UDC: 616.216.1-006.04 DOI: 10.2298/SGS1202110M

Recurrent Solid Ameloblastoma of the Maxillary Sinus: A Case Report

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SUMMARY

Introduction Ameloblastomas are clinically the most important type of odontogenic tumors. Solid or multicystic form most commonly affects mandible, it is highly aggressive and shows high rates of recurrence. The aim was to report aggressive behavior of a rare maxillary solid ameloblastoma, emphasizing the clinical, tomographic and histological aspects.

Case Report A young and asymptomatic patient, presenting a solid ameloblastoma initially located in the maxillary sinus with rapid spreading to the adjacent tissues, had early recurrence despite radical surgical approach.

Conclusion Multicystic or solid ameloblastoma has lower incidence in maxilla and extremely aggressive behavior, justifying careful follow-up of the patients.

Keywords: ameloblastoma; odontogenic tumors; maxillary sinus

INTRODUCTION

Ameloblastomas are the most common neoplasms that originate from odontogenic epithelium accounting approximately 10% of all odontogenic tumors. There are three types of ameloblastomas including two of intraosseous (solid/multicystic and unicystic ameloblastoma) and one of extraosseous origin (peripheral ameloblastoma) [1, 2]. A new classification that included desmoplastic type as an additional variant of ameloblastoma was suggested by the World Health Organization in 2005. Solid/multicystic type corresponds to 86% of all ameloblastomas. This variant is characterized by locally aggressive behavior, no gender preference, high prevalence among young adults and it is mainly located in the posterior mandibular region [3, 4].

The expansion of the solid/multicystic type through medullary spaces, although slow, can culminate in cortical bone erosion. Radiographically, this erosion is characterized by a multilocular infiltrative aspect and characteristic soap bubble-like appearance [5]. An association with retained teeth and root resorption has been reported. Surgical approach with wide resection is the most appropriate treatment because of its high recurrence rate [6, 7, 8]. The diagnosis should be confirmed histologically. A follicular, plexiform, acanthomatous or desmoplastic pattern is typically found, although basaloid and granular cell (desmoplastic) variations might be seen in the histological section of this tumor [9].

The aim of the paper is to report the case of a young patient with extensive solid/multicystic ameloblastoma involving the right maxillary sinus. Asymptomatic course and aggressiveness of the tumor as indicated by its short-term recurrence after surgery are discussed.

CASE REPORT

A 21-year-old male presented with a 6-month history of progressive enlargement of the right posterior region of the maxilla. The patient reported that he himself removed a tooth from the region two weeks earlier and the adjacent teeth were mobile. He was asymptomatic and medical history was insignificant. Physical examination revealed discrete facial asymmetry and an ulcerating exophytic and erythematous mass size about 1.6 cm in width was detected in the alveolar region of the extracted tooth (Figure 1).

A computed tomography (CT) scan showed an enormous opaque area that involved the entire right maxillary sinus and extended into the nasal fossa and orbital cavity on the same side. In addition, the right third molar was displaced superiorly in the direction of the floor of the orbit (Figure 2). An incision biopsy was performed and histological examination revealed islands and anastomosing strands of odontogenic epithelium in a fibrous stroma. Columnar and hyperchromatic basal cells with vacuolated cytoplasm and reverse polarization of the

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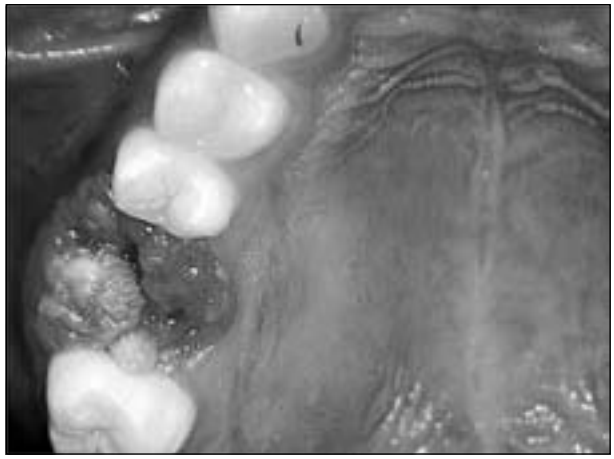

Figure 1. Persistent exophytic mass in the region of the right upper first molar
Slika 1. Egzofitična tumorska masa u predelu gornjeg desnog prvog molara

nuclei were lined up in a palisade pattern at the periphery. The diagnosis was compatible with solid/multicystic ameloblastoma showing features of both the follicular and the plexiform variant (Figure 3).

The patient was referred to the head and neck surgery service for complete excision of the tumor by partial resection of the right maxilla. The surgical specimen consisted of a tissue mass approximately 7×5 cm in size associated with a tooth. Histological analysis confirmed the diagnosis of solid/multicystic ameloblastoma. Despite the radical surgical approach, new tomography scans revealed recurrence of the tumor after eight month follow-up. The patient was therefore submitted to a new surgical intervention that resulted in almost complete resection of the right maxilla and part of the hard palate (Figure 4). Bone scintigraphy with the radioactive tracer technetium-99m did not show increased staining in the treated region. A maxillofacial prosthesis was fabricated for the restoration of masticatory, esthetic and phonetic functions in order to improve the quality of life of the patient.

DISCUSSION

Ameloblastomas are certainly one of the most clinically relevant odontogenic tumors [10]. This benign neoplasm consists of a mixture of ameloblastic epithelium and mesenchyme arising from the remnants of the outer and inner enamel epithelium and dental lamina [1, 11]. The tumor is slow growing but locally aggressive facilitating the infiltration of medullar spaces and sometimes culminating in cortical bone resorption [3, 12]. The current classification of ameloblastomas includes four distinct variants: solid/multicystic, unicystic, extraosseous or peripheral, and desmoplastic (the most recently described variant) [4].

Although small tumors may be diagnosed incidentally in asymptomatic patients by radiography, symptoms such as swelling, malocclusion, tooth loss, pain and paresthesia can be present in cases of more extensive tumors [7]. Ameloblastomas primarily affect the jaws, with a

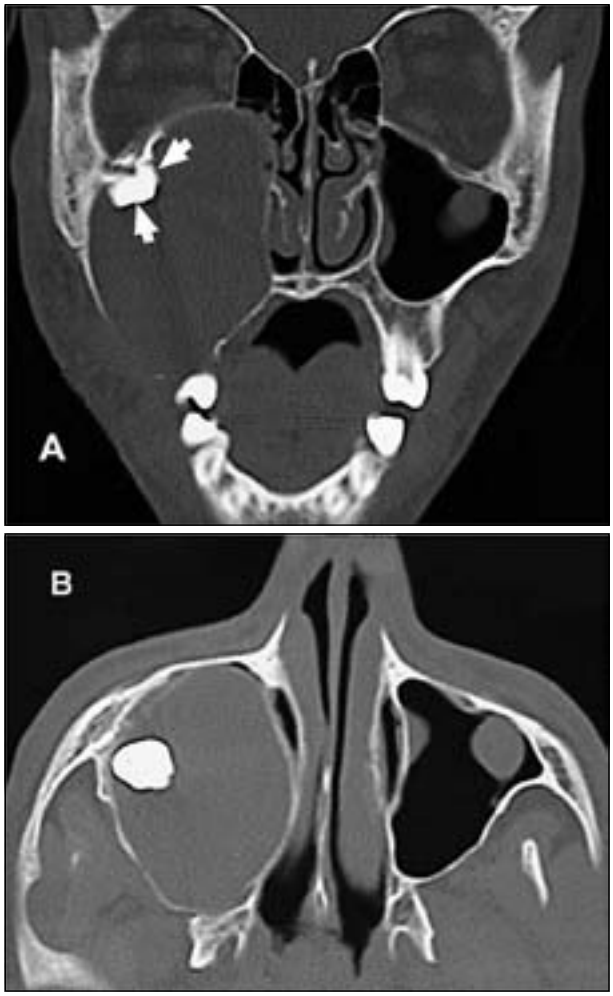

Figure. 2. Coronal (A) and axial (B) computed tomography scans showing a mass in the region of the right maxillary sinus that caused superior displacement of the tooth
Slika 2. Koronarni (A) i aksijalni (B) snimak kompjuterske tomografije koji pokazuje tumor u regiji desnog maksilarnog sinusa koji je izazvao dislokaciju zuba nagore

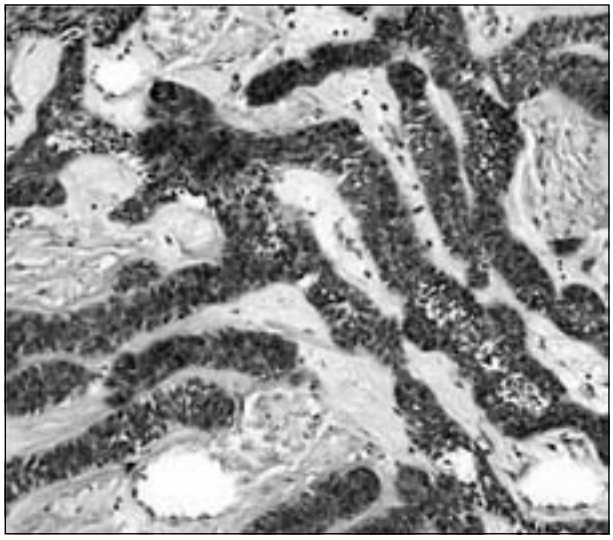

Figure 3. Hematoxylin-eosin stain (20×) showing anastomosing cords of odontogenic epithelium in a fibrous stroma. Note also the presence of stellate reticulum-like cells in some cords.
Slika 3. Bojenje hematoksilin-eozinom (20×) pokazuje anastomotične trake odontogenog epitela u fibroznoj stromi. Zapažaju se ćelije koje podsećaju na ćelije zvezdastog retikuluma u nekim trakama.

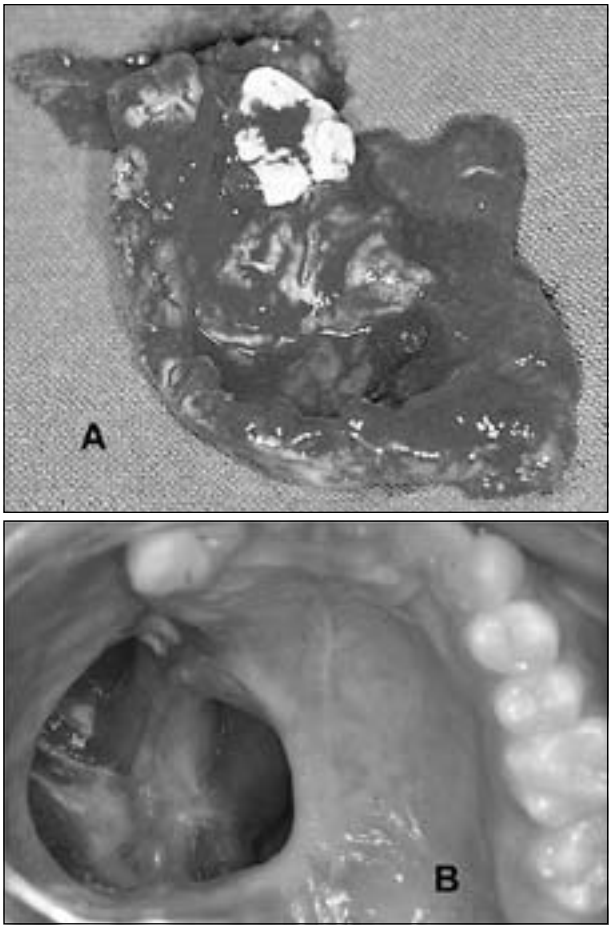

Figure 4. Macroscopic view of the tumor associated with the tooth removed during the first surgery (A) and postoperative view of the surgical pocket remained after the second surgery (B)
Slika 4. Makroskopski izgled tumora povezanog sa zubom uklonjenim tokom prvog hirurškog zahvata (A) i postoperacioni izgled hirurške šupljine koja je ostala nakon druge intervencije (B)

preference for mandibular posterior region. They are characterized by high recurrence rates and rare metastatic potential [8, 13]. In the present study, although the patient was asymptomatic, swelling, tooth loss, and tooth mobility were observed. In addition, the tumor was located in the maxilla contrary to the data that 85% solid ameloblastomas is located in mandible. Tumor progression resulted in complete obliteration of the maxilla, displaced third molar and involved part of right orbital cavity and nasal fossa.

Radiographically, ameloblastomas classically appear as expansive, radiolucent, multiloculated cystic lesions with characteristic soap bubble-like appearance that may occasionally not be detected by CT [14]. CT plays an important role in determining the whole tumor volume and relationship with the surrounding structures including soft tissue. Cystic areas of low attenuation with scattered isoattenuating regions can also be seen, corresponding to the soft tissue components, beyond the thinning and expansion of the cortical plate with erosion through the cortex [6]. Impacted teeth are found in low number of cases [5, 9]. In the present case, CT identified an extensive, opaque, single lesion that caused superior displacement of the third molar and expansion of the cortical plates of the right infraorbital margin and nasal fossa. This nonspecific aspect of the lesion led to the inclusion of unicystic ameloblastoma and dentigerous cyst in differential diagnosis.

The current histological patterns of solid/multicystic ameloblastoma include the follicular, plexiform, acanthomatous, granular and basal cell type [4, 12]. The desmoplastic type is important histological subtype. However, a separate classification was proposed for solid ameloblastomas presenting the desmoplastic pattern because of their peculiar clinical and radiographic characteristics [9]. With respect to the prevalence, the follicular and plexiform types are the most common, in this order, followed by the mixed pattern that is characterized by combination of these two histological types present in a single lesion [15]. In the present case, histological examination showed features of the follicular and plexiform pattern, including islands and anastomosing strands of odontogenic epithelium lying in the fibrous stroma. The epithelium consisted of basal cells showing cytoplasmic vacuolization and reverse nuclei polarization. Loosely arranged cells were also present in the islands and cords, which resembled stellate reticulum and determined the follicular/plexiform histologic pattern.

The type of treatment for solid/multicystic ameloblastomas is certainly a determinant factor for the prognosis of these tumors considering their high rate of recurrence [8]. In contrast to the unicystic ameloblastomas, which usually respond well to conservative therapy, solid ameloblastomas require a radical surgical approach, with margins 1.5 to 2 cm beyond the radiographical limit. Combination with cryosurgery is an option to minimize any possibility of recurrence. Thus, good coordination between the surgeon and radiologist is crucial for the treatment success [12]. In the present study, although a radical surgical approach was chosen in the first intervention and the radiologist participated in the surgical planning, the tumor relapsed after 8 months demonstrating the aggressiveness of these lesions. The location of solid ameloblastomas is also an important factor for the prognosis of these tumors.

In the present case, spongy architecture of maxillary bone allowed rapid spreading of the tumor, which invaded the maxillary sinus and the part of orbital cavity and nasal fossa. These findings support a radical treatment instead of a simple enucleation followed by the curettage. Finally, radiographic and clinical postoperative follow-up should be performed twice per year for the first 24 months and annually up to 10 years after surgical resection, a period of high probability for tumor recurrence.

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Received: 20/04/2012 • Accepted: 28/05/2012

Rekurentni solidni ameloblastom maksilarnog sinusa – prikaz slučaja

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KRATAK SADRŽAJ

Uvod Ameloblastomi su klinički najvažnija vrsta odontogenih tumora. Solidni ili multicistični oblik najčešće je lokalizovan u donjoj vilici, vrlo je agresivan i pokazuje visok procenat recidiva. Cilj rada bio je da se opiše agresivno ponašanje retkog solidnog ameloblastoma u gornjoj vilici, s osvrtom na klinički, radiološki i histološki aspekt.

Prikaz slučaja Kod mlade osobe sa solidnim ameloblastomom lokalizovanim u maksilarnom sinusu uz brzo širenje na susedna tkiva, ali bez simptoma oboljenja, ispoljio se rani recidiv uprkos radikalnom hirurškom lečenju.

Zaključak Multicistični ili solidni ameloblastom je značajno ređi u gornjoj vilici, vrlo je agresivnog ponašanja i zahteva pažljivo nadgledanje bolesnika.

Ključne reči: ameloblastom; odontogeni tumori; maksilarni sinus

UVOD

Ameloblastomi su najčešća vrsta tumora koja potiče od odontogenog epitela, a čini ukupno oko 10% svih odontogenih tumora. Klasifikacija ameloblastoma obuhvata tri tipa: dva intraosealnog porekla (solidno-multicistični i jednocistični ameloblastom) i jedan ekstraosealnog porekla (periferni ameloblastom) [1, 2]. Svetska zdravstvena organizacija je 2005. godine predložila novu klasifikaciju koja uključuje i dezmoplastični tip, kao dodatnu varijantu ameloblastoma. Solidni (multicistični) tip je najčešći i javlja se u 86% slučajeva svih ameloblastoma. Ovaj tip ispoljava lokalno agresivno ponašanje, ne postoji polna predispozicija i uglavnom se otkriva u zadnjem delu mandibule [3, 4].

Širenje solidnog (multicističnog) tumora kroz medularne prostore, premda sporo, može izazvati eroziju kortikalne kosti. Radiološki, solidni ameloblastom ima tipičan izgled lezije koja je multilokularna, infiltrirajuća i s prepoznatljivim izgledom mehura sapunice [5]. Često može sadržati neiznikli zub i izazvati resorpciju korena. Hirurški pristup sa širokom resekcijom tkiva je najprikladniji način lečenja zbog visokog procenta recidiva ovog tumora [6, 7, 8]. Dijagnozu treba potvrditi histološki. Folikularni, pleksiformni, akantozni ili dezmoplastični izgled tkiva je uobičajen histološki nalaz, iako se mogu uočiti i bazaloidne i granularne ćelije [9].

Cilj rada bio je da se prikaže slučaj mladog bolesnika s opsežnim solidno-multicističnim tipom ameloblastoma lokalizovanim u desnom maksilarnom sinusu. Asimptomatski tok, agresivnost tumora i brzi recidiv nakon operacije su prodiskutovani.

PRIKAZ SLUČAJA

Dvadesetjednogodišnji muškarac je došao na kliniku sa šestomesečnom istorijom progresivnog proširenja zadnjeg dela gornje vilice sa desne strane. Potvrdio je da je sam izvadio zub iz te regije dve nedelje pre dolaska na kliniku, a susedni zubi su bili mobilni. Nije imao nikakve subjektivne simptome, a medicinska istorija je bila bez značajnih promena. Pregledom tkiva utvrđene su diskretna asimetrija lica i ulcerozna egzofitična i eritematozna masa veličine oko 1,6 cm u regiji alveolarnog grebena na mestu izvađenog zuba (Slika 1).

Kompjuterizovana tomografija (CT) je otkrila da postoji neprovidna regija koja je pokrivala čitav desni maksilarni sinus, nosnu jamu i očnu duplju s iste strane. Osim toga, desni umnjak je bio potisnut nagore, ka podu orbite (Slika 2). Nakon incizijske biopsije, patohistološki pregled je pokazao ostrvca i anastomozirane trake odontogenog epitela u fibroznoj stromi. Kolumnarne i hiperhromatične bazalne ćelije s vakuoliziranom citoplazmom i obrnutom polarizacijom nukleusa bile su palisadno poređane na periferiji. Dijagnoza je glasila: solidni, multicistični ameloblastom s obeležjima folikularnog i pleksiformnog tipa tumora (Slika 3).

Bolesnik je upućen na operaciju glave i vrata, kako bi se tumor potpuno odstranio, sa delimičnom resekcijom gornje desne maksile. Hirurški uzorak se sastojao od tkiva veličine 7×5 cm i zuba. Histološka analiza potvrdila je dijagnozu solidnog, multicističnog ameloblastoma. I pored radikalnog hirurškog pristupa, CT snimak je nakon osam meseci pokazao recidiv tumora. Druga hirurška intervencija je obuhvatila gotovo potpunu resekciju desne maksile i deo tvrdog nepca (Slika 4). Koštana scintigrafija s radioaktivnim tehnecijumom (99m) nije ukazala na povećanu senku na tretiranom području. Bolesniku je napravljena maksilofacijalna proteza za obnovu mastikatorne, estetske i funkcije govora, radi poboljšanja kvaliteta života.

DISKUSIJA

Ameloblastomi su sigurno jedan od klinički najvažnijih odontogenih tumora [10]. Ovaj benigni tumor se sastoji od ameloblastičnog epitela i mezenhima koji potiče od ostataka spoljašnjeg i unutrašnjeg gleđnog epitela i dentalne lamine [1, 11]. Tumor sporo raste, ali je lokalno agresivan, lako zauzima medularne prostore, što može dovesti do resorpcije kortikalne kosti [3, 12]. Sadašnja klasifikacija ameloblastoma uključuje četiri različita tipa: solidno-multicistični, jednocistični, ekstraosealni ili periferni i dezmoplastični (najnoviji tip) [4].

Iako se mali tumori mogu dijagnostikovati slučajno kod bolesnika bez simptoma oboljenja pomoću radiološkog snimka, simptomi kao što su otok, ortodontske anomalije, gubitak zuba, bol i parestezija mogu postojati kod većih tumora [7]. Ameloblastomi se najčešće lokalizuju u vilicama, naročito u zadnjem delu mandibule. Odlikuje ih visok procenat recidiva i retko metastaziranje [8, 13]. Iako prikazani bolesnik nije imao nikakve subjektivne simptome, postojali su objektivni znaci, poput otoka, gubitka i mobilnosti zuba. Osim toga, tumor je bio lokalizovan u gornjoj vilici, suprotno podacima da se 85% solidnih ameloblastoma nalazi u donjoj vilici. Napredovanje tumora dovelo je do potpune obliteracije gornje vilice i dislokacije umnjaka, šireći se na pod desne orbite i nosne jame.

Radiološka slika ameloblastoma je tipična, prostrana radiolucentna i multilokularna cistična lezija s karakterističnim izgledom mehura sapunice, ponekad teško uočljiva čak i pomoću CT [14]. CT ima važnu ulogu u otkrivanju čitavog volumena tumora i njegovog odnosa s okolnim strukturama, uključujući meko tkivo. Cistična područja s niskom atenuacijom vazduha s rasutim regijama slične atenuacije takođe se mogu uočiti, što odgovara mekotkivnoj komponenti, pored istanjivanja i širenja kortikalne ploče s erozijom kroz korteks [6]. Impaktirani zubi nalaze se u malom broju slučajeva [5, 9]. U ovom slučaju pomoću CT je ustanovljena velika lezija visokog opaciteta koja je izazvala pomeranje umnjaka nagore i širenje kortikalne ploče infraorbitalne ivice i nosne jame sa desne strane. Ovakav nespecifičan izgled lezije doveo je do uključivanja jednocističnog ameloblastoma i folikularne ciste u diferencijalnu dijagnozu.

Histološki izgled solidnog, multicističnog ameloblastoma obuhvata folikularni, pleksiformni, akantozni, granularni i bazilarni tip ćelija [4, 12]. Dezmoplastični tip je takođe važan histološki podtip. Međutim, posebna klasifikacija je predložena za solidni ameloblastom dezmoplastičnog tipa zbog svojih specifičnih kliničkih i radioloških obeležja [9]. S obzirom na učestalost, folikularni i pleksiformni tip ameloblastoma su najčešći, a potom sledi mešoviti tip, koji se odlikuje kombinacijom ova dva histološka tipa u jednoj leziji [15]. U ovom prikazu histološki pregled je pokazao osobine folikularnog i pleksiformnog tipa, uključujući ostrvca i anastomoze odontogenog epitela u fibroznoj stromi. Epitel se sastojao od bazilarnih ćelija koje su pokazivale citoplazmatičnu vakuolizaciju i obrnutu polarizaciju nukleusa. Labavo poređane ćelije su takođe bile u obliku ostrvaca i traka, dajući sliku koja podseća na zvezdasti retikulum i određuje folikularno-pleksiformni histološki model.

Način lečenja bolesnika sa solidnim, multicističnim ameloblastomom je svakako odlučujući faktor za prognozu ovih tumora, s obzirom na njihov visok procenat recidiva [8]. Za razliku od jednocističnih ameloblastoma, koji odlično reaguju na konzervativnu terapiju, solidni ameloblastom zahteva radikalni hirurški pristup, sa ivicama 1,5-2 cm izvan radiološke granice. Kriohirurgija je dodatna opcija, da bi se smanjila mogućnost recidiva. Dobra koordinacija između hirurga i radiologa je presudna za uspeh lečenja bolesnika [12]. Iako je kod prikazanog bolesnika preduzet radikalni hirurški zahvat već pri prvoj intervenciji, a radiolog učestvovao u planiranju hirurškog lečenja, stanje bolesnika se pogoršalo nakon osam meseci usled vidno povećane agresivnosti tumora. Mesto solidnih ameloblastoma je takođe važan činilac u prognozi ovih tumora.

U ovom prikazu bolesnika porozni izgled gornje vilice pogodovao je brzom širenju tumora, koji je zahvatio maksilarni sinus, deo orbite i nosne jame. Ovi rezultati govore u prilog potrebi za radikalnom operacijom umesto jednostavne enukleacije i kiretaže tumora. Radiološko i kliničko postoperaciono praćenje bolesnika neophodno je dva puta godišnje u prva 24 meseca, a onda jednom godišnje do 10 godina (vreme mogućeg recidiva tumora) nakon hirurške resekcije.