ABSTRACT
Introduction: the keratocystic odontogenic tumor (KOT) is a relatively common oral and maxillofacial lesion that arises from rests of dental lamina. It has an aggressive behavior including high rates of recurrence, rapid growth, and extension into adjacent tissues. Various treatment modalities, and thus differing recurrence rates after treatment, have been reported. Due to the very thin and friable lining, characteristic of the tumor, enucleation can be difficult undertaking and, for this reason it is associated with the highest recurrence rates.

Aim: we describe a case of a large KOT in the mandibular body, where due to the presence of an unusual thick lining, removal of the tumor as a single piece was successful.

Conclusion: this case shows that large KOT can be treated in a conservative approach. Due to the possible recurrences many years after initial treatment, a life-long follow-up schedule is mandatory.

Keywords: Keratocyst; Lining; Oral Surgery.

RESUMO
Introdução: o tumor odontogênico ceratocístico (TOC) é uma lesão oral e maxilofacial relativamente comum que surge de restos da lâmina dental. Ele tem um comportamento agressivo, incluindo altas taxas de recorrência, crescimento rápido e extensão para os tecidos adjacentes. Várias modalidades de tratamento e, assim, diferentes taxas de recorrência após o tratamento, foram relatadas. Devido ao revestimento muito fino e fráível, característica do tumor, a enucleação pode ser difícil e, por esta razão ele é associado com as maiores taxas de recorrência.

Objetivo: o presente trabalho descreve um caso de um grande TOC no corpo mandibular, onde, devido à presença de um revestimento de espessura incomum, a remoção do tumor como uma peça única foi bem sucedida.

Conclusão: o presente caso demonstra que TOC de grandes proporções podem ser tratados de maneira conservadora. Devido à possibilidade de recidiva em muitos anos após o tratamento inicial, acompanhamento a longo prazo é obrigatório.

Palavras-chave: Tumor Odontogênico Queratocístico; Revestimento; Cirurgia Oral.
INTRODUCTION

The keratocystic odontogenic tumor (KOT) is defined as a benign uni or multicystic, intraosseous tumor of odontogenic origin with a characteristic lining of parakeratinized stratified squamous epithelium and potential for aggressive, infiltrative behavior1. This tumor, previously considered an odontogenic cyst was first reported by Philipsen in 1956 and attracted interest because of its specific histopathological features and its high recurrence rate2-3.

Keratocystic odontogenic tumor occurs over a wide age range, and cases have been recorded as early as the first decade and as late as the ninth4. In most series there has been a pronounced peak frequency in the second and third decades, with figures ranging from 40% to 60% of patients in this age group. The tumor usually is found more frequently in men than in women5-6.

Several studies concerning the clinical presentation of the KOTs have been conducted and most of them have shown that the mandible is involved more often than the maxilla, with higher incidence in the posterior body and ascending ramus3,5-8.

Patients with KOTs may complain of pain, swelling, or discharge. However, some are unaware of the lesion until they develop pathologic fractures; this occurs because KOT tend to extend into the medullary cavity and clinically observable expansion of the bone occurs late5. The aggressive behavior of the tumor penetrating the surrounding soft tissues9, base of skull10, orbit and infratemporal fossa11 have been reported. Sometimes KOT occurs in association with basal cell nevus syndrome, and frequently there are multiple KOTs12.

Radiographically, KOT appears as a unilocular or multilocular radiolucency with a scalloped contour3. Unilocular KOT can be located periapically, simulating periapical cyst7 and can also present as a small radiolucency between teeth simulating a lateral periodontal cyst6. Displacement of unerupted teeth and inferior and superior displacement of the mandibular canal can be observed8.

The typical histologic features of the KOT include a thin, uniform layer of epithelium with little or no evidence of rete ridges, a well-defined basal cell layer with palisading cuboidal or columnar cells, and a corrugated, keratinizing luminal surface that is primarily parakeratinized. The presence of small “daughter” or “satellite” cysts may be observed in the connective tissue wall of the KOT13. The tumors linings are thin and fragile, being difficult to enucleate in one piece5,14.

In the literature there is a controversy regarding the treatment of KOT: some surgeons advocate conservative therapies whereas others are in favor of an aggressive treatment15. The goal is to choose the treatment modality that carries the lowest possible risk of recurrence and the least morbidity, while still eradicating the lesion9.

The present article presents a case of KOT in the right mandibular body of a male patient where due to the presence of an unusual very thick lining what should be the incisional biopsy turned to be the complete enucleation of the tumor.

CASE REPORT

A 51 year-old male presented to the Stomatology Service of Pontificia Universidade Católica do Paraná, Curitiba/PR, Brazil for evaluation of a gradually enlarging hard mass in his right cheek. He reported that the swelling had gradually increased in size over a period of 4 months and at that time he was stating severe
discomfort due to the bad taste that was occurring in consequence of persistant pus drainage inside his mouth. He denied any neurosensory deficit associated with right inferior alveolar nerve. His medical history was unremarkable except for accentuated tobacco use.

Clinical evaluation showed a swelling along all the right mandibular body, with the presence of small discharge of purulent fluid from an opening in the oral mucosa in the posterior area of the alveolar ridge (Fig. 1). Patient presented only his anterior teeth, from left canine to right first pre-molar, and all of them had severe carious lesions.

The panoramic radiograph showed a unilocular radiolucent lesion, irregularly corticated with scalloping of the cortex, extending from the molar to the cuspid area (Fig. 2).

After obtaining an informed consent, the patient was treated with amoxycilin for seven days to control the infection and then the teeth were all removed. An
incisional biopsy was scheduled fifteen days later. Initially, a needle aspiration biopsy was performed and no fluid was aspirated. During the incisional biopsy as soon as the lining was reached, keratin discharge from the lesion was observed (Fig. 3). Unexpectedly, the cyst lining was very thick allowing the entire lesion enucleation (Fig. 4). After that, peripheral ostectomy of the area was performed. Postoperative evaluation of the patient did not show any neurological damage of the inferior alveolar nerve.

Figure 3. Transoperative view: As soon the lining was reached, keratin discharge was observed.

Figure 4. Transoperative view showing the very thick lining.
Histologic examination showed a cyst lined by parakeratotic stratified squamous epithelium with variations in thickness, approximately 2-6 cell layers (Fig. 5). The fibrous connective tissue wall contained sparse inflammatory cells. The pathologic diagnosis was KOT.

Figure 5. Histopathologic examination: thin and regular parakeratinized stratified squamous epithelium without rete pegs and exhibiting the flat interface between the epithelium and connective tissue. There is an intense descamation of keratin in the lumen of the cyst.

The patient returns every six months for follow-up and now at two-year follow-up no clinical or radiological evidence of recurrence was observed (Fig. 6). He’s now using an inferior total prosthesis, and after the completion of five-year follow-up, implants are planned to give him a better masticatory function.

Figure 6. Panoramic radiograph of two-year follow-up showing complete bone neoformation with no signs of recurrence.
DISCUSSION

KOTs have been reported to occur most commonly in the second and third decade of life but may be diagnosed in people of any age. Van Rensburg et al. found that the age at the diagnosis was higher than the usually reported; most patients were in their sixth and fourth decade of life. In another study with 106 KOTs, the incidence was also highest in the older age groups, and the peak age incidence was approximately a decade younger in women than in men, a factor also discussed in a study of 430 cases of KOTs in the Northwestern USA. A male predominance has been reported for KOT in previous studies. In contrast, Maurette et al. found a male to female ratio of 1:2.1 in a series of 30 Brazilian cases. In other studies, the authors found that both genders were affected almost evenly, with no predilection for males or females.

The tumor involves the mandible far more frequently than the maxilla. Shear in a study of 125 KOTs, found 94 occurring in the mandible; another study with 183 KOTs, revealed that 70.5% were also in the mandible. The tumor can occur in any part of the mandible, but the majority arises in the angle and ascending ramus.

Even being considered a benign tumor, some of the characteristics of KOT make this pathology extremely serious. KOT tend to recur, can reach considerable dimensions, and can arise close to delicate and important anatomical structures such as inferior alveolar nerve. All these characteristics had suggested, in the past, that an aggressive surgical approach should be followed in order to completely eradicate the lesion.

The treatment of KOT remains controversial and various surgical modalities have evolved in an attempt to decrease the recurrence rate, including enucleation, curettage, marsupialization, decompression, enucleation and treatment of the bony defect with Carnoy's solution, liquid nitrogen cryotherapy and resection with or without loss of jaw continuity. Patient characteristics are important determinants, including the patient's general health status, age, reliability for follow-up, and whether the patient has basal cell nevus syndrome.

Resection has proved to be the most predictable treatment, but the morbidity associated with reconstructing continuity defects seems unwarranted in most instances for a disease with no real metastatic potential, although its aggressive nature. As long as more conservative treatment is used, recurrences should be anticipated. In a systematic review of the treatment and prognosis of KOT, the authors stated that for a routine KOT in a person who is likely to return for follow-up treatment, Carnoy's solution appeared to be the least invasive procedure with the lowest recurrence rate; for very large tumors, decompression followed by enucleation will also have a low recurrence rate. For patients who are unlikely to return for follow-up, the authors recommend resection of the lesion.

In the present case the initial treatment planning was an incisional biopsy followed by decompression and definitive removal by enucleation and curettage after the tumor volume reduction was reached. This planning was changed during the biopsy due to the unusual thick lining of the KOT that allowed the enucleation of the tumor in a single piece; this thickening probably occurred due to the chronic infection. It's well established in the literature that enucleation has the highest recurrence rates in the treatment of KOTs and this is partially explained by the characteristic thin and friable lining of the KOT that makes removal of the tumor as a single piece difficult.
Keratocystic odontogenic tumor: case report

Simple enucleation is no longer advocated as an appropriate method of treatment for KOT and, minimally, the recommendation is for curettage after tumor removal. Recently endoscopy has been used to assist the enucleation and curettage of a voluminous KOT of the mandible with good results. The endoscopic assistance allows the surgeon to explore accurately the operative field and the areas of difficult access, improving the complete removal of the tumor.

Due to possible recurrences many years after the initial treatment, long-term follow-up is mandatory. Stoelinga suggests a life-long follow-up schedule, regardless of the treatment given, every year for the first 5 years and every 2 years thereafter. As enucleation and curettage presents higher recurrence rates than other treatment modalities, even though studies have shown the enucleation of the tumor in a single piece can reduce this rate, our patient will return for follow-up every six months during the first 5 years and every year thereafter.

CONCLUSION

- In the light of previous clinical trials, this case report outlines that large KOT might be treated with conservative approach. In any case, long-term post-surgical follow-up is mandatory due to its aggressive nature and possible recurrences many years after the initial treatment.

REFERENCES

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