UNIVERSIDADE FEDERAL DO RIO DE JANEIRO

CENTRO DE CIÊNCIAS DA SAÚDE

FACULDADE DE ODONTOLOGIA

KERATOAMELOBLASTOMA: A VERY RARE LESION WITH AN UNUSUAL RECURRENCE

Rafael Luís Ferreira Netto Cardoso

Rio de Janeiro, RJ - Brasil

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Dissertação submetida à Banca Examinadora do Metsrado Profissional em Clínica Odontológica como parte dos requisitos para obtenção do título de Mestre em Clínica Odontológica.

Orientadora: Profª Drª Maria Elisa Rangel Janini

Rio de Janeiro, RJ - Brasil

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“Quem precisa de ordem pra moldar?
Quem precisa de ordem pra pintar?
Quem precisa de ordem pra esculpir?
Quem precisa de ordem pra narrar?
Quem precisa de ordem?
Agora uma fabulazinha
Me falaram sobre uma floresta distante
Onde uma estória triste aconteceu
No tempo em que os pássaros falavam
Os urubus, bichos altivos mas sem dotes para o canto, resolveram,
Mesmo contra a natureza, que haveriam de se tornar grandes cantores
Abriram escolas e importaram professores
Aprenderam dó-ré-mi-fá-sol-lá-si
Encomendaram diplomas e combinaram provas entre si
Para escolher quais deles passariam a mandar nos demais
A partir daí criaram concursos e inventaram títulos pomposos
Cada urubuzinho aprendiz sonhava um dia se tornar um ilustre urubu titular
A fim de ser chamado por Vossa Excelência
Passaram-se décadas até que a patética harmonia dos urubus-maestros
Foi abalada com a invasão da floresta por canários tagarelas
Que faziam coro com periquitos festivos e serenatas com os sabiás
Os velhos urubus, encrespados, entortaram o bico
E convocaram canários, periquitos e sabiás
Para um rigoroso inquérito
"Cada os documentos de seus concursos?" indagaram
E os pobres passarinhos se olharam assustados
Nunca haviam freqüentado escolas de canto pois o canto nascera com eles
Seu canto era tão natural que nunca se preocuparam em provar que sabiam cantar
Naturalmente cantavam
"Não, não, não assim não pode, cantar sem os documentos devidos
É um desrespeito à ordem!"
Bradaram os urubus
E em uníssono expulsaram da floresta os inofensivos passarinhos
Que ousavam cantar sem alvarás
Moral da estória:
Em terra de urubus diplomados não se ouve o canto dos sabiás”

Muito Obrigado (Letra: Fred 04 – Música: Mundo Livre S/A)

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RESUMO

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A denominação ceratoameloblastoma tem sido utilizada para descrever um grupo histológico heterogêneo de variantes do ameloblastoma, que tem em comum a formação de ceratina pelo epitélio ameloblastomatoso. Até o momento, vinte casos foram previamente reportados na literautra, dos quais cinco exibem um componente papilifero. Nós relatamos um novo caso de um tumor recidivado que se enquadra no espectro do keratoameloblastoma, o qual apresentava uma lesão expansiva, sólida, com calcificações internas, na fossa infratemporal direita, seis anos após uma hemimandibulectomia ipsilateral, de uma mulher branca de 46 anos. Ilhas de células colunares que lembram ameloblastoma ao redor de uma área central com células estreladas, algumas das quais completamente preenchidas por ceratina e outras exibindo células basais colunares a cuboidais com núcleo hipercromático, foram observadas na avaliação histológica do espécime. Nós revisamos o padrão clínico, histopatológico e radiográfico dos casos previamente publicados de ceratoameloblastoma, além do tratamento e acompanhamento realizado. Embora um pequeno número de casos tenha sido reportado, o comportamento biológico agressivo e altas taxas de recorrência sugerem que um manejo mais agressivo deve ser realizado. Ressecção com margens de segurança e análise histopatológica dessas margens são altamente recomendadas.

Palavras-chave: Tumores odontogênicos, ameloblastoma, ceratoameloblastoma, recidiva

ABSTRACT

The denomination keratoameloblastoma has been used to describe a histologically heterogeneous group of ameloblastoma variants which have in common the formation of keratin by the ameloblastomatous epithelium. Up to now twenty cases of keratoameloblastoma have been previously reported in the literature, of which five exhibited a papilliferous component. Here we report a new case of a relapsed tumor that fits the spectrum of keratoameloblastoma which presented as an expansile, solid lesion with internal calcification in the right infratemporal fossa six years after ipsilateral hemimandibulectomy of a 46-year-old white female. Islands of columnar cells resembling ameloblasts surrounding a central area with starry cells, some of them completely filled with keratin and others also showing columnar to cuboidal basal cells with hypercromatic nuclei were observed in the histological evaluation of the specimen. The clinical, histopathologic and radiographic features of keratoameloblastoma are reviewed so as treatment and follow up. Although only few cases have been reported, the biological aggressive behavior and the high recurrence suggest that a more aggressive approach should be performed. A resection with sufficient safety margins and histopathological analysis of surgical margins are highly recommended.

Keywords: Odontogenic tumors, ameloblastoma, keratoameloblastoma, recurrence

SUMMARY

ARTICLE

Keratoameloblastoma: a very rare lesion with an unusual recurrence

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ABSTRACT

The denomination keratoameloblastoma has been used to describe a histologically heterogeneous group of ameloblastoma variants which have in common the formation of keratin by the ameloblastomatous epithelium. Up to now twenty cases of keratoameloblastoma have been previously reported in the literature, of which five exhibited a papilliferous component. Here we report a new case of a relapsed tumor that fits the spectrum of keratoameloblastoma which presented as an expansile, solid lesion with internal calcification in the right infratemporal fossa six years after ipsilateral hemimandibulectomy of a 46-year-old white female. Islands of columnar cells resembling ameloblasts surrounding a central area with starry cells, some of them completely filled with keratin and others also showing columnar to cuboidal basal cells with hypercromatic nuclei were observed in the histological evaluation of the specimen. The clinical, histopathologic and radiographic features of keratoameloblastoma are reviewed so as treatment and follow up. Although only few cases have been reported, the biological aggressive behavior and the high recurrence suggest that a more aggressive approach should be performed and the patient must be aware for the importance of clinical control. A resection with sufficient safety margins and histopathological analysis of surgical margins are highly recommended.

INTRODUCTION

An unusual variant of ameloblastoma demonstrating ameloblastic islands filled with keratin and exhibiting varying degrees of keratinization was first described by Pindborg[1] as papilliferous keratoameloblastoma (PKA) in 1970

and further named keratoameloblastoma (KAB) by Altini et al[2] in 1976, respectively. Keratoameloblastoma is a very rare lesion and up to now after these previous reports, only four more cases[3-6] with papilliferous pattern and fourteen non-papiliferous[7-17] were published in English literature. Despite the similarity of names, keratoameloblastoma and papilliferous keratoameloblastoma are distinct morphologically. PKA is described as cystic spaces filled with necrotic debris and lined by papillary keratin infolding of odontogenic epithelium resembling ameloblastoma. The odontogenic epithelium consists of cells resembling stellate reticulum of the enamel organ and basal layer of tall columnar ameloblast-like cells showing palisading and reversal polarity[4,6]. KAB is histologically described as cystic follicles filled with parakeratin, orthokeratin, and necrotic material with calcification and lined by stratified squamous epithelium exhibiting hyperchromatic, palisaded basal cells with focal reverse polarity, subnuclear vacuolation and also peripheral areas resembling odontogenic keratocyst[4,11]. Both PKA and KAB generally presents as a mandibular painless swelling in adult male patients (Table I). The radiographic aspect is of a uni or multilocular lesion, sometimes irregular, with few cases showing calcification[5,11,17], leading to osteolysis and eroding cortical bone. Due to its high rate of recurrence (23,8%) a more aggressive approach should be considered. The most common treatment is resection. Curettage and enucleation also have been performed. Due to the rarity of the lesion, up to now the treatment and follow up cannot be associated with recurrence[11,16,17]. Follow up is well established only in eight cases[3,5,8,11,14-17]. Four of them[5,8,16,17] presented relapse of the tumor (two[8,16] were treated primary with enucleation

and two[5,17] by resection) and the other four remaining cases[3,11,14,15] showed no evidence of disease (follow up time varying 10 to 24 months with).

We report a case of keratoameloblastoma with an unusual pattern of recurrence and review the previously reported cases, with emphasis on the histological features, prognosis and follow up and treatment of each case.

CASE REPORT

A 46 years old white female was referred to the Stomatology Service of a public School of Dentistry with a chief complaint of a swelling on the right infratemporal fossa. The lesion was noted about a year before first clinical examination. The patient reported respiratory problems (long-term bronchitis) and a previous surgery to excise a mandibular tumor 6 years ago, which she could not precise the diagnosis. The extra-oral examination showed a firm well-circumscribed painless swelling in right infratemporal region, measuring about 7 cm on its longest axis and a surgical scar in the middle mandibular region (Figure 1). At intra-oral examination, the clinical absence of right left molars was observed. Immediate radiographic evaluation confirmed the absence of part of the body, ramus and right mandibular condyle. The original diagnosis, the slides of the specimen removed in the mandibular surgical intervention and a computed tomography imaging study (CT) were requested. The CT image showed a massive swelling in the right infratemporal region with osteolysis of the zygomatic arch and areas of calcification (Figure 2). The histopathological diagnosis of the mandible specimen was of KAB. Due to the rarity of this lesion, the slides were reviewed by three experienced Oral Pathologists, which confirmed the diagnosis. The patient was referred to a maxillofacial surgery

service of a public hospital where an incisional biopsy was performed. The histopathological examination revealed a solid lesion composed of islands of columnar cells resembling ameloblasts surrounding a central area with starry cells, some of them completely filled with keratin and others also showing columnar to cuboidal basal cells with hypercromatic nuclei (Figure 3). These features were the same observed in the mandibular lesion. The diagnosis of KAB was confirmed suggesting that the infratemporal lesion was a recurrence of the mandibular one. The suggested and performed treatment was the total removal of the infratemporal lesion with safety margins and reconstruction of the zygomatic arch with autogenous skullcap graft (Figures 4 and 5). Histopathological evaluation of the surgical specimen revealed the same features observed in the incisional biopsy and in the mandibular lesion. The findings corroborated the final diagnosis of KAB, supporting that the lesion was a recurrence of a previous similar tumor. The patient is under clinical and radiographic follow-up for 36 months with no signs of recurrence (Figures 6 and 7).

DISCUSSION

Ameloblastoma is the most common odontogenic epithelial tumor of the jaws and accounts for only 1% of all oral tumors and 10% of all odontogenic tumors[6,14,18]. Generally, it is slow-growing but locally invasive, with a high rate of recurrence if not treated adequately. Its incidence, combined with its clinical behavior, makes ameloblastoma the most significant odontogenic neoplasm[15]. It occurs in various forms and is classified into multicystic, unicystic, desmoplastic, and peripheral clinical types. Multicystic ameloblastoma is

histologically classified as follicular (spindle cell, basal cell, granular cell, and acanthomatous ameloblastoma) and/or plexiform. In addition, there is another rare subtype known as KAB[1,17,19,20]. Ameloblastoma is highly polymorphic, due to its ability to undergo various forms of metaplasia. The stimulus for the metaplastic change is poorly understood but has been attributed to the multipotentiality of odontogenic epithelium[12]. Although there is no evidence that any histological variation is more aggressive than anyother, unicystic ameloblastomas are generally associated with a lower post-operative recurrence rate than the multicystic types[10,21,22]. The lesion occurs mostly in the 4th or 5th decades of life, with no gender predilection, and in the posterior molar-ramus region and ascending ramus of the mandible[14,18]. Ameloblastomas can spread through the cancellous bone, causing osteolysis and perforation of the compact bone, beyond resorption of dental roots[5]. The keratocystic odontogenic tumor is a benign uni or multicystic, intraosseous potentially aggressive odontogenic tumor, with a characteristic lining of parakeratinized stratified squamous epithelium and an infiltrative behavior. Although this lesion presents a benign behavior, the WHO Working Group recommends the term keratocystic odontogenic tumor (KCOT) as it better reflects its neoplastic nature. It generally occur in the posterior region of the mandible of males from the first to the ninth decades with a peak of incidence in the second and third decades[20,23]. One of the most important clinical feature of the KCOT, as in the ameloblastoma, is its potential for locally destructive behavior, its recurrence rate and its tendency to multiplicity. Patients may complain of pain, swelling or discharge. These tumors may reach a large size prior to discovery and may penetrate cortical bone and involve adjacent structures. Adjacent teeth may be

displaced but root resorption occurs rarely. KCOTs may appear as small, round or ovoid unilocular radiolucencies or may be larger with scalloped margins. The radiolucencies tend to be well-demarcated with distinct sclerotic margins, but may be diffuse in parts. True multilocular mandibular lesions are not uncommon. CT scans may be helpful in detecting cortical perforation and assessment of soft tissue involvement. As it is a potentially aggressive lesion, patients should be carefully followed up after treatment because of the common presence of daughter cysts and a tendency for recurrence[20,24]. Rarely, a primary intraosseous squamous cell carcinoma can be derived from a KCOT, but metastasis have not been described[20,25,26].

The knowledge about clinicopathological behavior of ameloblastoma and KCOT may be helpful in the understanding of KAB. Some authors express their frustration about this entity based on the uncertain whether this tumor represents a KCOT with ameloblastoma foci or an ameloblastoma with KCOT areas, or perhaps a chimera[7,11]. There are few published KAB case reports. Up to now, in the English Language, twenty cases have been reported under the appellation KAB[1-17]. Among them, five presenting a variant called PKA[1,3-6]. This papilliferous variant was the first to be reported in 1970 by Pindborg as an unusual type of ameloblastoma with keratinization, consisting partly of keratinizing cysts and partly of tumor islands with a papilliferous appearance[1]. Six years later, Altini et al presented a similar lesion but without the papilliferous component, where numerous follicles of odontogenic epithelium were observed, many of which had undergone central cystic degeneration, were lined by a parakeratinized stratified squamous epithelium, and filled by desquamated parakeratotic cells[2]. Although some older reports bring to light lesions with

histopathological similarities[27,28], the terms PKA and KAB were initially used by these two authors. An additional case of PKA was described by Altini et al[3] in 1991, before the 1992 World Health Organization (WHO) classified it as other variations of ameloblastoma[19]. This classification made clear the difference between KAB and the acanthomatous pattern, where there is extensive squamous metaplasia, sometimes with keratin formation within the islands of tumor cells. Even though the acanthomatous type is often associated with keratinization, this is not the pathognomonic feature of this type of ameloblastoma. PKA and KAB are unique in that they show massive keratinization[17,19]. Acanthomatous changes and keratinization in ameloblastoma occur with different frequencies. While former is common, latter is rare[7].When the two described KAB are compared, the KAB “variant” indicates a lesion with a more extensive keratinization, while the PKA “variant” had to present microcysts lined by parakeratinized epithelium and contain keratin, while others showed a non-keratinized epithelium with a papilliferous pattern[19]. In 2005, although nine more cases had been reported (two papilliferous and seven non-papilliferous)[4-9], the latest edition of head and neck tumors book from WHO did not mentioned it as a particular entity. The only reference to it is as histopathologic differential diagnosis of primary intraosseous squamous cell carcinoma derived from KCOT[20]. Notwithstanding the fact that, by the literature it’s possible to have malignous transformation in KCOT, so as in ameloblastomas, the reports we have up to now on KAB do not show neither malignization nor metastasis[20,26,29]. Collini et al reported a PKA case with recurrence and suggested that, due to its biological behavior, it should be classified as a papillary ameloblastic carcinoma. This was based on the

presence of increased number of mitotic figures and extensive necrosis, what is usually considered a marker of malignancy[5]. However, Gardner observed that the diagnosis of ameloblastic carcinoma is clear if there are obvious dysplastic changes not observed in reported cases of KAB[30]. Whitt et al believe that the omission of this lesion in WHO's book most likely reflects an editorial decision to limit the classification to well-defined entities, rather than a retraction of prior nosology[11]. In this same work, Whitt et al was the first to try to label the thirteen cases previously published into four histological subtypes. Three of them[1,3,5] showed a "papilliferous histology"; two[2,9] a "simple histology"; five[7,8] a "simple histology with OKC-like features"; and three[4,10,11] a "complex histology" category[11]. Seven new cases were further reported, one with a papilliferous pattern[6] and six with a non-papilliferous pattern[12-17]. Regarding histopathological aspects, the twenty cases reported in the literature showed some points of special interest. Some authors[3,5,8,11] state that the first three published cases lacked typical ameloblastoma features. According to this statement, Norval et al[4] report would be the first to present a PKA, and Siar et al[7] the first to present a KAB. Norval et al reported a case as an unusual variant of KAB, once it shown only focal papilliferous content and under the author's belief that this lesion may be example of the acanthomatous ameloblastoma[4]. Kaku report misses some important aspects once it was published as an abstract[9]. Three reports[5,11,17] show presence of calcification into the lesion, what is not a common feature in ameloblastomas or KCOT. Additionally, root resorption was also shown in one report[6], what is a usual ameloblastoma finding but unusual in KCOT. None of the other nineteen cases have this presentation. Some authors[31,32,33] defends the existence of a separate entity named solid variant of

KCOT. It does not fit KCOT criteria and may be confused with KAB. A high rate of recurrence (23,8% approximately) is reported but in more than 50% of the previously reported cases the information about follow up is missing[1,2,4,6,7,9,10,12,13]. It can suggest that the recurrence rate could be even higher. Additionally it is difficult to correlate the recurrence with the histological pattern or the treatment for KAB. Among the five cases with reported recurrence, including the case here presented[5,8,16,17], the papilliferous pattern was only observed in one[5]. The treatment varied from enucleation or curettage and surgery for resection after initial enucleation or curettage, which had to be performed more than once to stop recurrences, in some cases. The presented case shows an unusual recurrence site, the right infratemporal fossa, after a hemimandibulectomy. This was also previously reported in another case[5] where the patient was submitted to two resections and died from lymphoma after a third recurrence was detected. A possible explanation for this site of recurrence is that the cells left in the TMJ surrounding area may have infiltrate the adjacent soft tissue.

Although we believe KAB and its papilliferous variant are not malignant lesions, care must be taken when a surgical plan is made. Its biological aggressive behavior and the high recurrence suggest that a more aggressive approach should be performed and the patient must be aware for the importance of clinical control. A resection with sufficient safety margins and histopathological analysis of surgical margins are highly recommended.

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TABLE

RN[1]	Author	A/G[2]	Location	Radiographic	Histopathologic	Treatment	Follow-up	Recurrence
1	Pindborg JJ	57/F	Right mandibular body and ramus	Multilocular	PKA[3]	unknown	unknown	unknown
2	Altini M et al	28/M	Anterior Maxilla	Multilocular	KAB[4]	Resection	unknown	unknown
3	Altini M et al	76/M	Right Mandible	Multilocular	PKA[3]	Resection	12 months	No
4	Norval EIG et al	26/F	Right Mandible	Irregular	PKA[3]	Resection	unknown	unknown
5	Collini P et al	62/M	Right mandibular ramus and condyle	Irregular with calcifications	PKA[3]	Resection (1st and 2nd surgeries)	39 months	Yes
6	Mohanty N et al	46/M	Right Posterior Mandible	Multilocular	PKA[3]	unknown	unknown	unknown
7	Siar CH et al	39/F	Left Anterior Mandible	Unilocular	KAB[4]	Enucleation	unknown	unknown
7	Siar CH et al	35/F	Right Maxilla	Ground glass	KAB[4]	unknown	unknown	unknown
7	Siar CH et al	35/M	Left Mandible	unknown	KAB[4]	Resection	unknown	unknown
7	Siar CH et al	30/M	Anterior Mandible	Multilocular	KAB[4]	Resection	unknown	unknown
8	Said-al-Naief NA et al	26/M	Right Posterior Maxilla	Unilocular	KAB[4]	Curettage (1st) Resection (2nd surgery)	6 months	Yes
9	Kaku T	35/M	Right body of Mandible	Unilocular	KAB[4]	unknown	unknown	unknown
10	Takeda Y et al	76/M	Left body of Mandible	Multilocular	KAB[4]	Resection	unknown	unknown
11	Whitt JC et al	45/M	Anterior Maxilla	Unilocular with calcifications	KAB[4]	Curettage	10 months	No
12	Adeyemi BF et al	38/M	Right Posterior Mandible	Multilocular	KAB[4]	Resection	unknown	unknown
13	Sisto JM et al	35/F	Right Posterior Mandible	Multilocular	KAB[4]	Resection	unknown	unknown
14	Ketabi MA et al	21/F	Right Anterior Mandible	Unilocular	KAB[4]	Enucleation	12 months	No
15	Raj V et al	22/F	Right Posterior Mandible	Unilocular	KAB[4]	Resection	24 months	No
16	Palaskar SJ et al	65/F	Anterior Mandible	Unilocular	KAB[4]	Enucleation (1st) Resection (2nd surgery)	4 months	Yes
17	Lee C et al	56/M	Right Maxilla	Irregular with calcifications	KAB[4]	Enucleation (1st-4th) Resection (5th-7th surgeries)	40 months	Yes
*	Netto R et al	46/F	Right infratemporal fossa	Solid with calcifications	KAB[4]	Resection (1st and 2nd surgeries)	36 months	Yes

Table I – Summary of clinical and radiologic features of previously reported cases of PKA and KAB, including the present case

FIGURES

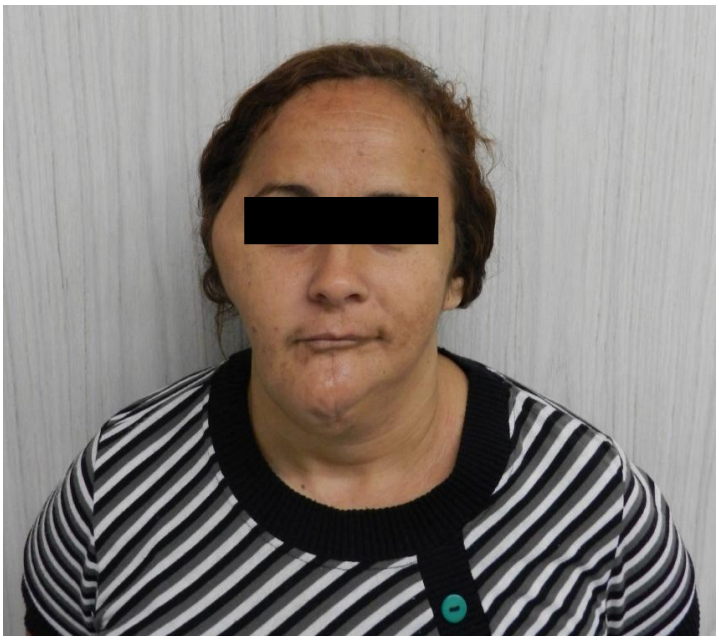

Figure 1 - Extra-oral examination: Firm well-circumscribed swelling in right infratemporal region measuring about 7 cm on its longest axis and surgical scar in the middle mandibular region

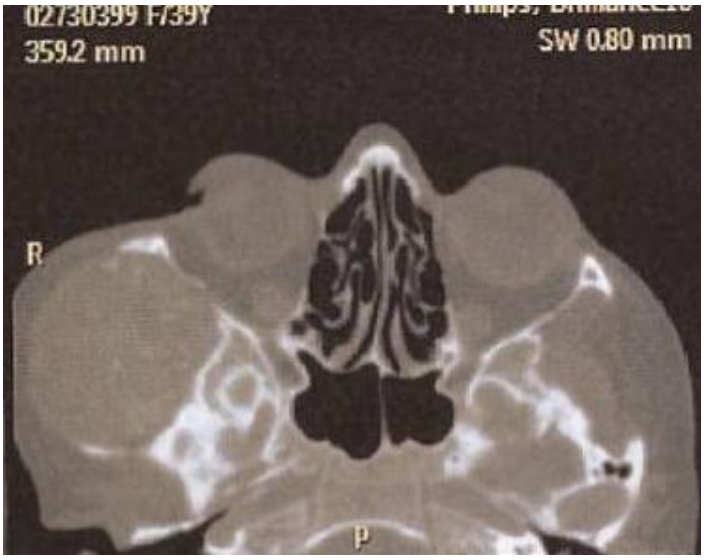

Figure 2 – CT Scan which exhibited an expansile, solid lesion, with internal calcification in the right infratemporal fossa

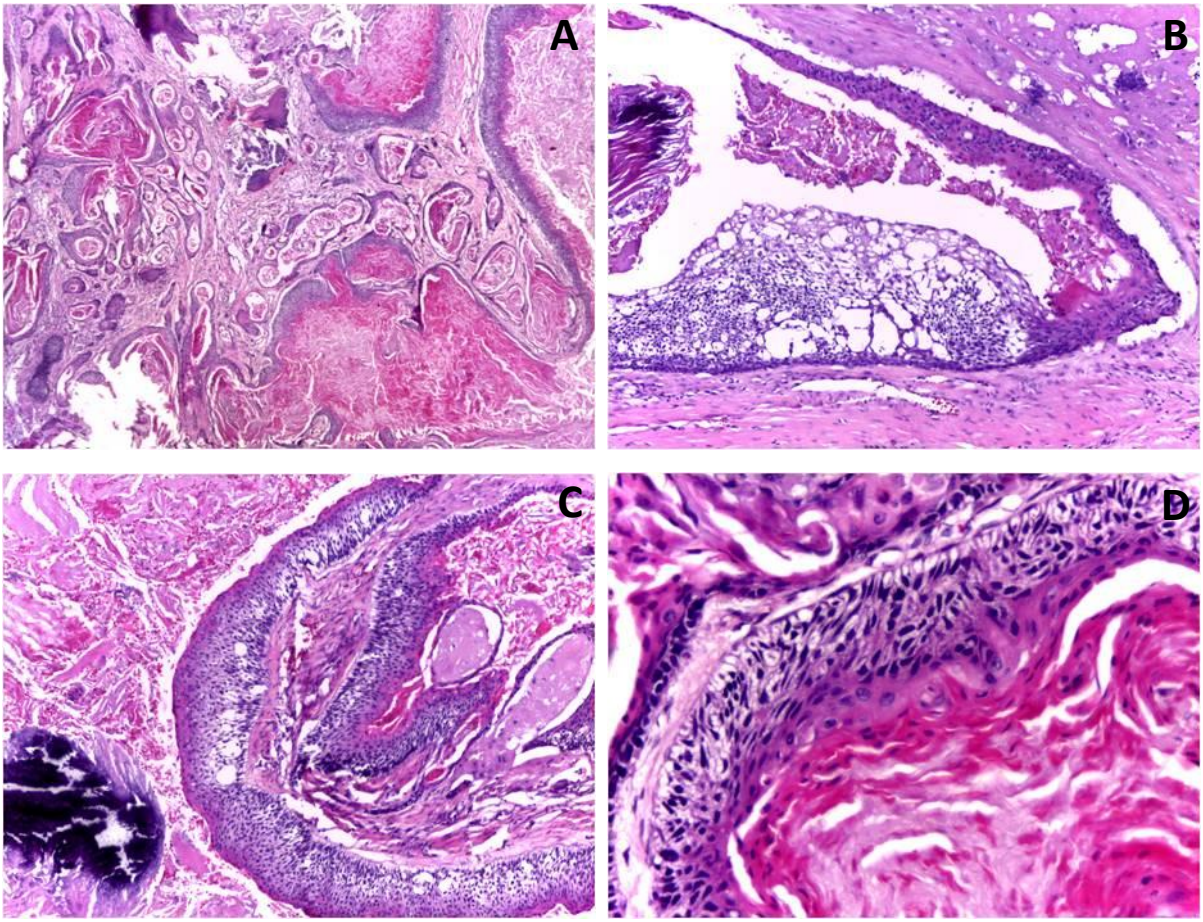

Figure 3 – Histopatological features of Keratoameloblastoma (infratemporal lesion). **A,** Cystic proliferation of odontogenic epithelium. Keratin and cellular debris are observed filling the cystic lumen. The lining epithelium presentes variable thickness and multiple cell layers. Some cystic areas are similar to ameloblastoma and other to odontogenic keratocystic tumor (OKT) (HE, 40x). **B and C,** The epitheluim shows a basal cell layer with palisade colunar cells with polarized nuclei. The intermidiate layer shows cells ressembling stellate reticulum of enamel organ, but some areas are similar to OKT. The superficial cell layer presents keratinized cells. A calcification area can be observed into the surrounding connective tissue (HE, 100x). **D,** High power view showing the linning cystic epithelium organized similar to an ameloblastoma island. An intense keratinization is observed in the lumen (HE, 400x).

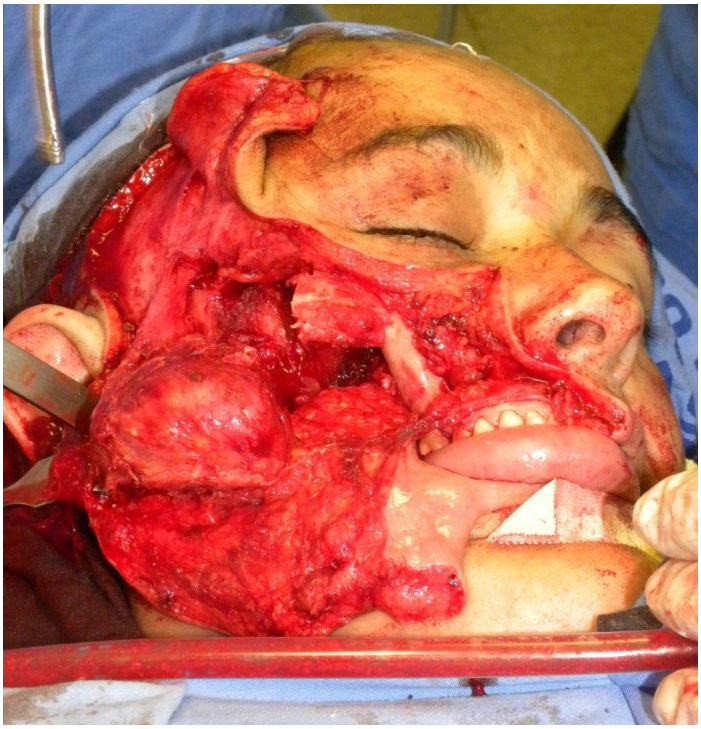

Figure 4 – Total removal of the infratemporal lesion with safety margins

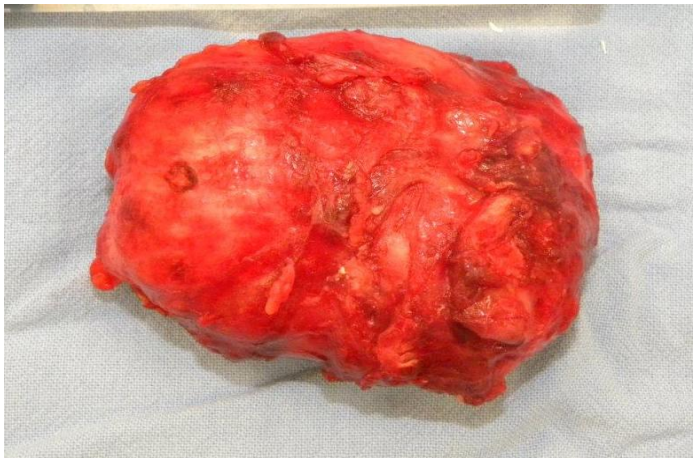

Figure 5 – Surgical specimen

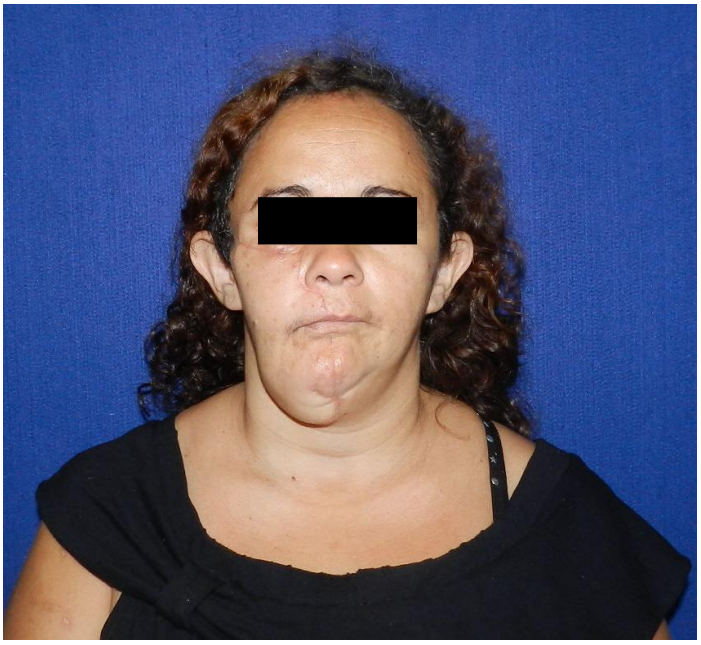

Figure 6 - Clinical follow-up of 36 months

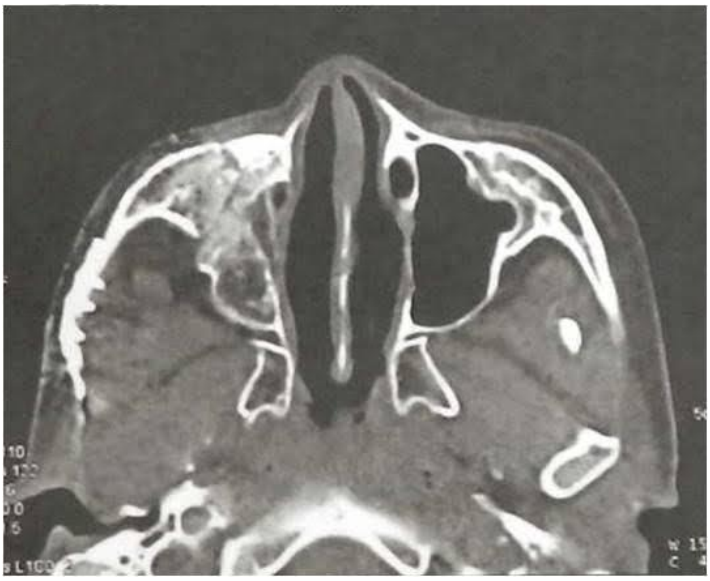

Figure 7 - Radiographic follow-up of 36 months