CASE REPORT

Management of large mandibular ameloblastoma – a case report and literature reviews

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Abstract
Ameloblastoma is a slow growing benign tumour of the jaw and patients usually present late after the tumour achieved considerable size to cause facial disfigurement. Diagnosis mainly from tissue biopsy and characteristic findings on plain X-rays does assist in differentiating between types of ameloblastoma. The challenges in the management of this tumour are to provide complete excision as recurrence may occur in incomplete removal and also to reconstruct the bony defect in order to give reasonable cosmetic and functional outcome to the patient.

Introduction
Benign mandibular swellings can be due to a wide variety of lesions and can be divided into odontogenic and nonodontogenic origin. Among these are ameloblastoma, radicular cyst, dentigerous cyst, keratocystic odontogenic tumour, central giant cell granuloma, fibro-osseous lesions and osteomas. Ameloblastoma is the commonest benign tumour of odontogenic origin which developed from epithelial cellular elements and dental tissues in their various phases of development. It is generally a slow-growing but locally invasive tumour. Its peak incidence is in the 3rd to 4th decades of life and the male to female ratio is 1:1. It is often associated with an unerupted third molar (Gerzenshtein et al., 2006). It may present as a result of routine radiographic examination finding. Eighty percent of ameloblastomas occur in the mandible and majority is found in the angle and ramus region. There are three forms of ameloblastomas, namely peripheral, unicystic, and multicystic tumors. Multicystic ameloblastoma was commonly seen among all and represent 86% of cases. Peripheral tumors are odontogenic tumors, with the histological characteristics of intraosseous ameloblastoma that occur solely in the soft tissues covering the tooth-bearing parts of the jaws. Unicystic tumors include those that have been variously referred to as mural ameloblastomas, luminal ameloblastomas, and ameloblastomas arising in dentigerous cysts (Chana et al., 2004). The challenge in managing ameloblastoma is in achieving complete excision and reconstruction of the defect when the tumour is large. We present a case of a large mandibular ameloblastoma and review of the literatures on the choices of reconstruction.

Case report
An 18-year-old Malay girl (Figure 1) presented with a history of a painless left mandibular swelling of two years duration which was progressively increasing in size. The swelling was associated with paraesthesia of her left side of face mainly over the jaw and she also had slight difficulty in chewing due to the effect of the mass over the left mandible. Apart from minimal...
malocclusion noted, there were no problem related with articulation as well as deglutition.

On physical examination, she was medium build size and clinically appeared pink. There was a large firm swelling over the left mandibular region which was non tender extending from the left angle of mouth anteriorly to the anterior border of sternocleidomastoid muscle posteriorly and causing left facial asymmetry. The skin overlying the mass was normal in color without any dilated superficial veins. There was no enlarged lymph node over the neck region. There was no trismus and intraorally, she had good dental hygiene and all the present dentition were vital. However, the floor of mouth was raised on the left side but with intact and smooth buccal mucosa. Panoramic X-ray revealed a cystic lesion located in the left side of mandible extending from angle to ramus and was associated with unerupted third molar (Figure 2).

Incisional biopsy of the oral cavity mass was done under local anaesthesia with minimal bleeding and it was reported as pseudoepitheliomatous hyperplasia. CT scan of the neck suggestive of a benign lesion or low grade malignant mass with differential diagnosis of ameloblastoma, aneurysmal bone cyst and lymphoma (Figure 3). Though the biopsy result did not showed specific features of ameloblastoma, the clinical history and physical examination along with radiological findings were highly suggestive of ameloblastoma. She was then planned for segmental mandibulectomy via lip splitting incision and then to stabilise the mandible with titanium plate. Intraoperatively, after temporary tracheostomy was performed, the skin flap was raised and retracted laterally (inferior flap) and superiorly (superior flap) exposing the mandible underneath. The solid tumour was then noted to involve the whole of left mandible measuring about 8 x 6 cm which include the body, ramus and head of condyle. In view of this extensive tumour involvement, the surgical plan was changed to hemimandibulectomy whereby the mandible was split at midline and left hemimandible was removed en bloc with the tumour.

Therefore the initial plan to bridge the bony gap created with titanium plate was not possible as there was no bony remnant left on the left side and the wound was closed primarily without reconstruction (Figure 4). Post operative period was uneventful and tracheostomy tube was decannulated on day five and suture were removed on day seven post operative. She was on Ryle’s tube feeding till day 12 and thereafter started on soft diet after the tube was removed. She was also referred to occupational therapist for chewing exercise. She was discharged home on day 15 post operatively. Post operative histopathological result revealed sheets of odontogenic epithelium with basal layer of cuboidal cells and suprabasal stellate reticulum like cells exhibiting cystic degeneration (Figure 5). Intervening area of loose stroma with minimal...
infiltration of inflammatory cells and haemorrhagic area were also seen. The histopathology was interpreted as ameloblastoma and some of these histologic features are in accordance with the criteria for a diagnosis of unicystic ameloblastoma (Li et al., 2000).

At her first follow up one month later, she had no significant complaint except for slight tightness of her left cheek. There was small depression over her left cheek but it was not apparent when she had her headscarf on. She defaulted some of the subsequent follow up due to inconvenience that she had as she was enrolled to continue her study at a local university. She managed to turn up about two years later and there was no recurrence noted during clinical examination. She had no difficulties in swallowing, chewing and maintain good voice quality. We advised for reconstruction of the cheek defect but she refuses and was happy with her present status.

Discussion

Ameloblastoma in the mandible can progress to great size and cause facial asymmetry, displacement of teeth, loose teeth, malocclusion, and pathologic fractures. Tumor size may ranges from 1 to 16 cm at presentation which result from expansion of bone and invasion into soft tissue. Typically ameloblastoma present as painless slow growing mass and in this case it took about two years before the patient developed symptoms such as significant facial asymmetry, malocclusion and difficulty in chewing. This patient also had paraesthesia over the left cheek particularly over the distribution of mandibular division of trigeminal nerve. Other clinical presentations of this disease were pain or anesthesia of the affected area. Becelli et al. (2002) studied 60 patients whom were confirmed with mandibular ameloblastoma and found out that about half of them showed typical symptoms such as swelling of the affected region (38.3%), paraesthesia of the innervated region of the mandibular nerve (13.3%) and alteration in dental occlusion in 10% of the cases. Radiographically, ameloblastoma appear as radiolucent lesion that may have either a unilocular or multilocular appearance. It may expand the cortical plate which gives rise to a paper-thin and soap bubble appearance on panoramic X-ray as well as CT scan (Fig. 2). Bilkay et al. (2004) in retrospective analysis of 100 patients with benign mandibular lesion has found that 78% of the cases had a radiolucent lesion and 83% of this had cysts with well-defined borders.

Ameloblastoma was known for its high recurrence rate if excision was incomplete. Therefore the treatment of choice is surgical excision with wide free margins. The traditional approach for a mandibulectomy is through a lip-splitting incision and though it has the disadvantage of post operative morbidity; it gives a better exposure for complete tumour removal. Some authors such as Derderian et al. (2004) use a less invasive incision which avoid troublesome outcome of the lip-splitting. They utilize a ridson incision and this was combined with intra-oral incision which gives a less post operative morbidity and more cosmetic outcome. Shirani et al. (2007) in a series of 7 patients introduced a new technique of removal of large ameloblastoma with immediate reconstruction by using only an intra-oral incision. It has the advantages of removing and repositioning of the mandible intra-orally and therefore allows removal of the lesion and reconstruction procedure to be done simultaneously. Facial scar and damage to the marginal mandibular nerve that innervate the lips can also be avoided via this technique.

Compared to its multicystic counterpart, the unicystic ameloblastoma tend to be less aggressive and has lower recurrence rate. Even though some authors advocate a more conservative approach such as enucleation and curettage of this tumour, a large lesion with erosion of the mandibular cortex certainly requires a more aggressive approach for complete removal of the tumour. Eppley (2002) in his review of 60 mandibular ameloblastoma cases have shown that there was no recurrence of those cases treated via en bloc resection as compared to enucleation and curettage in which the recurrence rate was as high as 25% to 50%.

Reconstruction of large mandibular defects represents a challenge to head and neck reconstructive surgeons. The mandible is both functionally and cosmetically important structure of the head and neck, contributing to facial appearance, chewing, speech and swallowing. In this case, we experienced difficulties in reconstructing the defect as we do not have plastic reconstructive unit in our centre to do the reconstruction aspect and furthermore the parent refused referral to other centers due to transportation and logistic problems. There are different methods of mandibular reconstruction for large defect that have been described in literatures and among all, microvascular surgery has become the preferred option. Four donor sites i.e, fibula, iliac crest, radial forearm, and scapula have become the primary sources of vascularized bone and soft tissue for the oral reconstruction. Among all these, fibula has multiple advantages including bone length and thickness, donor site location permitting flap harvest simultaneously with tumor resection because both teams are at different end of the table, and minimal donor site morbidity and therefore should be considered as a choice in reconstruction (Disa and Cordeiro, 2000). Yilmaz et al. (2008) did a comparison between vascularised iliac crest flap (24 cases) and vascularised free fibular flap (13 cases) and noticed that less complication rate and superior functional and aesthetic results were achieved for those with fibular flap. Ghana et al. (2004) in their series of 10 cases utilized vascularised...
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fibula flap with simultaneous placement of osseointegrated dental implants and claimed it is the ideal treatment for large ameloblastoma. Becelli et al. (2002) elucidate two phases in the reconstruction process which are first, the phase of reconstruction of the surgical defect with free or autogenous bone graft or revascularized autogenous bone graft and the subsequent phase which is carried out to obtain prosthetic restoration by means of endosseous implants.

Another method of reconstruction is internal distraction osteogenesis as has been popularized by McCarthy et al. (1992). Among the large series advocate of this technique were González-Garcia et al. (2008) who did 10 cases. They achieved successful distraction in eight patients with one patient failed and the other did not complete due to tumour recurrence. With the advancement of biomaterial engineering, researchers are now looking at other method of reconstruction and one of the latest technique was using bioimplant containing BMP-7 as described by Clokie and Sándor (2008). Ten patients with major mandibular defects following resection of biopsy-proven ameloblastoma lesions or osteomyelitis of the mandibular body or ramus were included in this study. The resection defects were spanned with rigid reconstruction plates to hold the remaining mandibular segments in the correct position. The defects were filled with a bioimplant containing bone morphogenetic protein-7 (BMP-7) in a demineralized bone matrix (DBM) suspended in a reverse-phase medium to effect sustained BMP delivery. Radiographic evidence of mandibular bone formation was found in all cases and at the end of 1 year, functional and esthetic reconstruction of the mandible was complete.

In conclusion, en-bloc tumour resection reduces the chance of tumour recurrence but resulted in large mutilating bony and soft tissue defects as indicated by our experience and also in many other series. The challenge in the management of large ameloblastoma of the mandible is not only to excise the tumour completely in order to prevent recurrence but also to provide the best reconstruction method.

References


