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Ameloblastoma of the mandible

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Abstract

Ameloblastoma is a locally aggressive, slow-growing, benign neoplasm that arises primarily, but not exclusively in the mandible and maxilla. While the exact cell of origin of this tumor remains controversial, its tumorigenesis is thought to represent an aberrant form of embryogenesis whereby cells of the tooth germ undergo formation of a tumor. This neoplasm is therefore classified as an odontogenic tumor. Regardless of its size, this tumor is capable of significant destruction of the jaws and infiltration of the surrounding soft tissues of the head and neck region. Due to the relatively painless nature of these tumors, persistent growth with clinically apparent facial deformity will occur when patients do not seek consultation and appropriate surgical treatment in a timely fashion. The result may also include clinically significant functional impairment. This notwithstanding, large ameloblastomas may be surgically removed while providing functional and cosmetic reconstructions thereby preserving quality of life in such patients. This case report reviews the workup and surgical ablation of a large ameloblastoma of the mandible in a patient who first became aware of its development twenty years earlier.

Keywords

solid/multicystic ameloblastoma; facial deformity; functional impairment; selective embolization of tumor vasculature

Introduction

Odontogenic tumors represent a biologically, radiographically, and clinically diverse group of benign and malignant tumors that arise in the jaws and associated soft tissues. These tumors are uncommon and account for only 1-4% of all specimens processed and diagnosed in oral and maxillofacial pathology laboratories worldwide [1]. Large series of odontogenic tumors reveal that approximately 97-99% of all odontogenic tumors are benign [1]. While almost uniformly slow-growing, these tumors are also highly destructive neoplasms. In addition, the surgical removal of locally aggressive benign odontogenic tumors with appropriate linear and anatomic barrier margins, as indicated, leads to definitive, curative surgical management of these neoplasms provided that negative surgical margins are realized.

The ameloblastoma is a benign, locally aggressive odontogenic tumor that has received significant attention in the international literature, likely due to its relatively common incidence in terms of the collective assessment of odontogenic tumors. In most tertiary care centers, the ameloblastoma is encountered as the commonest odontogenic tumor. The ameloblastoma is most commonly diagnosed in the late third and early fourth decades with men and women being equally affected [2]. A painless

swelling and slow interval growth are typically reported by patients. The mandible is affected four times more commonly than the maxilla. The ameloblastoma has been classified according to three anatomic variants including the solid/multicystic, unicystic and peripheral variants. The solid/multicystic ameloblastoma is not only the most common variant of this tumor, but also the most frequently discussed in terms of acceptable treatment [3, 4]. It is the variant of the ameloblastoma that typically presents as a multilocular radiolucency of the jaws as noted on plain film and sophisticated imaging studies such as CT scans. The unicystic ameloblastoma commonly presents as a unilocular radiolucency of the jaws and most commonly in young patients. Additionally, this variant of the ameloblastoma has three subtypes including the luminal, intraluminal, and mural subtypes. Finally, the peripheral ameloblastoma is rarely encountered clinically, and presents as a soft tissue mass adjacent to the jaws. Large case reviews indicate that approximately 92% of all ameloblastomas are solid/multicystic, while 6% and 2% are unicystic and peripheral, respectively [2].

Surgery is the standard approach to the management of the solid/multicystic ameloblastoma. Historically, the extent of surgical removal was a subject of controversy with conservative enucleation and curettage of the solid/multicystic ameloblastoma being recommended by some surgeons [5, 6, 7] while segmental resection with the inclusion of linear and anatomic barrier margins being recommended by others [8, 9]. In the final analysis, definitive surgical removal of the tumor with the realization of negative surgical margins offers the patient a predictably high likelihood of cure of their neoplasm. In fact, any surgical approach short of resection is likely to result in the identification of persistent tumor at unpredictable times postoperatively [9].

Case Presentation

A 41 year old man (figure 1a-d) was referred to the author's clinic with a twenty year history of a progressively enlarging mass of the right face. Oral examination (figure 1c) supported the origin of the mass to be from the mandible that was confirmed by panoramic radiography (figure 1d). The patient had previously undergone transoral incisional biopsy of the mandibular tumor that did not secure the histopathologic diagnosis. An ameloblastoma was suspected due to the location and chronicity of the mass as well as its multilocular radiolucent character. The patient became briefly lost to follow-up and represented to the author approximately two years later (figure 2). A repeat panoramic radiograph (figure 2c) was obtained at that time that demonstrated progressive interval growth of the mandibular tumor. Additional plain films of the mandible (figure 3a, b) obtained at that time permitted additional radiographic assessment of the tumor. The patient was subjected to computed tomograms (CT) of the maxillofacial region (figure 4a-c) that supported the development of three-dimensional reconstructions (figure 4d). A repeat incisional biopsy of the mass was performed transorally that identified solid/multicystic ameloblastoma. Due to the large size of the tumor, a decision was made to perform a diagnostic angiogram (figure 5a) and selective embolization of surrounding vasculature so as to reduce blood loss at the time of surgery. The right internal maxillary artery was embolized with three coils. The right facial and lingual arteries were similarly embolized with multiple coils. A post-embolization angiogram identified effective diminution of peri-tumor vasculature (figure 5b). The patient was prepared for the operating room within 24 hours so as to minimize the development of collateral blood flow to the tumor.

Surgical Procedure

The patient underwent a nasoendotracheal intubation in a fiberoptic awake fashion without difficulty, thereby obviating the need for a tracheotomy. A skin incision was designed inferior to the palpable subcutaneous tumor in the right neck while sacrificing a small skin paddle that was adherent to the tumor (figure 6). The soft tissues of the right neck were widely dissected and the platysma muscle was transected and reflected from the tumor bed as this muscle was not invaded by the tumor. The dissection about the tumor bed was performed while identifying and maintaining the pseudocapsule on the tumor (figure 7). A disarticulation segmental resection was performed that observed a 1 cm linear margin in bone in the left mandible and without tumor spillage (figure 8a, b). A specimen radiograph was obtained to provide additional information regarding the radiographic character of the tumor (figure 9). Hemostasis was obtained with electrocautery in the residual tissue bed (figure 10), and stabilization of the mandibular defect was performed with a reconstruction bone plate with an affixed condyle and with four bicortical screws in the remaining contralateral mandibular segment (figure 11). Soft tissue reconstruction of oral lining was performed with an anterolateral thigh free flap. No complications were encountered during the surgery and the patient was extubated on the first postoperative day.

Pathologic analysis of the specimen

Examination of the gross bivalved specimen (figure 12) demonstrated areas of solid tumor and numerous cystic spaces, consistent with the solid/multicystic ameloblastoma. Final microscopic sections resulted in a diagnosis of solid/multicystic ameloblastoma with negative soft and hard tissue margins (figure 13a, b, c). No signs of malignancy were identified in the specimen.

Discussion

The solid/multicystic ameloblastoma of the jaws may develop to a large size for a variety of reasons including the patient's fear of surgery or the lack of access to care for surgical removal. Unimpeded growth of the tumor will therefore occur under these circumstances. The molecular events of tumor growth in the cell cycle permit the identification and designation of tumor doubling times that are required for the doubling of tumor size [10,11]. A two year doubling time was appreciated in the patient discussed in this case report based on an evaluation of his clinical and radiographic presentations in two distinct time points separated by two years. As was noted in this patient, large ameloblastomas may represent intimidating clinical and radiographic presentations. As such, some authors have speculated as to the role of radiation therapy in the management of the solid/multicystic ameloblastoma [12]. In fact, Atkinson et al reviewed 10 patients where radiation therapy was administered in the treatment of ameloblastomas and reported that the ameloblastoma is radioresponsive [12]. This notwithstanding, surgical therapy represents the standard of care of the management of ameloblastoma, including those tumors that are large. As performed in the management of the present case, the execution of preoperative embolization of vasculature surrounding the tumor and a methodical approach to dissection of the anatomic barriers surrounding the tumor resulted in minimal intraoperative blood loss and tumor free margins. Despite the large size of the tumor, this patient was able to be reconstructed in a fashion consistent with the preservation of quality of life and function. The use of the reconstruction bone plate permitted support of the patient's facial form and the ability to maintain his remaining dentition in an anatomic and functional occlusion. The ability to secure the suprahyoid musculature to the

reconstruction bone plate permitted predictable support of the patient's tongue and maintenance of his posterior airway such that tracheotomy was not required. It is not unreasonable to apply such principles to similar cases and to expect similar results. Finally, the presence of this tumor for twenty years did not result in its malignant transformation.

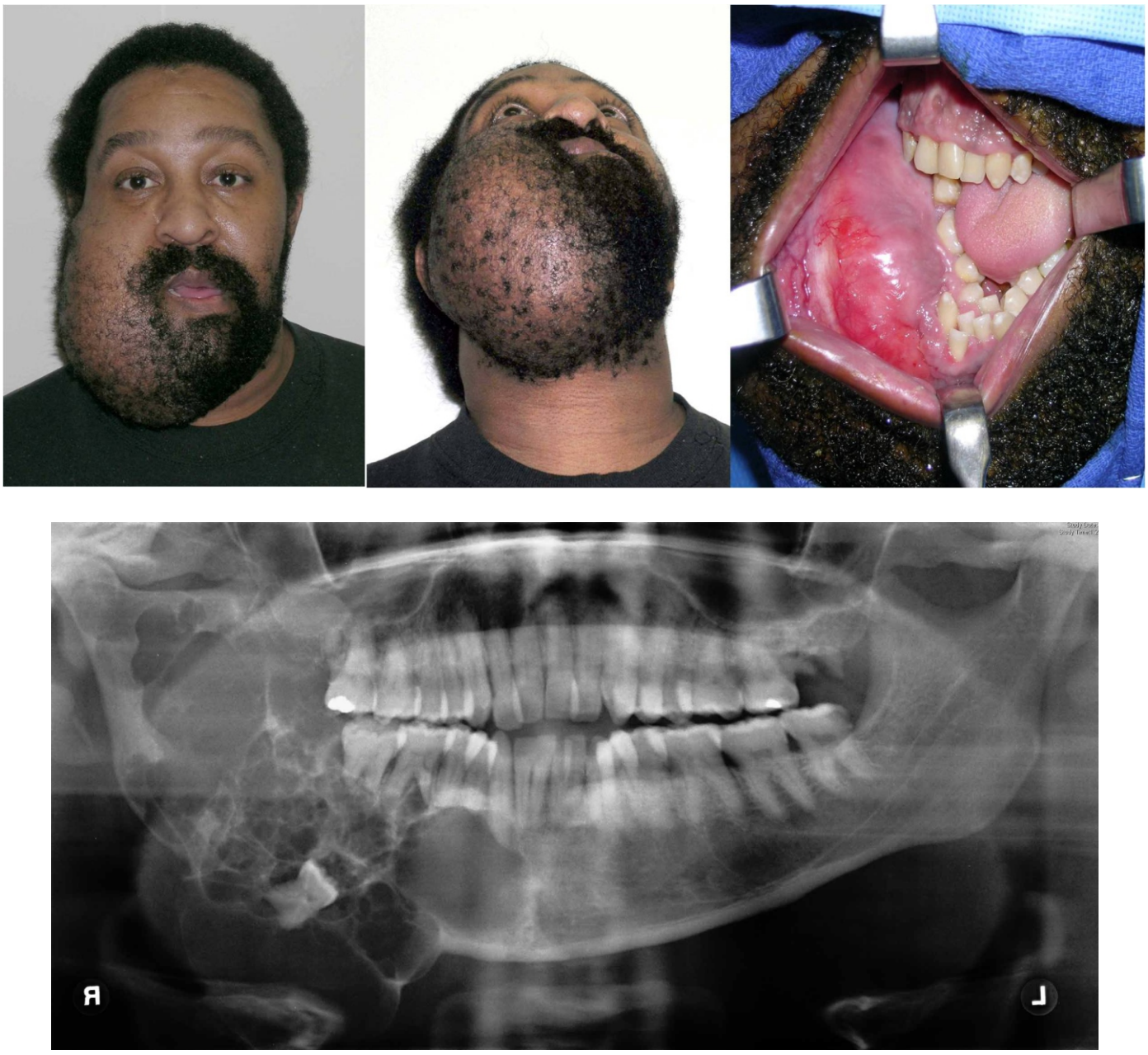


Figure 1: The patient at presentation. Significant expansion of the face is noted with obvious facial disfigurement (figures 1a and b). Of particular note is that the anatomic barrier of skin remains intact associated with this seemingly benign process. Examination of the oral mucosa (figure 1c) reveals expansion and increased vascular markings, presumably induced by the tumor. In addition, the tumor's creation of divergence of the position of the teeth is also noted. The panoramic radiograph (figure 1d) reveals an expansile multilocular radiolucency of the right mandible with root resorption of some of the right mandibular teeth. The right mandibular wisdom tooth is noted within the tumor, supporting the presence of an odontogenic tumor, most likely an ameloblastoma.

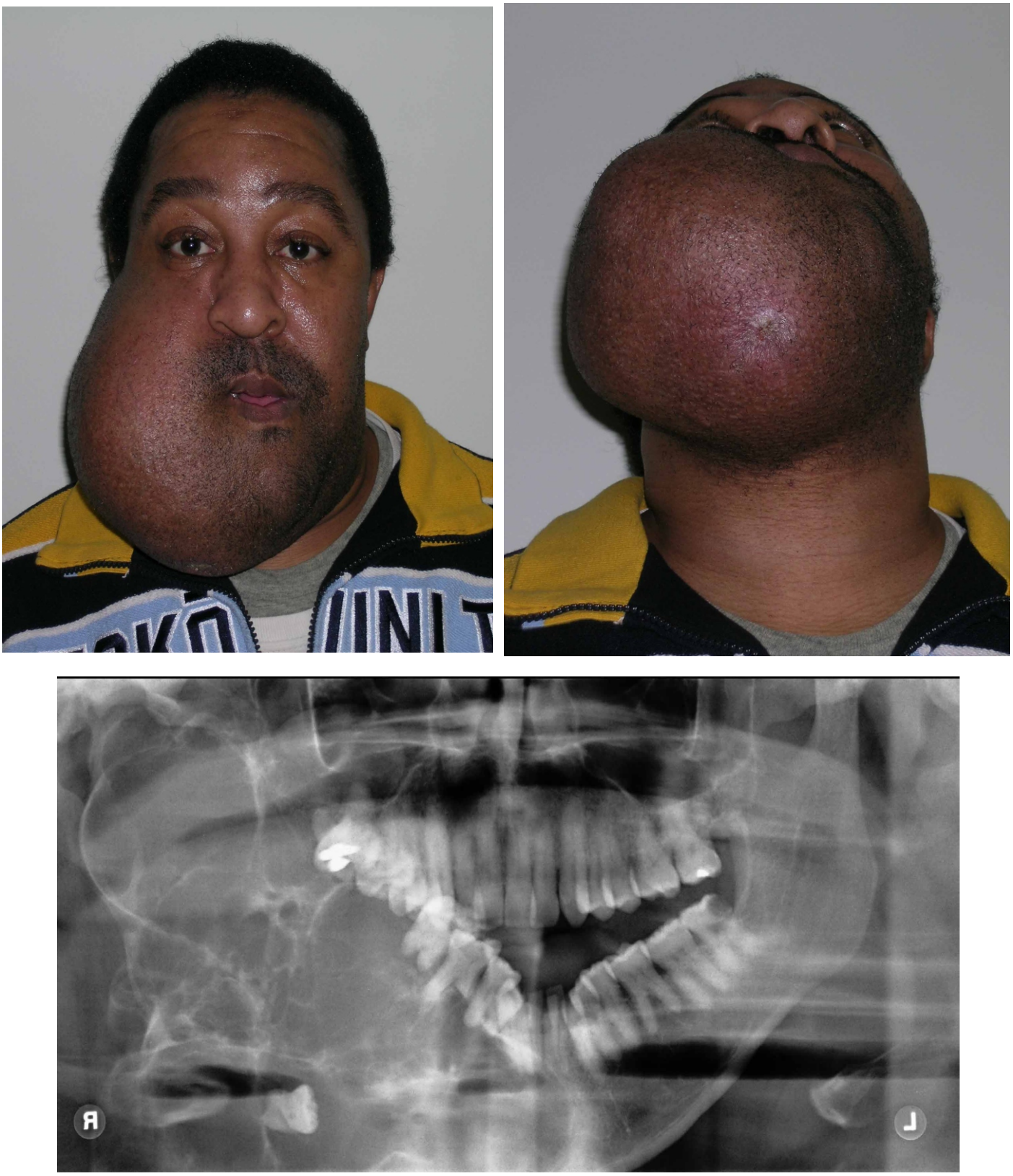
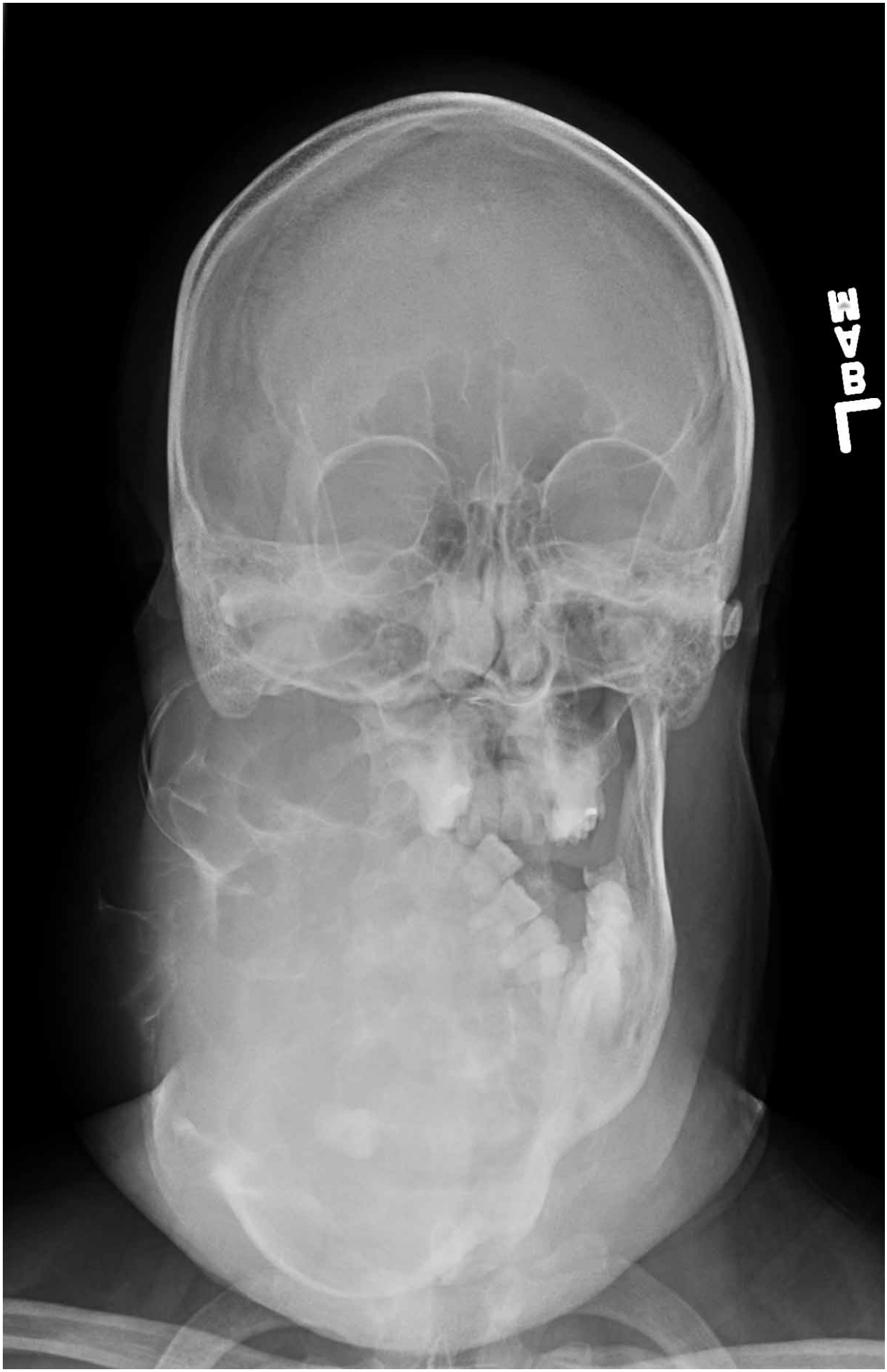


Figure 2: The patient's facial appearance two years later. Increased interval clinical growth of the tumor is appreciated (figures 2a and b). The repeat panoramic radiograph demonstrates interval increased growth of the tumor (figure 2c).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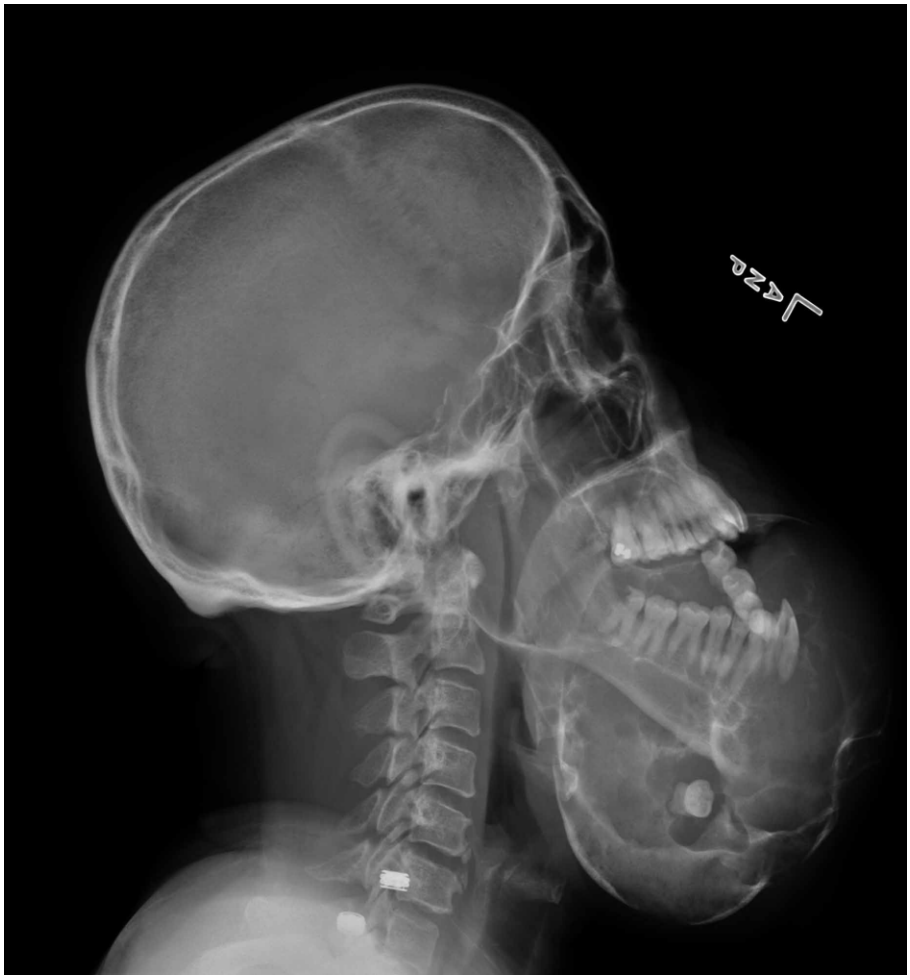
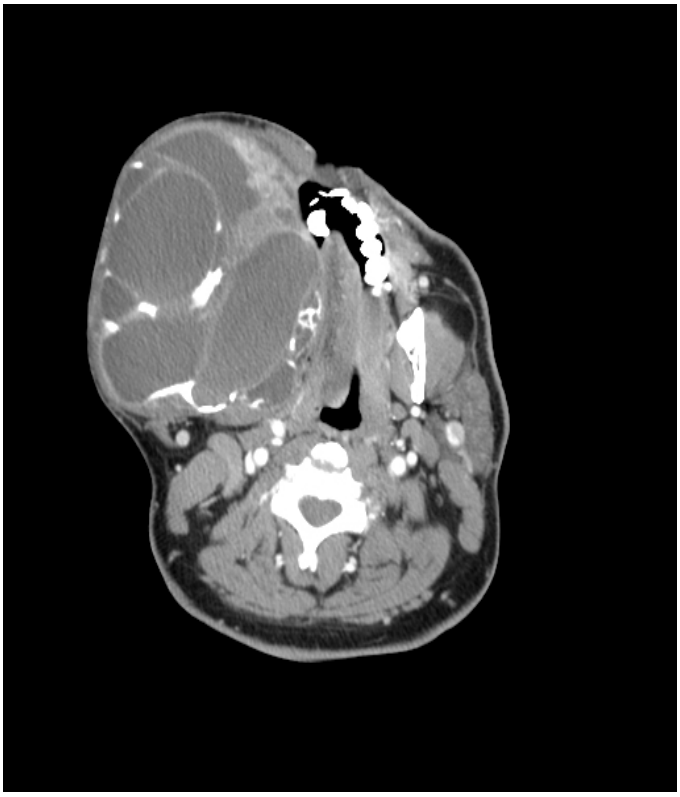
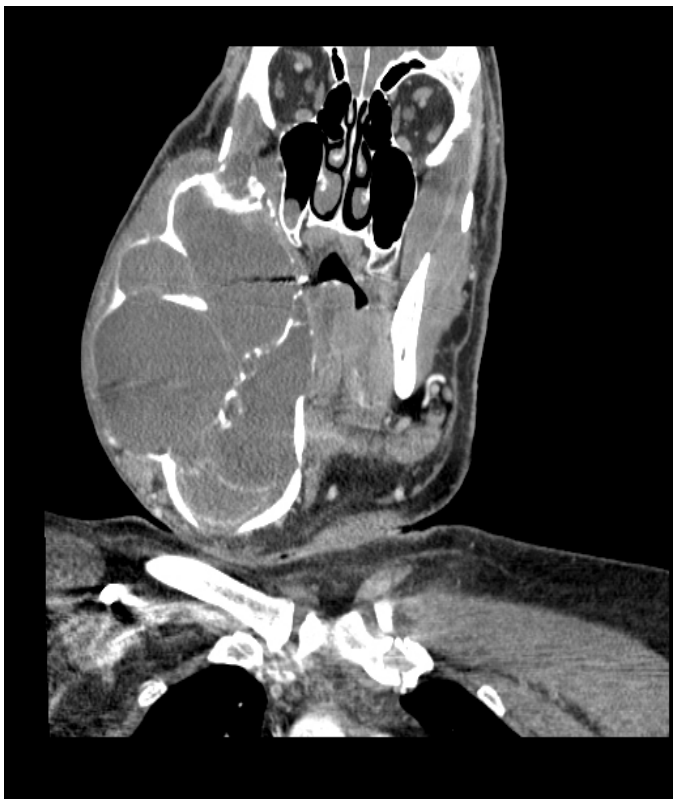


Figure 3: Additional posterior-anterior (figure 3a) and lateral (figure 3b) plain films of the mandible demonstrate the expansile nature and epic proportions of this ameloblastoma.





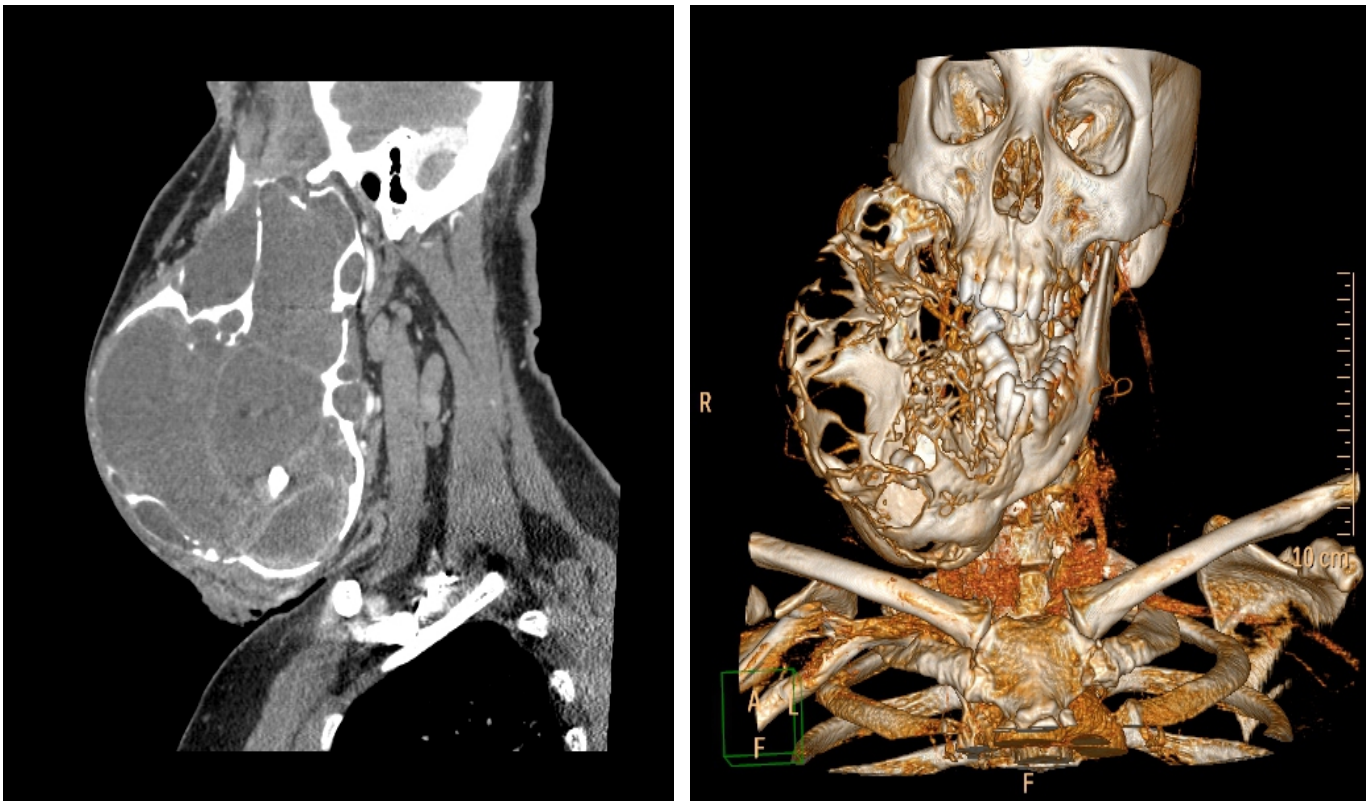


Figure 4: Computed tomograms reveal significant expansion of the right mandible and cortical perforation by the tumor in axial (figure 4a), coronal (figure 4b), and sagittal (figure 4c) projections. In addition, the three dimensional reconstructions of the patient's CT scan (figure 4d) provide a formidable appreciation of the destructive and expansile nature of the tumor.

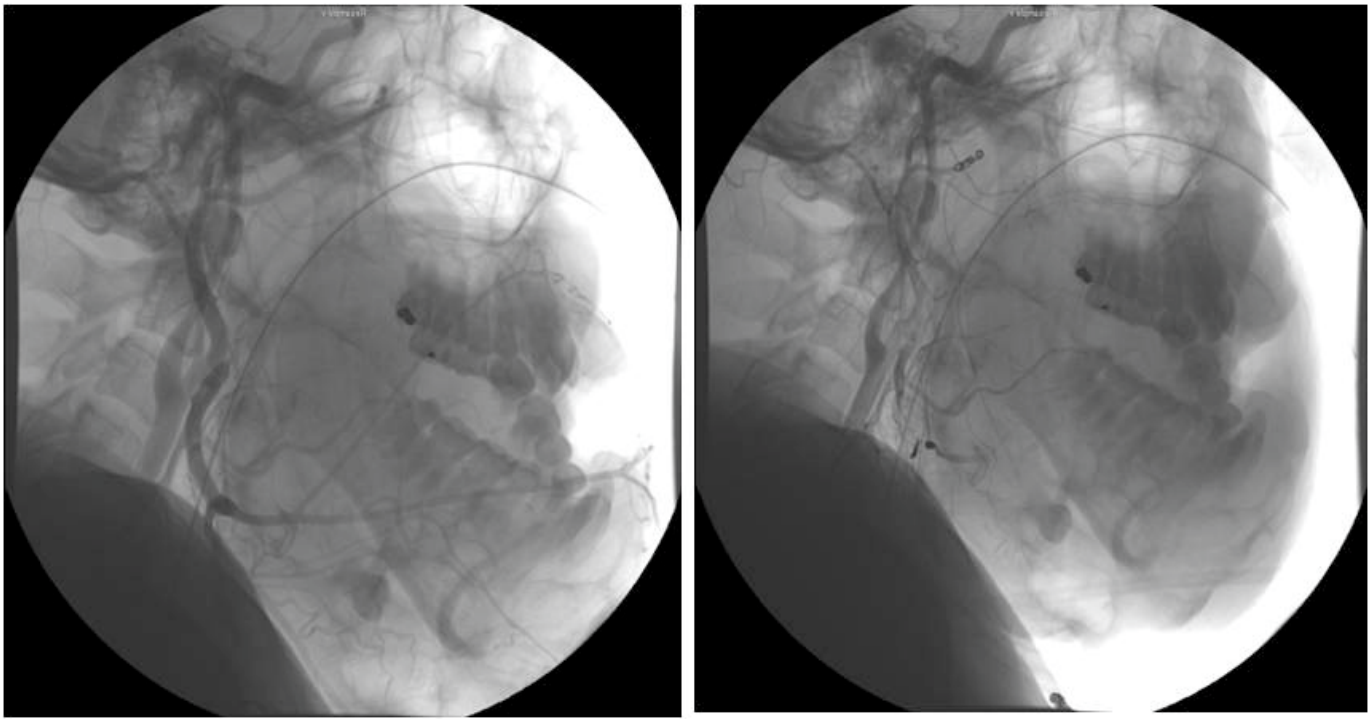


Figure 5: Preoperative diagnostic angiogram (figure 5a) reveals impressive peri-tumor vasculature. Preoperative coil embolization of the maxillary, facial, and lingual arteries (figure 5b) was performed so as to minimize the intraoperative blood loss.

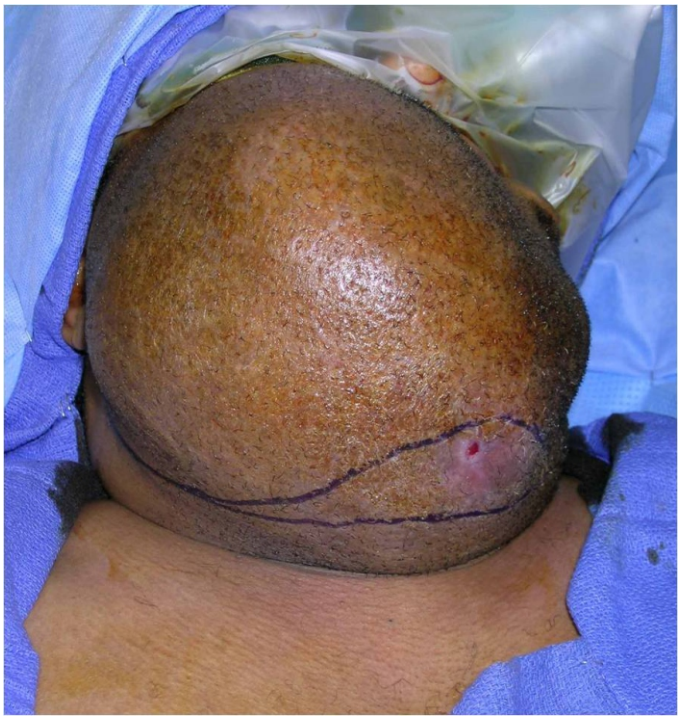


Figure 6: Wide transcutaneous exposure was performed for ablation of this tumor that includes the intentional sacrifice of skin in the submental region of the neck.

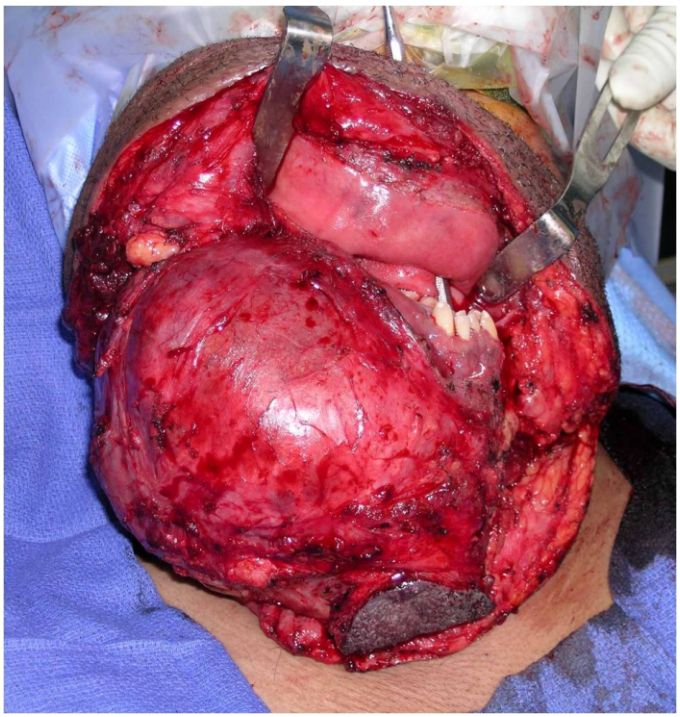
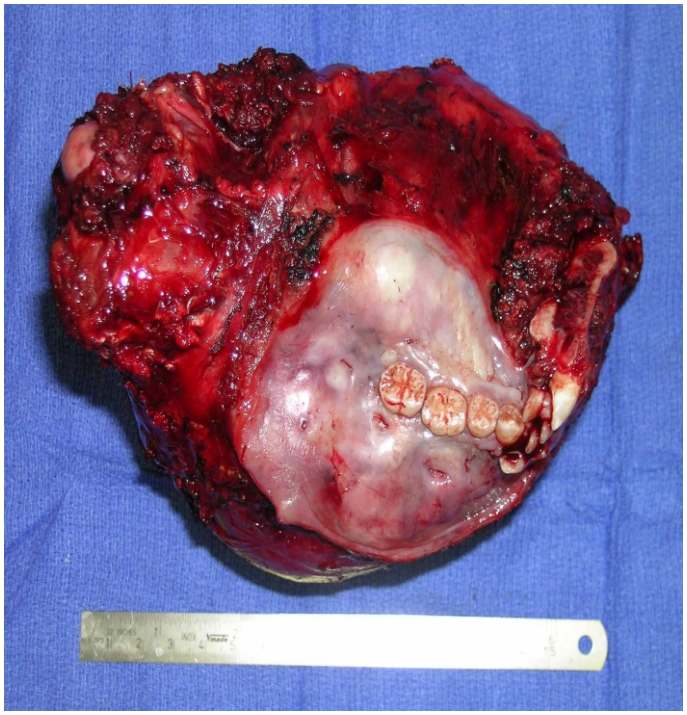


Figure 7: A dissection of the tumor pseudocapsule is performed around the tumor without tumor spillage.



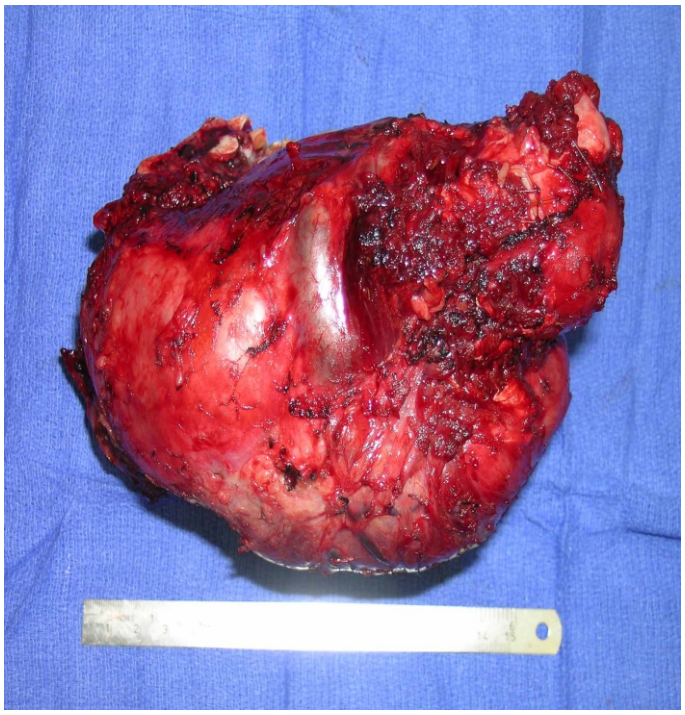


Figure 8: The gross specimen (figures 8a and 8b) from the disarticulation segmental resection of the right mandible.

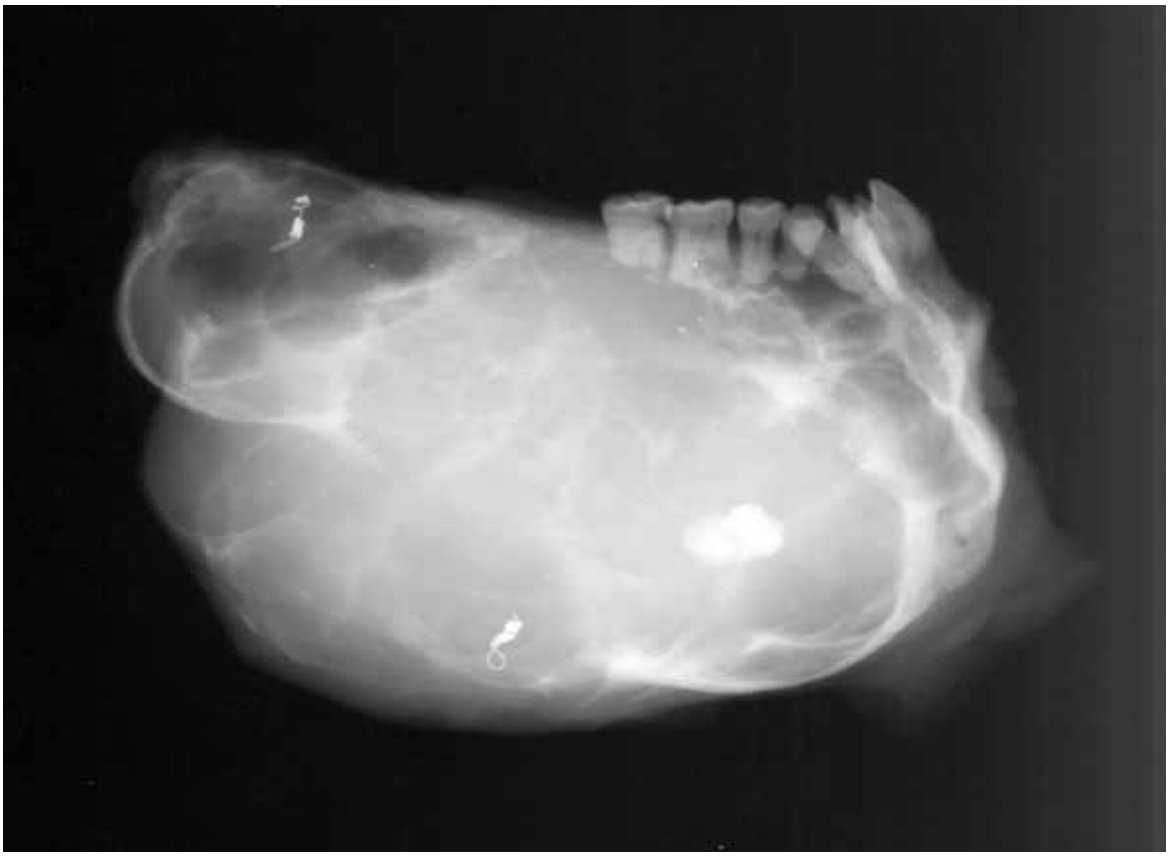


Figure 9: The specimen radiograph provides additional appreciation for the extent of the tumor as well as the root resorption of the right mandibular teeth, indicative of the aggressive nature of this tumor.

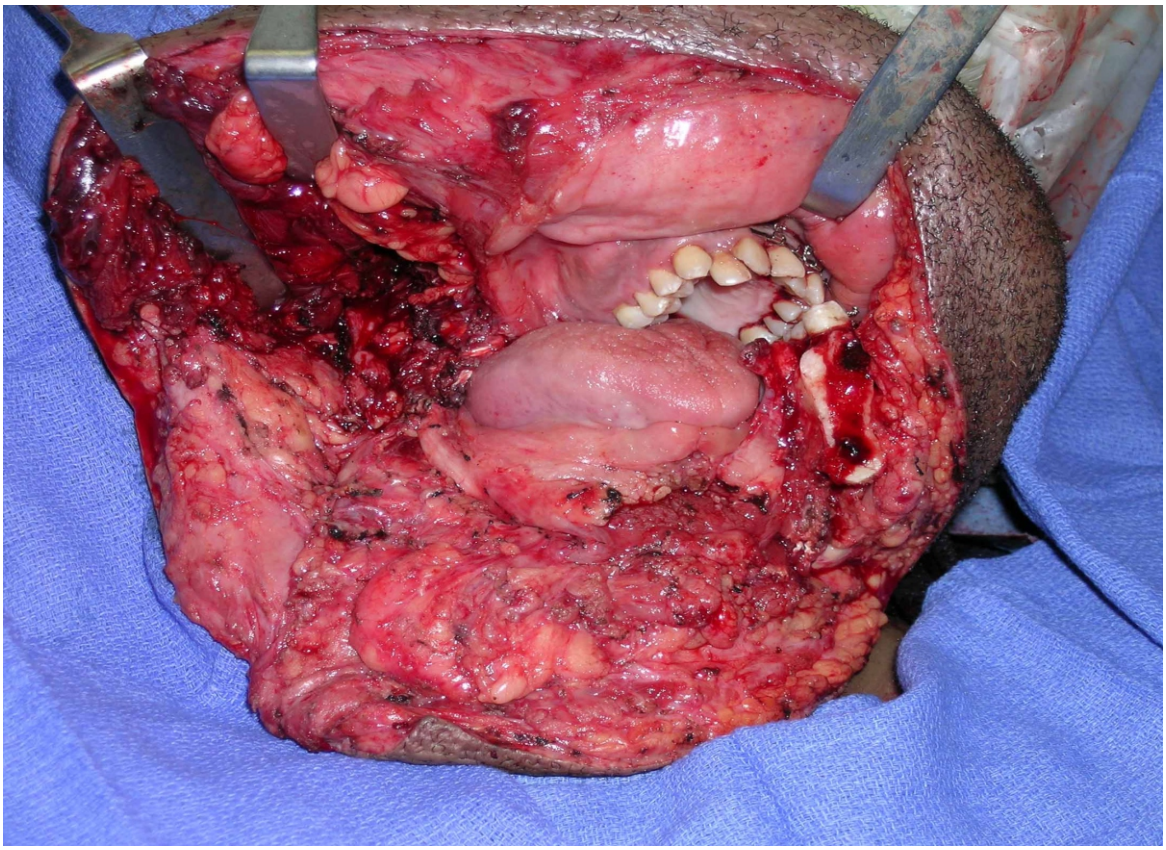


Figure 10: The bony and soft tissue defect following tumor ablation.

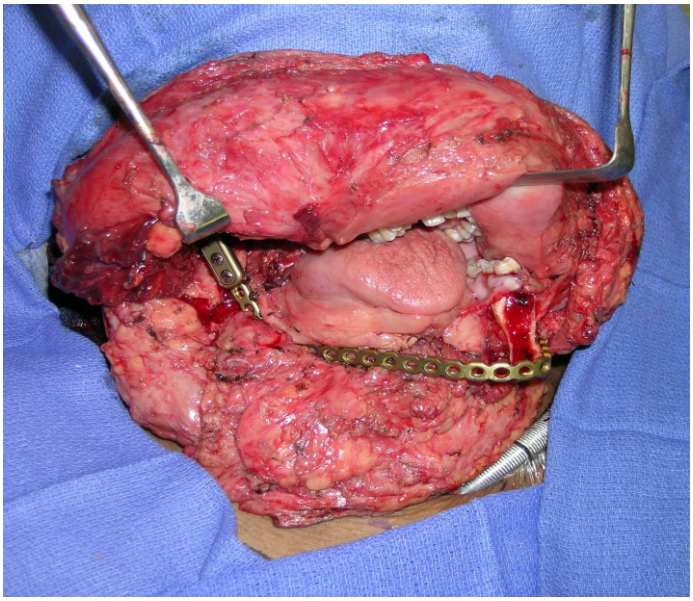


Figure 11: Reconstruction of the mandibular defect is accomplished with a reconstruction bone plate secured to the remaining mandible with four bicortical screws and an affixed condyle for pseudoarticulation in the glenoid fossa.

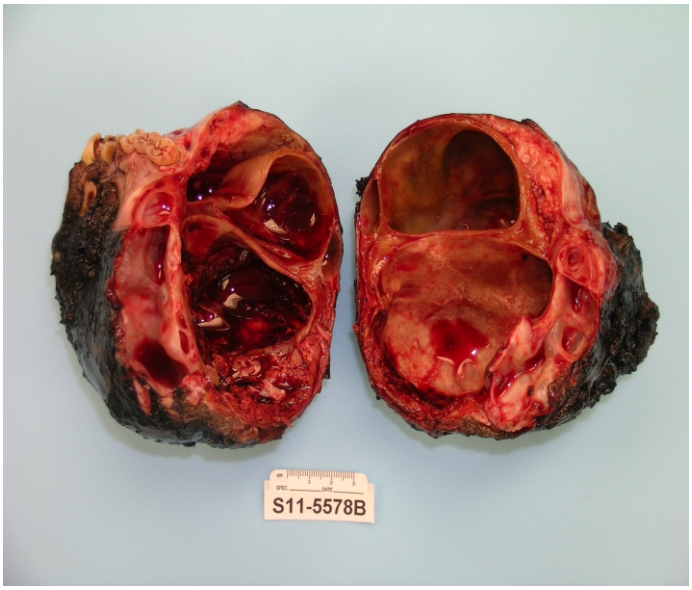


Figure 12: The specimen is bivalved in the pathology laboratory. The tumor notably demonstrates cystic and solid components, consistent with the nomenclature of this tumor.

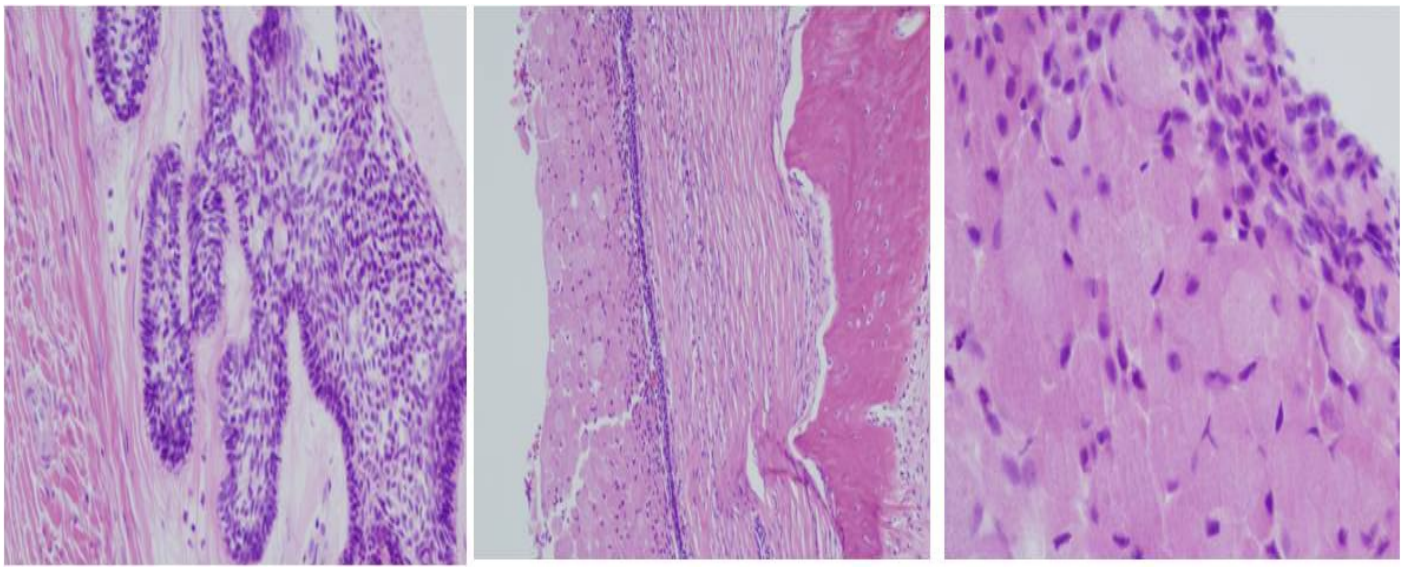


Figure 13: The histopathology of the tumor reveals the presence of a follicular ameloblastoma (figure 13a) with bone destruction (figure 13b) and the presence of granular cells in the tumor (figure 13b and 13c). No signs of malignancy were identified microscopically.

Conclusion

Ablative surgeons should rely on time-honored techniques for the removal of large ameloblastomas of the jaws. While adhering to these techniques, gratifying results can be expected and realized and cure of even very large ameloblastomas can be anticipated. The delay of surgical treatment of these tumors permits an estimate of their doubling time that proclaims slow growth. Chronicity of the tumor should not translate to the identification of malignant transformation of this benign tumor.