriptive studies and case reports; or reports of expert committees.			

Source: Adapted from U.S./Canadian Preventive Services Task Force

GRADES OF RECOMMENDATION

Α	At least one meta analysis, systematic review or RCT or evidence rated as good or directly applicable to the target population		
В	Evidence from well conducted clinical trials, directly applicable to the target population and demonstrating overall consistency of results; or evidence extrapolated from meta analysis, systematic reviews or RCT.		
С	Evidence from expert committee reports, or opinions and or clinical experiences of respected authorities; indicates absence of directly applicable clinical studies of good quality.		

Source: Modified from the Scottish Intercollegiate Guidelines Network (SIGN)

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GLOSSARY

Ameloblastoma	A benign but locally aggressive tumour of odontogenic epithelium arising from the mandible, or less commonly from the maxilla.		
Carnoy's solution	Slightly yellow liquid with chloroform odour.It is composed of 3 ml of chloroform (30%),6 ml of absolute ethanol (Ethyl Alcohol 60%),1 ml of glacial acec acid (Acec Acid 10%), and1 g of ferric chloride.		
Central giant cell granuloma	A rarely aggressive idiopathic benign intraosseous lesion that occurs almost exclusively in the jaws. This osteolytic lesion histologically consists of pro-liferaon of fibrous tissue, hemorrhagic focuses, hemosiderin deposits, osteoclast-like giant cells, and reactive bone formation.		
Cryotherapy	A treatment in which the lesion is frozen using liquid nitrogen.		
A thickened epithelial band along the margin of to gum, in the embryo, from which the enamel organs a developed.			
Dredging	A conservative surgical procedure in which, after deflation and enucleation or only enucleation, repeated dredging is applied to accelerate new bone formation by removing out the scar tissue from the bony cavity.		
Enucleation	Removal of entire lesion or tumour from an enveloping cover or sac in such a way it comes out clean and whole.		
An epithelial remnant of Hertwig's sheath in periodontal membrane, which sometimes devinto dental cyst.			

Exenteration	Surgical removal of the inner organs, commonly used to indicate radical excision of the contents of a body cavity.		
Malocclusion	Teeth which are not properly aligned.		
Marsupialisation	Marsupialisation is the surgical technique of cutting a slit into a cyst and suturing the edges of the slit to form a continuous surface from the exterior to the interior of the cyst. Sutured in this fashion, the cyst remains open and can drain freely.		
Maxillary A surgical obturator replaces missing dentoal and/or palatal structures. They are usually us close oronasal and/or oroantral communic following ablative cancer surgery or trauma.			
Metastasize To form new foci of disease in another part of t			
Obturator	A prosthetic device serves to close an opening in the body.		
Odontogenic cyst	Jaw cyst that is formed from tissues involved in odontogenesis (tooth development).		
Odontogenic myxoma	An uncommon benign tumour of jaw, apparently arising from the mesenchymal portion of tooth germ.		
Odontogenic tumour	Tumours in the jaws that arise from odontogenic (tooth forming) tissues		
Odontomas A tumour originating from a tooth and containing dental tissue (as enamel, dentin, or cementum).			

Osseointegrated implants	The process of bone growth right up to the implant surface. No soft tissue connects the bone to the surface of the implant. No scar tissue, cartilage or ligament fibres are present between the bones and implant surface. The direct contact of bone and implant surface can be verified microscopically.		
Ostectomy	Excision of bone or part of bone.		
Pedicle flap	A flap consisting of the full thickness of the skin and the subcutaneous tissue, attached by tissue through which it receives its blood supply.		
Prognosis	A prediction of the probable course and outcome of a disease/ the likelihood of recovery from a disease.		
Recurrence	Return of the disease after treatment and after a period of time.		
Resection	Surgical removal or excision of a portion of an organ or structure.		
Unicentric	Pertaining to or having a single centre (as of origin or dispersal).		

8.8. Age

Because the craniofacial bones, teeth, and soft tissues of children and adolescents are not completely developed, aggressive surgery has the potential to cause irreversible deformities. Clinicians must not only treat the disease but also consider the aesthetic and functional results in the light of the postoperative growth and development of these young patients. Although excisional surgery achieves low recurrent rates, ^{48, level III} it may cause facial deformity and dysfunction, affecting a young patient's physical and psychological development. Conservative treatment can achieve good results, either curing the disease or effectively controlling the progression of the disease until maturity allow for complete excision. ^{49, level III}

Recommendation 2

 Solid and multicystic ameloblastomas should be treated by surgical resection with a 1cm-1.5cm margin.

Grade B

 Unicystic ameloblastomas should be treated by enucleation coupled with an adjunctive procedure with Carnoy's solution or liquid nitrogen.

Grade B

Maxillary ameloblastomas should be treated by resection.

Grade B

1. INTRODUCTION

Ameloblastomas are tumours that originate from odontogenic epithelial tissue which are involved in the tooth formation and classified as benign odontogenic tumours by the World Health Organization (2005).^{1, level III} It is the most commonly encountered odontogenic tumour when odontomas are excluded. ^{2, level 14} They rarely develop into a malignant growth and metastasize but they can result in lesions that lead to severe abnormalities in the jaw and face. ^{3, level III} Due to variation in the method of treatment, recurrence rate ranges from 3.6% to 100%. ^{4-9, level III}

2. AETIOLOGY

It is generally agreed that ameloblastoma arise from remnants of odontogenic epithelium that failed to regress during embryonic development. They may arise from rests of dental lamina, epithelial rest cell of Malassez or enamel forming organ. If these cell rests are situated outside the bone in the soft tissues of gingival and edentulous alveolar mucosa, they may give rise to peripheral ameloblastoma. It is also agreed that solid multicystic ameloblastoma may arise as a result of neoplastic changes in the lining or wall of a non-neoplastic odontogenic cyst, in particular dentigerous and odontogenic keratocysts. ^{4, level III; 10, level II-3}

3. CLASSIFICATION AND HISTOLOGICAL FEATURES

World Health Organization (WHO) Classification of Odontogenic Tumours, ameloblastoma has been distinctly classified into 2 groups: benign ameloblastomas and malignant ameloblastomas. 2, level III

Table 1: Classification of ameloblastoma

TYPE OF AMELOBLASTOMA		CLINICAL FEATURESAND HISTOPATHOLOGY	
		A: BENIGN VARIANTS	
Ameloblastoma (SMA) High rate of recurrence if not adequal 2,11-12, level III It has 2 basic histopathological patt and plexiform, along with other variation the spindle cell ameloblastoma ameloblastoma, granula ameloblastoma, granula ameloblastoma. For the pathologist, it is essential variety of histological pattern in order the tumour as ameloblastoma. Ho patterns have no value in determinant.		High rate of recurrence if not adequately excised.	
2	Unicystic Ameloblastoma (UA)	 Presents as a cyst. 3 histological sub variants. ^{2-4,level III; 15,level II-3} Luminal (ameloblastomatous cyst lining) Intraluminal (protruding into cyst cavity) Mural (invading into cyst wall as islands of ameloblastoma or focal invasion from the ameloblastomatous lining) Distinction is important in predicting chances of recurrences. 	

 Luminal and intraluminal forms are confined by fibrous cyst wall May be removed completely if enucleated Thus, these UA variants enjoy good unlike the mural form which prognosis behaves like a SMA. 2; 11-12; 4, level III It is also important to be reminded that a preoperative incisional biopsy of UA is often not representative for the entire lesion and the true nature (whether the tumour has breached the fibrous cyst wall elsewhere) may only be evident when the entire specimen is available for microscopic examination. 2, level III Finally, microscopic diagnosis of UA can sometimes be difficult, and may not satisfy the criteria of Vickers and Gorlin or Robinson and Martinez. 13; 8-9, level III If the tumour is associated with an unerupted tooth it can be difficult to distinguish from dentigerous cyst clinically and radiologically. 2-9, level III Extraosseous counterpart of the intraosseous 3 Peripheral SMA. Ameloblastoma Occurring in the soft tissues covering the tooth (PA) bearing parts of the jaws. Features the same histomorphological cell types and patterns seen in SMA. 2, level III; 16, level II - 3 This variant lacks the persistent invasiveness of the SMA.

		Do not tend to recur if adequately excised. 2; 11-12; 4, level III; 10, level II - 3		
4	Desmoplastic Ameloblastoma (DA)	 Have specific clinical, imaging and histological features. ^{2, level III} Often found in the anterior/premolar regions of jaws with about 50% presenting radiologically as mixed radiolucent-radiopaque lesion similar to that of benign fibro-osseous lesions. Histologically, consists of irregularly shaped islands, stellate and cords of odontogenic epithelium embedded in desmoplastic connective tissue stroma. ^{2, level III ; 17- 18, level II - 3} 		
	B: MALIGNANT VARIANTS			
1	Metastasizing Ameloblastoma	 An ameloblastoma that metastasizes in spite of a benign histologic appearance. Diagnosis can only be made retrospectively, after the occurrence of metastatic deposits which are mostly seen in lungs. Thus, it is the clinical behaviour and not histology that justifies a diagnosis of metastasizing ameloblastoma. ^{2, level III} 		
		 Characterized by malignant cytologic features in 		

4. EPIDEMIOLOGY

4.1. Relative frequency

One of the largest retrospective study of ameloblastoma published in 1995 noted that the relative frequency of ameloblastoma in correlation to odontogenic tumours is between 11 and 92 percent, and that ameloblastoma forms 1 percent of all tumours and cyst of the jaws. ^{10, level II-3}

In one of the earliest study of ameloblastoma in Malaysia, it was observed that the tumour formed 1.1 percent of all oral pathology cases reported. ^{5, level III} When the frequency of ameloblastoma was correlated to all odontogenic tumours and cysts, Malaysia was noted to have a rate of 12.4 percent as compared to neighbouring Thailand with 6.7 percent, and Japan with 3.3 percent. ^{19, level III}

4.2. Racial and geographical distribution

The geographical differences noted in Siar and Ng's earlier study in 1993 has also been described in other reports. ^{2; 19, level III; 10, level II-3} Racial differences in the distribution of ameloblastomas were also observed with relative frequency of 24.8% seen in Caucasians, 34.4% in Blacks and 38.4% in Asians. ^{10, level II-3} In Malaysia, racial differences in the distribution of ameloblastoma among the various ethnic groups has recently been reported, with Malays accounting for 47.6%, Chinese 34.8%, Indians 7.0%, and remaining ethnic groups 10.6%. ^{14, level III}

4.3. Age

In Malaysia, patients diagnosed with ameloblastoma had a wide age range of between 7 to 85 years, with mean age of diagnosis at 30.3±16.3 years, and peak incidence in the second decade of life. ^{14, level III}

The younger patients seen in Malaysia when compared to the average age of 35.9 years, concur with Reichart et al's (1995) findings that patients from developing countries present with the disease at an earlier age as compared to those from industrialized countries. ^{10, level II - 3}

Ameloblastoma is uncommon in children. The occurrence was reported between 8.7% - 19.7% with the average age at diagnosis of 14.5 years, and less than 10% of childhood cases occurred under the age of 10 years. 20-24, level III It is suggested that the slow growth of this tumour could indicate that many adults diagnosed in their mid-30s had tumours since childhood. 13, level III

4.4. Gender

The gender ratio (male:female) when all ameloblastomas are considered in Malaysia is 1.4:1, while the ratio reported in one of the largest series of ameloblastoma was 1.14:1. 10, level II-3; 14, level III

4.5. Sites

Local studies of ameloblastoma have noted that the tumour predominantly occurs in the mandible (90.6%-91.5%), located mainly in the body and posterior region of mandible. ^{19, 14, level III} There are some variations in localization among the subtypes of ameloblastoma. The SMA and UA variants are predominantly found in the body and posterior part of mandible while the DA shows a preference for the anterior jaw segment. ^{14, level III}

5. CLINICAL CHARACTERISTICS

5.1. Clinical examination

The diagnosis of the ameloblastoma is achieved by the clinical examination based on the signs and symptoms, radiological and the histological examination of the lesion. All lesions must be biopsied. For cystic lesion of more than 3cm, an incisional biopsy is to be done first, especially if it perforates the cortical bone. ^{25, level III}

5.2. Signs and Symptoms

Ameloblastoma is usually unicentric, slow growing, locally invasive and will infiltrate through the medullary spaces and can erode cortical bone and extend into adjacent tissue, if left untreated. Patients commonly present with painless swelling of jaw and additional symptoms can include malocclusion, pain, tooth mobility, ill-fitting dentures, ulceration, paraesthesia and/or anaesthesia of the affected area. ^{26-27, level III} Siar et al, 2011 reported that painless swelling was the most common complaint (73.3%) among Malaysian. Other symptoms are numbness (5.2%), soft tissue growth (4.4%), discharging sinus (3.0%) and a non-healing extraction socket (0.7%). ^{6, level III} Posterior maxillary tumours can obliterate the maxillary sinus and subsequently extend intracranially. ^{28, level III}



Figure 1: Large mandibular ameloblastoma extending from left anterior region to right angle of the mandible.



Figure 2: Radiographic appearance on OPG.

A multilocular radiolucency was the most commonly encountered radiographic presentation in the series of case study done by Siar et al, 2011, for the Malaysian population (36.8%). Sixteen cases (6.7%) presented with root resorption and 8 (3.4%) were associated with unerupted teeth. ^{6, level III} Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) may be helpful in establishing the extent of the lesion, particularly when located in the maxilla. ^{29, level III}

Computed Tomographic (CT) images, usually show an expansile, radiolucent, multiloculated cystic lesion, with a characteristic "soap bubble-like" appearance. Other CT findings also include cystic areas of low attenuation with scattered isoattenuating regions, representing soft-tissue components. Thinning and expansion of the cortical plate with erosion through the cortex can be seen. ³⁰, ¹⁴, level III



Figure 3: Coronal view of CT scan of the left mandibular ameloblastoma.

5.3. Recurrence Rate

Recurrence can occur up to 21 years after treatment; more than 50% of recurrences are encountered within the first 5 years after treatment. ^{6, level III} There is weak evidence showing that resection resulted in the lowest recurrence rate (3.6%), followed by enucleation with application of Carnoy's solution (16%). Enucleation alone resulted in the highest recurrence rate (30.5%). ^{4, level III} The mainstay of treatment is surgery, with wide resection is recommended due to the high recurrence rate of solid/multicystic ameloblastomas. The recurrence rate after resection is 13–15%, as opposed to 90–100% after curettage. ^{7-9, 8, level III}

6. INVESTIGATIONS

6.1. Radiological Investigation

Conventional radiographs, Computed Tomographs (CT) and Magnetic Resonance Imaging (MRI) have been predominantly used for evaluation of ameloblastomas. The typical features of multilocular or unilocular radiolucency in conventional radiographs are not pathognomonic of ameloblastoma and may also indicate other odontogenic tumours/cysts such as keratocystic odontogenic tumours. 31, level III

Ultrasound can be as a supplementary non-radiation diagnostic method for mandibular ameloblastoma. It can also be used to distinguish cystic from solid contents in the tumour. ^{32, level III} In addition, colour Doppler flow imaging (CDFI) may be used to determine the active proliferation of the tumour. ^{32, level III} However ultrasound and CDFI are not routinely used.

6.2. Radiographic Features

In most cases, ameloblastoma have a characteristic but not diagnostic radiographic appearance. The neoplasm usually appears as a unilocular radiolucent area or a multilocular radiolucent area with a honeycomb appearance. Resorption of the adjacent tooth roots is not uncommon. In many cases of an unerupted tooth, most often a mandibular third molar, is associated with the tumour. ^{27, 33, level III} 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.

6.3. Histological Investigation

The importance of an accurate preoperative histological diagnosis in the treatment of ameloblastoma cannot be over emphasized. The individual histologic patterns have no effect on the clinical behaviour of the tumour. ^{2, 11-12, 4, level III} The tumour has been separated into several subtypes of ameloblastoma, each with its own distinct biological behaviour requiring different forms of treatment. ^{11-12, 4, 2, level III}

It is generally accepted that there is no relationship between the individual histologic patterns and the behaviour of the tumour or its prognosis. Histologic pattern is not to be confused with the designation of solid or multicystic, unicystic or peripheral types, as these descriptors have considerable impact on the patients' treatment and prognosis. ^{26-27, 6, 33, level III}

7. DIAGNOSIS

A differential diagnosis of ameloblastoma is achieved by the clinical features and radiological investigations. Clinically it presents as a painless, slow growing swelling of the jaw with bucco-lingual bony expansion. The mobility of teeth will also be observed if a substantial amount of bone resorption has occurred.

An orthopantomogram will show radiolucency which is unilocular or multilocular with evidence of resorption of the adjacent teeth. Further radiological investigations such as CT scan or MRI can further evaluate the extent of the tumour prior to surgery. 34, level III

A definitive diagnosis of ameloblastoma is made on the basis of histologic features which remains as the most reliable means of diagnosis as compared to clinical radiological features alone. ^{35, level III}

Fine needle aspiration cytology (FNAC) has also been found useful and minimally invasive in achieving a diagnosis of ameloblastoma in some cases.

36-39, level III Nevertheless FNAC is not widely practised.

Recommendation 1

Definitive diagnosis of ameloblastoma is made based on the histologic features with clinical and radiological correlations.

Grade C

8. MANAGEMENT

The options for management of ameloblastomas are;

- enucleation with or without adjunctive procedures
- ii. dredging
- iii. resection

8.1. Enucleation

Enucleation refers to surgically shelling the lesion out of the bone. The aim is to remove the whole cyst/tumour without leaving any visible remnants behind. If remnants were expected to have been left behind, enucleation can be coupled with adjunctive procedures like peripheral ostectomy, application of Carnoy's solution or liquid nitrogen cryotherapy. Recurrence rates with adjunctive procedures are lower compared to enucleation alone. 40, level III Enucleation alone shows higher recurrence rates (30.5%) 8, level III compared to enucleation plus adjunctive procedures (16%), or wide excision, even for the unicystic variety. 40-41, level III

8.2. Enucleation plus adjunctive procedures

The most common adjunctive treatment was the application of Carnoy's solution. This method has a lower recurrence rate (16%) compared to enucleation alone or marsupialization for unicystic ameloblastoma. 42, 2-3 level III

8.3. Dredging

Dredging is a conservative surgical procedure in which following enucleation, repeated dredging/curettage is applied to accelerate new bone formation by removing the scar tissue from the bony cavity. Dredging is carried out at 2-3 month intervals until tumour cells are not seen in scar tissue removed by 2 consecutive dredging. Evidence base for this procedure is small but recurrence rates are comparable to enucleation coupled with adjunctive procedures. ^{41, level III}

Dredging or curettage is a procedure where enucleation of the lesion is followed by repeated curettage of scar tissue from the bony margins to eradicate any residual tumour cells and promote new bone formation. Dredging is applied at 2-3 month intervals and stopped when tumour cells are not present in microscopic examination of the scar tissue removed by two consecutive dredging.

In very large cysts/tumours, marsupialisation maybe carried out first to reduce the size of the lesion, before enucleation or dredging is undertaken. Marsupialisation (decompression) refers to surgically removing the superficial bony wall of the lesion, and suturing the incised edge of the cyst/tumour to the adjacent mucosa. Most lesions do reduce in size and some may completely resolve without further treatment. 43, level III

8.4. Resection

Resection refers to either segmental resections of the mandible or maxilla without maintaining continuity or marginal resection with preservation of bony continuity. This method of treatment will usually require some form of reconstruction by non-vascularised or vascularised grafts and flaps, depending on the site and size of the resultant defect. Surgery should be performed with a 1cm to 1.5cm bony margin free of tumour. ^{44, level III} A more radical approach is necessary for solid or multicystic ameloblastomas. ^{45-46, level III} Regardless of subtype, this method of treatment results in the lowest rate of recurrence in all age groups. ^{40-47, 48, level III}

8.5. Choice of Treatment

The goal of treatment in the management of ameloblastomas should be the eradication of the disease with no recurrence. Ameloblastomas, although locally invasive and aggressive, are essentially benign

in nature, and thus it is advocated to be treated as such, with adoption of a more conservative approach to management. ^{49-51, level III} However more recent evidence indicates such conservative methods like enucleation appear to result in high recurrence rates. ^{43; 25, level III} Several factors need to be considered when deciding on the method of treatment for ameloblastomas. They include:

- Subtypes
- ii. Site
- iii. Age of patient

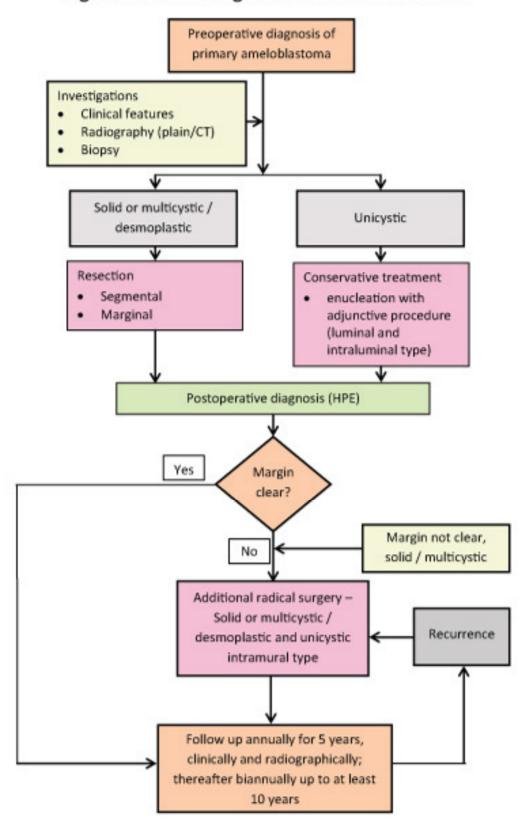
8.6. Subtype

Ameloblastomas are classified into 3 subtypes: solid and multicystic, unicystic and peripheral (which are not included in this review). In general, ameloblastomas are slow growing, locally invasive benign tumours. However the solid and multicystic variant tends to be locally aggressive with a high recurrence potential if inadequately excised. The unicystic ameloblastoma is thought to have recurrence potential, but is less aggressive and may respond to enucleation and/or curettage. 52, level III

8.7. Site

In general, although histologically identical, maxillary lesions can be more troublesome than mandibular lesions. The maxilla lacks the thick confining cortical plates of the mandible, thereby allowing spread of the tumour to surrounding vital structures. ^{44, level III} Spread to the posterior wall of the maxillary sinus, pterygomaxillary space, greater palatine canal and base of skull have been described. ^{53-54, level III} Adjunctive procedures like Carnoy's solution and liquid nitrogen are more difficult to apply to the maxilla as they cannot normally be applied in the sinus or nasal cavity, which is usually involved.

Algorithm for management of Ameloblastoma



8.9. Management in Children

Ameloblastomas in children and adolescents are thought to be rare. They account for approximately 10-15% of all reported cases of ameloblastoma. 55-56, level III As the tumour grows slowly, some authors have proposed the theory that ameloblastoma probably starts to develop in childhood. 57, level III The unicystic ameloblastoma is the most commonly reported type of ameloblastoma in children. 58, level III

The treatment of ameloblastoma is controversial and presents some special problems in children. Incomplete growth of jaws and the prognosis of the tumour in children make the surgical consideration different from adults as it can cause irreversible deformities and poor aesthetic results in light of postoperative growth and development of young patients.

Consequently, some advocate that ameloblastoma is treated conservatively with decompression, enucleation and peripheral ostectomy as well as periodic long-term follow up. ^{57-59, level III} The basis for this conservative approach was suggested as the majority of ameloblastoma in children are unicystic which tend to have much lower rates of recurrence (10-25%) than the multicystic or solid types. ^{7, level III}

Whilst the conservative surgical approach results in a good appearance and functional recovery, a more aggressive surgical approach such as marginal or segmental resection may be considered when the condition recurs or the ameloblastoma is of multicystic or solid types. ^{58, level III}

Recommendation 3

Decompression followed by enucleation can be effective in controlling progression of disease until maturity allows for complete excision or even curing the disease. Good compliance for regular follow-up is important for successful treatment when the conservative approach is chosen.

Grade C

9. RECONSTRUCTION

Reconstruction is the procedure to re-establish anatomy and function of the facial structures. It is needed for aesthetic and function of the jaws after resection of maxilla or mandible. In current practice, there are several means of reconstruction.

9.1. Reconstruction of resected maxilla

Reconstruction of maxillary defects could be carried out by several options namely maxillary prostheses, local pedicle flaps, soft tissue free flaps and vascularized bone flaps. 60-62, level III Prostheses could be used either alone or in addition to surgical flaps, depending on the defect. 63-64, level III

The prosthesis restores the oronasal separation, which is fundamental for speaking and swallowing. Dentition can be included for cosmesis and chewing. The surgical complexity and length of procedure is less with obturators than with tissue reconstruction.



Figure 4: Maxillary reconstruction using full prosthesis

9.2. Reconstruction of Resected Mandible

Loss of mandible continuity results in alteration in speech, swallowing and mastication, and in the appearance of the patient. 65, level III

9.3. Reconstruction With Vascularized Bone Graft

With the advent of microsurgery, many reconstructive surgeons have adopted vascularised bone grafts such as rib, metatarsal, iliac, radial, scapular and fibular. Fibula became a popular graft because of a straight forward dissection which facilitates a two-team approach. Furthermore, this flap provides enough long as well as strong bicortical bone of excellent quality which can endure physical stress, i.e. in terms of mastication, necessary for the reconstructed mandible after placement of osseointegrated implants. ^{66-68, level III} Today, the trend is toward early reconstruction. Full dental reconstruction is impossible without osseointegrated implants because conventional dentures are difficult to adapt to the transplanted bone. ^{69, level III}

9.4. Reconstruction With Non-vascularized Bone Graft

A variety of methods has been proposed for mandibular reconstruction using non-vascularised bone grafts or alloplastic implants.

Non-vascularized iliac crest bone grafts for segmental reconstruction of the mandible is the method of choice on the condition that the defect is at the posterior (ramus-body) region. ^{70, level III}



Figure 6: Mandibular reconstruction using non-vascularised bone graft from iliac bone.

9.5. Reconstruction With Reconstruction Plate Only

The reconstruction plate provides a predictable, safe and efficient means for the surgeon to maintain mandibular continuity. ^{63, level III} Although microsurgical bone transplant is still the gold standard, mandibular reconstruction plates (MRP) were for several years considered a fast and safe way to restore mandibular continuity without using bone graft. New plate designs with locking screws have been recently introduced, and these may overcome some of the complications related to previous designs. ^{64-71, level III}

Durable mandibular reconstruction with plates offers the possibility of reconstituting the mandible in patients with a poor prognosis or medically compromised which are not fit for a long surgery. Reconstruction plate is used when predicted life expectancy is low and when medical conditions preclude prolonged general anaesthesia. 72-73, level III

Key massage

The choice of the reconstruction should be discussed between the patient and the surgical team.

10. FOLLOW UP

Long term follow-up is important. ^{66, 69-70, 64, 72-73, level III} The first five years is critical. ^{62-66, 69-70, 64, 73, level III} Thereafter biannual follow-up for at least 10 years is recommended. ^{74, level III} Radiographs are to be taken during followup. Recurrence may occur even after 20 years. ^{7-8, level III}

11. PROGNOSIS

Prognosis is determined by site, spread, type and method of surgical treatment. 66-70, 64, 72-73, level III

11.1. Site

Maxillary ameloblastomas have poorer prognosis and therefore radical surgery is highly recommended. 75, 10, level III

11.2. Spread

Some studies have shown correlation with the extent of the initial tumour, multiple recurrences and surgical interventions to the potential for metastatic spread. ^{76, level III}

11.3. Type and Method of Surgical Treatment

Essentially most studies showed that the prognosis is more dependent on the method of surgical treatment rather than the histologic type of tumour. Solid ameloblastomas with multilocular radiographic image presented a significantly higher incidence of recurrence. The level III A ruptured mandibular basal cortical bone indicated a three times higher risk of recurrence compared with cases of preserved or expanded cortical bone the lowest incidence of recurrence. The level III Treatment by resection/radical surgery showed the lowest incidence of recurrence. The level III Many authors recommend a margin of 1.5 - 2 cm beyond the radiological limit to ensure all microcysts are removed. The level III microcysts are removed.

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