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Case Report

Case report of a massive giant ameloblastoma and its management

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ABSTRACT

Ameloblastoma is a benign odontogenic tumor. It is slow growing, locally aggressive, rare tumor, accounting for 1% of all the jaw tumors. Here, we report a case of a Giant ameloblastoma in a 38-year-old gentleman who presented with a massive tumor of the mandible, possibly the largest reported in literature. The tumor had caused significant facial disfigurement and functional impairment leading to nutritional deficiency, anemia and dehydration induced elevated renal parameters. It was neglected by the patient for over ten years, due to the fear of surgery and financial constraints. Pre-operative evaluation included 3-Dimensional computerized tomographic imaging of the head and neck and optimization of deranged hematological & renal parameters and high blood pressure. Surgical treatment entailed complete tumor resection with adequate margins and microvascular reconstruction with fibular free flap in a single stage. This case of a giant multicystic ameloblastoma of the mandible, possibly the largest of its kind, was managed by complete surgical resection and a simultaneous single stage reconstruction with complete financial aid provided by our institution for the entire treatment. Finally, providing the patient, an acceptable functional, aesthetic, and oncological outcome.

Keywords: Giant Ameloblastoma, Largest reported, Wide resection, Single stage osteocutaneous reconstruction

INTRODUCTION

Ameloblastoma is a benign odontogenic tumor.[1] It is a slow growing, locally aggressive tumor, accounting for 1% of all jaw tumors.[2] It is primarily seen in adults in their 3rd to 5th decades of life with equal sex predilection and can grow to varying sizes causing significant facial disfigurement.[3] Giant ameloblastoma is rarely reported in the present times. The largest reported, by Michael et al in 2010 was a 24×19×15 cm recurrent giant, multicystic maxillo-mandibular ameloblastoma.[4] Here, we report a case of a giant ameloblastoma, possibly the largest reported in literature, neglected by the patient for over ten years, due to fear of surgery and its outcomes, and financial constraints. Being a not-for-profit tertiary care centre, we were not only able to give him the best possible surgical treatment i.e., complete tumor resection with reconstruction, but also were able to provide complete financial aid for the entire treatment process.

CASE REPORT

A 38-year-old, thin-built gentleman was brought by our hospital's community outreach unit with a gradually progressive, painless swelling of the lower jaw for ten years. As it increased to the size of a football, he sought medical help, however, in view of the enormous size and other medical issues, surgery was deferred at various other centers. Social stigma and financial constraints had gradually pushed him into clinical depression. Before coming to us, he had also taken native medicinal treatment for a short duration.

Figure 1: Clinical picture of the patient showing large 25×22 cm swelling of the mandible with few residual teeth in the lower jaw and fair mouth opening.

Clinical examination revealed a large, non-tender swelling with variable consistency measuring 25×22 cm involving the entire body of the mandible, up to the angles bilaterally, with overtly stretched lower lip and overlying skin. Intra-orally, floor of the mouth and lower gingivobuccal sulcus too were significantly stretched with loss of multiple teeth from the lower jaw, with few displaced residual teeth still attached to the distorted mandible with no obvious ulcer/lesions in the oral cavity. He had a fair mouth opening, with no restriction of tongue movements and was able to consume a soft diet (Figure 1).

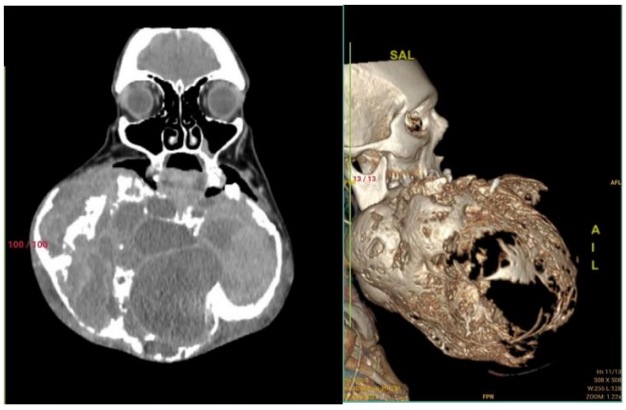

Figure 2: Plain and 3-Dimenstional CT showing huge expansile lytic lesion of size 25×22×15 cm involving the entire body of the mandible, till the angles bilaterally with internal multilocular cystic and few solid enhancing components.

Evaluation with a three-dimensional CT of head and neck (Figure 2), revealed a huge expansile lytic lesion of size 25×22×15 cm involving the entire body of the mandible, with overtly stretched floor of mouth muscles, with loss of fat planes. The expanded mandibular cortex was partly thickened and partly invisible and had internal multilocular cystic and few solid enhancing components; features suggestive of a giant Ameloblastoma of the mandible.

Work-up revealed low hemoglobin, thrombocytosis, hypertension, and a raised creatinine level. On further evaluation, in consultation with a hematologist, the anemia was found to be nutritional, for which he received blood transfusion prior to the surgery. No obvious cause for the thrombocytosis was identified. A physician's consult was sought, and he was started on an antihypertensive. The raised serum creatinine was thought to be pre-renal or secondary to the native medicines taken by the patient, however, it was corrected with adequate fluid resuscitation.

A multi-disciplinary team meeting, involving the surgical oncologist, plastic and reconstructive surgeons, radiation oncologist and radiologist was held. A preoperative biopsy was deferred in view of the extensive lytic nature of the tumor and very few significant solid areas that could be targeted for the biopsy. As the entire body of mandible was involved and the CT imagining could not delineate the floor of mouth muscles clearly, prophylactic tracheostomy for adequate airway protection was planned.

After ensuring adequate preoperative optimization, he was taken up for surgery i.e., wide local excision of the mandibular tumor and, single stage reconstruction with free fibula osteocutaneous flap. Due to the enormous size of the tumor, mask ventilation and laryngoscopy was difficult, hence an awake fiber optic nasal intubation was done under an airway block. Simultaneous mandibular resection and fibula free flap harvest were done by two teams. The gigantic size of the tumor had led to an altered local anatomy along with large, dilated vessels, which made resection of the tumor a challenging task. The entire body of the mandible involved by the tumor was resected, ensuring an adequate margin on either side.

The harvest of fibula osteocutaneous flap was performed in the standard manner based on peroneal vessels from the left lower limb, i.e, 6 cm of fibula bone was preserved proximally to safeguard the Common peroneal nerve and 6 cm of bone was preserved distally to maintain ankle stability. Around 24 cm of segment of bone was safely harvested to obtain adequate pedicle length. Once harvested, multiple osteotomies were performed as per requirement i.e., 2 cm of bone for reconstruction of each ramus and 7 cm for the central body of mandible and plating was done (Figure 3). Refashioned fibula was then anchored to bilateral rami of the mandible using titanium plates and screws while maintaining an adequate maxillo-

mandibular relationship and the skin paddle was used to drape the lingual surface of the bone (Figure 4). Microvascular anastomosis was performed between the peroneal artery and the facial artery, and the two venae comitantes were anastamosed with the Internal Jugular vein. Excess local soft tissue was excised and plantaris tendon was used as a static sling to maintain oral competence. Covering tracheostomy was done for airway protection, in view of extensive mandibular and floor of mouth resection, at the end of the surgery.

Figure 3: Harvested free fibula osteo-cutaneous flap fashioned into shape of mandible and pre-plated for fixation.

Figure 4: Fixation of the osteocutaneous flap to the native residual mandible bone, maintaining the maxillo-mandibular relationship for good occlusion and cosmesis.

The patient was monitored in the intensive care unit in the immediate postoperative period which was uneventful. He received nasogastric tube feeds in the immediate postoperative period and was then started on oral liquid feeds on postoperative day 10 which he tolerated well. Tracheostomy was decannulated once the surgical site edema resolved and after ensuring adequate tongue movements and swallowing reflexes. He was discharged from the hospital on postoperative day 13.

Figure 5: Excised specimen weighing 5.44 kgs.

Figure 6: Gross specimen in cross section showing multi-cystic lesion with focal solid areas.

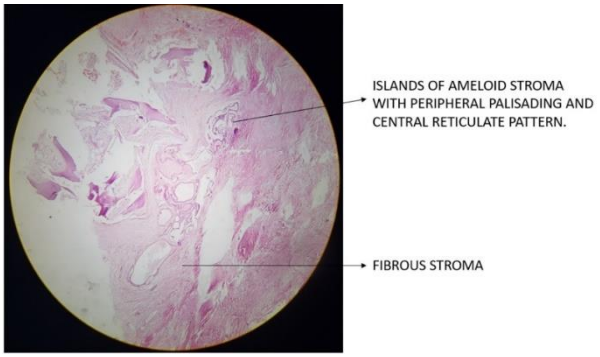

Figure 7: Histopathlogical microphotograph depicting the characteristic findings in a conventional multicystic ameloblastoma.

Final histopathology reported a gross tumor weighing 5.4 kg, (Figure 5) measuring 25×20×16 cm, cut section showing a multicystic lesion with expanded mandibular bone with focal solid areas (Figure 6) and histology confirmed a conventional multicystic ameloblastoma

(Figure 7) with reactive regional nodes with 2.5 cm margin.

At four months follow-up, good oral competence and restored mandibular shape were observed. There is no sign of recurrence.

As a tertiary care, not-for-profit centre, we were able to help the patient in a holistic manner, both medically and financially. The whole cost of hospitalization including the cost of implants was borne by the institute with the help of donors. With additional financial aid, we hope to aid in dental rehabilitation with osseo-integrated implants or dental prostheses in the future.

DISCUSSION

Ameloblastoma is a rare epithelial odontogenic tumor that occurs in the mandible and maxilla.[3] The exact etiology of ameloblastoma is still unknown, multiple cellular pathways are said to be involved.[6] No predisposing factors have been identified so far, as per the literature review. They are classified based on histology. As per the classification by The World Health Organization (WHO), there are 4 subtypes: 1) solid/ multicystic/ desmoplastic; 2) unicystic; 3) extraosseous/ peripheral; and 4) metastasizing ameloblastoma.[7] Among them, the most common is the solid/ multicystic ameloblastoma (75-86 %), and our case falls into this subtype.[8] Histologically they can be follicular or plexiform types.[8] Mandibular ramus is the most common site, and it is a slow growing tumor. Ameloblastoma are benign tumors but locally invasive, thereby have a high recurrence rate if not treated appropriately.[9] Rarely, it grows to gigantic sizes. However, there is no size definition for a giant ameloblastoma.

The diagnostic work usually includes imaging– Computerised Tomography (CT), MRI/ OPG, and biopsy in selected cases. On imaging, they appear as multilocular or unilocular lesions. The multiloculated tumors have a honeycomb or a soap bubble appearance.[10] The CT scan of our patient revealed a large, multilocular lytic tumor, involving the entire body of the mandible, with loss of teeth in the entire segment.

There are many treatment modalities for ameloblastoma, which include conservative treatment like enucleation, curettage, cryotherapy, radiotherapy, marginal resection and radical treatment, complete surgical resection- the most ideal treatment. Size of the tumor, location, type, age, and general condition of the patient all play a role in determining the right treatment modality. Preoperative biopsy is recommended whenever possible, especially if conservative treatment is being planned to confirm the histological type and risk of recurrence. Review of literature reveals a high recurrence rate following conservative treatment (29.3-93%), than radical treatment (0-21%).[11] Unicystic ameloblastoma and extra osseous/ peripheral tumors are suitable for conservative treatment. Solid/ multicystic and desmoplastic tumors warrant a complete surgical resection with adequate margins of resection. A minimum of 1-1.5 cm is the margin required for resection of these tumors, to decrease the risk of recurrence.[12]

We anticipated the following problems during the surgery. Difficult airway and the risk of shared airway between the surgical and anaesthesia teams which were managed with awake fibre-optic intubation followed by prophylactic tracheostomy. Blood loss, for which we ensured adequate blood and blood product availability prior to the surgery. Lack of clear plane between floor of mouth muscles and extremely stretched soft-tissue cover.

Figure 8: 3 month post operative picture of the patient. He has an acceptable functional, aesthetic, and satisfactory oncological outcome.

Our patient underwent complete resection of the involved body of mandible, along with the adjacent thinned out soft tissue, leaving behind both rami, to which the neo-mandible was anchored with plates and screws. The fibula bone along with the pedicle was harvested from the left leg under tourniquet control and was refashioned by making multiple osteotomies, to create a neo-mandible. As per the final histopathology report, we had achieved a 2cm margin on either side of the tumor. Post operative recovery was uneventful and aesthetic outcome was satisfactory with good oral competence and mouth opening at 3 months follow-up (Figure 8).

CONCLUSION

This case of a giant multicystic ameloblastoma of the mandible, possibly the largest of its kind, was managed by complete surgical resection and a simultaneous single stage reconstruction with complete financial aid provided

by our institution for the entire treatment. Finally, providing the patient, an acceptable functional, aesthetic, and oncological outcome.

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