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CASE REPORT

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Solid variant of odontogenic keratocyst in a 22-year-old man: Report of a case

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Key Clinical Message

In conclusion, we can focus on histologic features such as stellate reticulum, reverse polarization of basal cell layer nuclei, and luminal lamellated keratinization as distinguishing factors of ameloblastoma and SKCO. If there is any clinically doubt, molecular testing could be helpful.

Abstract

"Solid odontogenic keratocyst" is a rare variant of odontogenic keratocyst, which usually involves mandible. This case was presented as a unique variant of odontogenic keratocyst in an unusual site of left maxilla with extension to the maxillary sinus.

KEYWORDS

keratocyst, odontogenic cyst, solid variant of odontogenic cyst

1 | INTRODUCTION

Odontogenic keratocyst (OKC), which was first described by Philipsen, is a developmental odontogenic cyst with characteristic features including an epithelial lining of rather uniform thickness, palisaded basal cells, and a thin corrugated layer of parakeratin.[1,2]

Odontogenic keratocyst shows a high recurrence rate, which can be primarily attributed to the friable lining, which makes it difficult to be completely removed and additionally high proliferative activity of epithelial lining and extension to bony spaces, which makes complete enucleation difficult.

Although OKC was introduced as a neoplastic lesion in WHO Blue Book 2005, the lesion is moved back into the cyst categories in 2017 WHO classification.[3] The first description of "Solid odontogenic keratocyst," (SOKC) which is a rare variant of odontogenic keratocysts, was reported by Ide et al. in 2003.[1] The invasive and destructive-growth behavior along with histopathologic features reflects that solid lesions can be regarded as a true neoplasia.[2,3]

Some investigators believe that solid odontogenic keratocyst and ameloblastoma, especially keratoameloblastoma, have common histologic features, but it may need more case reports to be proven.[4]

The aim of this case is to show SOKC as a unique variant of odontogenic keratocyst, located in an unusual site of the jaws that should be separated based on its histological features and should not be mistaken with other intraosseous tumors, such as keratoameloblastoma.

2 | CASE REPORT

In October 2020, a 22-year-old man was referred to Head and Neck Surgery Department of Amir-Alam hospital with left facial swelling for 2-year duration. Also, the patient reported a history of trauma to the left facial area.

The extraoral examination of the patient revealed mild swelling in the left infraorbital area with normal-looking skin.

Also, intraoral examination showed mild and firm swelling in the left side of maxilla with purple discoloration of overlying gingiva and purulent discharge led to halitosis.

2.1 | Radiologic findings

Cone beam computed tomography (CBCT) images through the whole maxilla showed expansile multilocular lesion measuring 40 × 57 × 60 mm in the inferior aspect along with some wispy septation. The lesion has expanded to anterolateral and posterolateral walls of maxillary sinus as well as moderate expansion of medial wall of maxillary sinus toward the nasal cavity. Additionally, displacement of molar teeth was observed (Figures 1 and 2).

The differential diagnosis was made as "calcifying odontogenic cyst", "central giant cell granuloma", and less commonly "calcified odontogenic fibroma" and "ameloblastoma." Then, the patient was referred for an incisional biopsy.

2.2 | Microscopic findings

Microscopic diagnosis of the incisional biopsy was reported as an "infiltrative odontogenic tumor arising in odontogenic keratocyst." Additionally, SOKC and keratoameloblastoma are suggested as the definite diagnosis that deferred to microscopic assessment of the excised lesion.

The resected specimen was embedded totally, and histologic examination of prepared slides showed solid components with clefting spaces (Figure 3). Also, the lesion consisted of some small and large cysts that resembling odontogenic keratocyst. The epithelial lining of the cyst is characterized by palisaded hyperchromatic basal cell layer in which some areas of the luminal surface showed a corrugated parakeratin formation. The lumen of the cyst contained prominent keratinous material.

We found foci of solid islands arranged back to back within the underlying fibrotic stroma (Figure 4). High-power magnification of the mentioned islands showed bland-looking cells with low mitotic rate and basal cells arranged in a palisading pattern, surrounded by chronic inflammatory cells (Figure 5). No evidence of epithelial dysplasia, stellate reticulum-like change, and reverse polarity/ apical vacuolization is seen in epithelial lining.

2.3 | Follow-up

A postoperative panoramic radiograph and CT scan showed no radiographic tumor remnants. No clinical evidence of recurrent disease was found during 9 months after definite surgery.

3 | DISCUSSION

Odontogenic keratocyst is a developmental odontogenic cyst originating from dental lamina cells.

Solid variant of odontogenic keratocyst is extremely rare, and because of few reported cases of this lesion, there are no enough data in the literature.[3,5,6]

First, Ide et al. described SOKC in 2003.[1] The information obtained from the published cases such as high rate of recurrence and potentially destructive growth illustrated

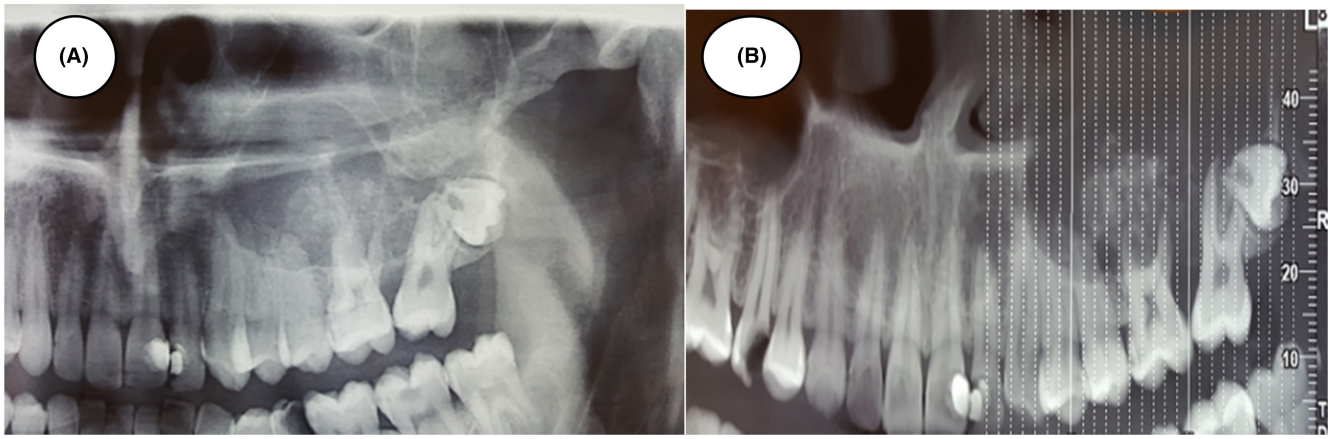

FIGURE 1 Preoperative panoramic radiograph. (A) Well-defined expansile lesion in the peri-apical and inter-radicular areas of the upper right molars and peri-molars. (B) Accumulation of multiple calcifications in inferior aspect.

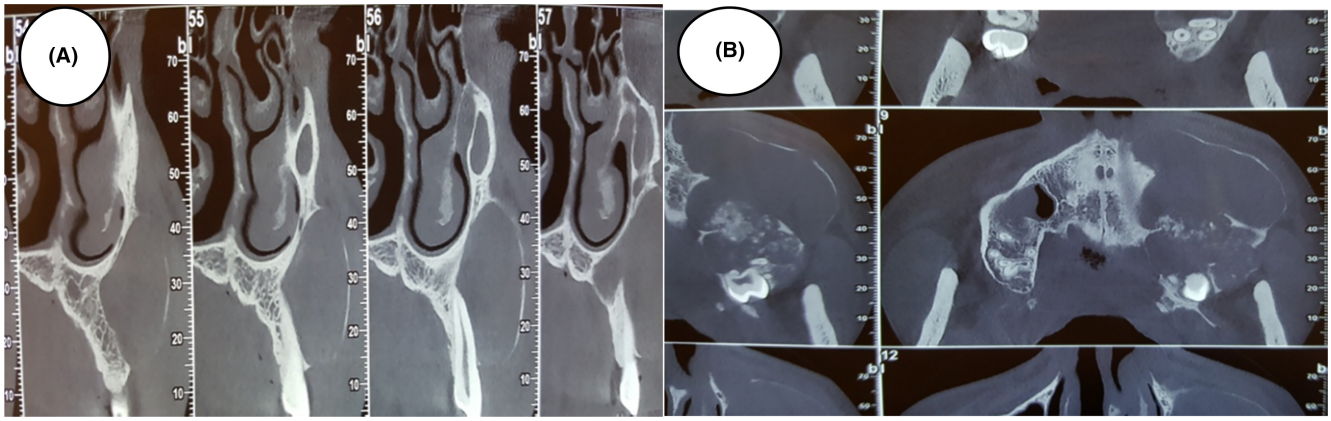

FIGURE 2 (A) Lesion has expanded antrolateral and posterolateral wall of maxillary sinus. (B) Expansion of medial wall of maxillary sinus toward nasal cavity.

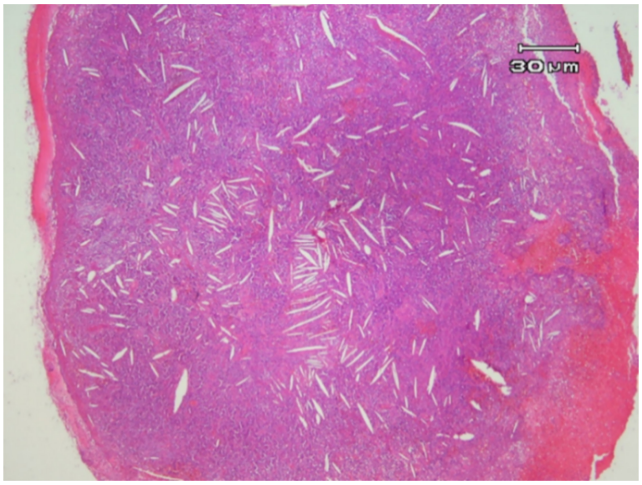

FIGURE 3 Low-power examination shows solid component with cholesterol clefts.

that this variant may be neoplastic and it is different from others.[3,5] Following this, Vered et al. in 2004 and Kawano in 2013 insisted that these variants show aggressive nature.[1,3] On the contrary, some may claim that although there are scant numbers of this variant, in some cases of them, they show nonaggressive nature. Despite genetic and behavioral evidence, some groups deny the neoplastic nature of these lesions. Or at least some of the existing evidence indicates that not all cases of solid KCOT would be uniformly more aggressive than the cystic variant.[1,7] All in all, in 2007, Daley et al. found that radiographically well-defined margin lesions usually show nonaggressive behavior.[3,7] Presumably, the neoplastic OKC family has a broad histopathologic spectrum from benign cystic form to cytologically malignant type and solid-cystic form occupying an intermediate position, may be nearer to the benign end.[8] We could find only nine case of SKCOT in the English literature until 2022, and they show predilection of mandible (7/9) over the maxilla. Most of them were radiolucent multilocular lesions, and three cases were ill-defined.[9]

On microscopic examinations, they contain solid and cystic components infiltrating into the bone lined by parakeratotic stratified squamous epithelium of a uniform thickness (2–10 cells thick) with palisaded nuclei in reverse polarity without any dysplastic changes. The surface epithelium is corrugated. The lumens of cysts are filled with layers of keratin, desquamated squames, and/or necrotic material.[2,9,10] The supporting connective tissue may contain varying degrees of chronic inflammatory reactions with giant cells and thick collagen fibers. These fibers demonstrate polarization colors of greenish-yellow range.[2]

Histologically, SKCOT is similar to the keratoameloblastoma.[9]

In WHO classification of tumors and cysts, there are odontogenic cysts with solid variants in one hand, and on the other hand, there are solid odontogenic neoplasms with cystic variant such as ameloblastoma.[2]

Ameloblastoma is a benign but locally invasive neoplasm. Two rare histologic variants of ameloblastoma are papilliferous keratoameloblastoma and keratoameloblastoma.[11] The term "keratoameloblastoma" was first described by Pindborg in 1970, and the first confirmed case was in 1976 by Altini et al.[12] Later, some investigators better defined this entity in 1993 focusing that acanthomatous changes in ameloblastoma are common, while keratinization is rare, supporting the term keratoameloblastoma (KA).[8,12] The 2017 WHO classification of benign odontogenic tumors loosely defined it as well-defined entities 12 and defines as "extensive keratinization in ameloblastoma."[2] It contains reverse polarizing palisading nuclei in the basal layer without any parakeratinization.[1,11] Some authors demonstrate that KA had a tendency to mandible over the maxilla, especially

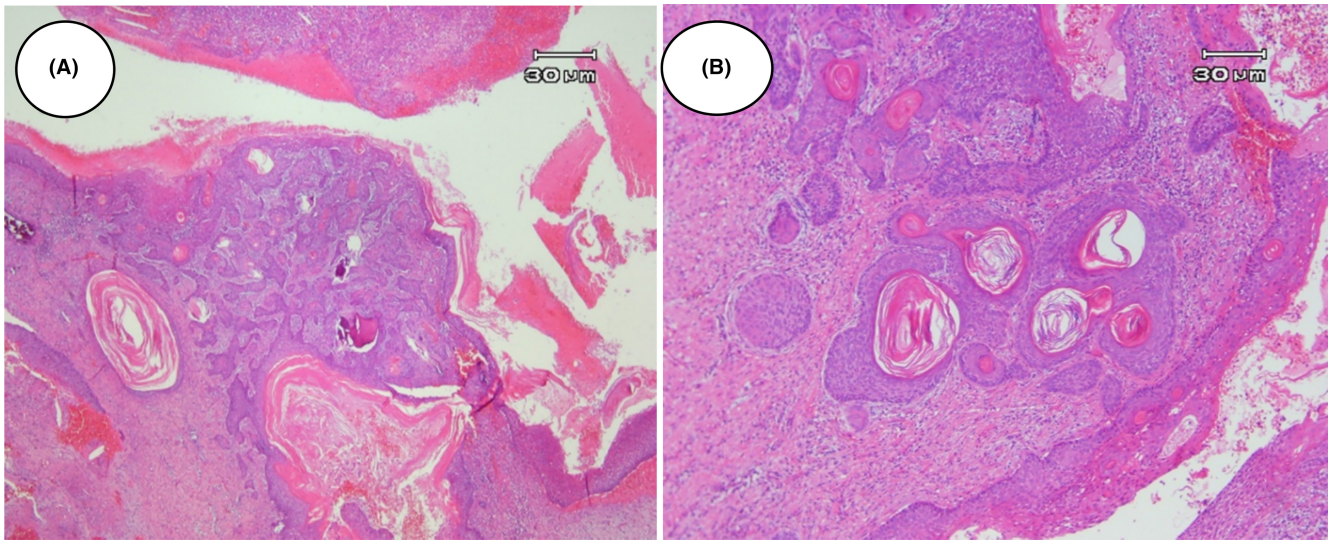

FIGURE 4 (A) Large cyst with luminal parakeratin layer demonstrating solid component of odontogenic epithelium packed in stroma, adjacent to cyst. (B) Several satellite cyst varying in size infiltraring adjacent stroma. The wall of cyst lined by parakeratotic stratified squamous epithelium with corrugated surface and palisaded appearance of basal cells (hematoxylin–eosin stain: original magnification ×4 to ×10).

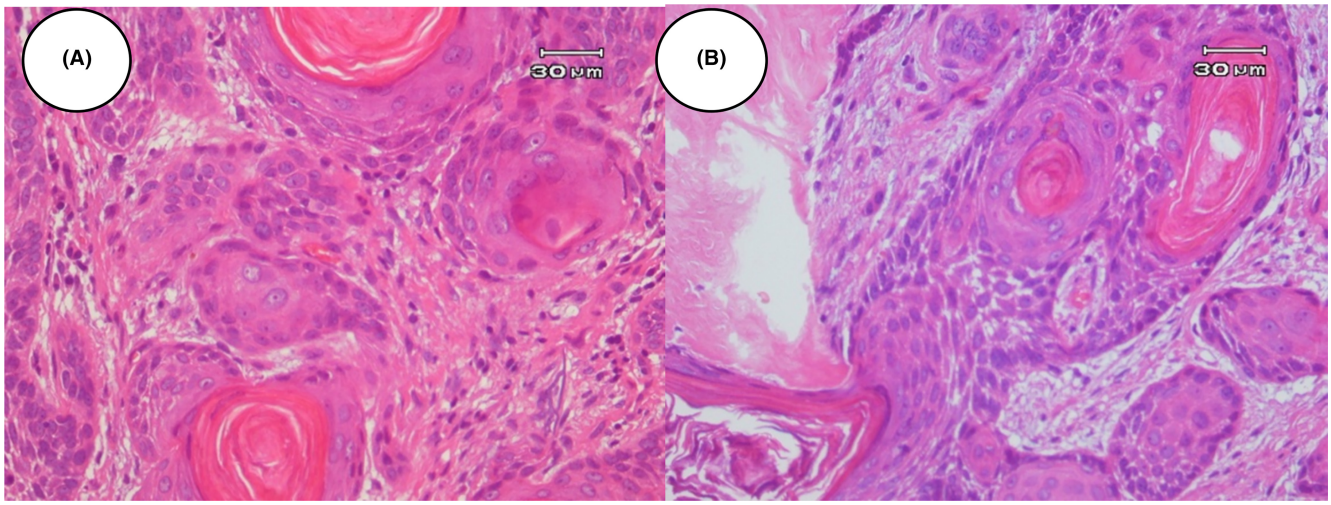

FIGURE 5 Surrounded by few inflammatory cells (hematoxylin–eosin stain: original magnification ×40) (A) and high-power examination of solid islands of Figure 4 illustrates bland-looking epithelial cells with low mitotic rate (B).

multilocular cases, and they will often show signs of aggressiveness such as cortical destruction, distinguishing from OKC.[12]

Keratoameloblastoma demonstrates a cyst lined by multilayered epithelium comprised of tall columnar cells with palisading and subnuclear vacuolization. Several solid keratin epithelial follicles lined in fibrous connective stroma with varying degrees of chronic inflammation.[11,12] Acanthomatous ameloblastoma demonstrates squamous metaplasia with keratinization and calcification keratin pearls in the center of ameloblastoma follicles, while the keratinization of SKCOT is located on the surface of stratified squamous epithelium lining.[6] Some cases show scattered dystrophic calcification and foreign body giant cell response.[12]

Palisading of the basal cell is a common feature of both KCOT and ameloblastoma. But stellate reticulum and reverse polarization of basal cell layer nuclei are the criteria of ameloblastoma and are found at least focally in most ameloblastoma. So, some may argue that the term ameloblastoma should be kept for cases that fulfill these criteria.[1] Some theories suggest that SKCOT should be regarded as ameloblastoma.[4] They believe the epithelium of solid variant of KCOT provides suitable ground where ameloblastoma can arise.[3,9] Indeed, the epithelial lining of OKC shares both phenotype and gene profiles

with ameloblastoma lining cells.[4] In fact, the presence of solid epithelial follicles containing keratin plugs is the evidence that shows ameloblastoma can develop on SKCOT.[3] Amir Shuster et al. believed that diagnosis of KCOT is simple for expert pathologists, but reaching a diagnosis of solid KCOT could be more challenging.[1] Given the fact that both lesions share some common histologic features such as a solid mass, keratin-packed cyst, and displaying hyperchromatic basal cells with palisading, making a definite diagnosis could be challenging. It has also been reported that the columnar basal layer of the lining epithelium of OKC may exhibit polarization making a distinction from ameloblastic epithelium even more difficult.[1,3] So, in some cases, we should better report the final diagnosis as a solid variant of OKC with ameloblastic change.[3]

Geng et al. focused on histologic features and luminal lamellate keratinization as distinguishing factors.[12] While Zhang et al. demonstrated molecular testing could be helpful, when available. Only single mutation of PTCH1 was found between nine cases and BRAF mutation was negative in all nine cases of SOCK, a mutation found in up to 90% of KA.[12] Following this, it was found that calretinin staining is positive in stellate reticulum-like epithelium and ameloblastoma while none of SKCOT showed positive stain.[9,10,12]

Primary intraosseous carcinoma (PIOC) resembling keratinizing squamous cell carcinoma (SCC) is another keratinizing malignant tumor of oral cavity. These may arise de novo, from odontogenic cysts. Keratinizing squamous cell carcinoma is an aggressive malignant lesion with bone destruction. Its histologic appearance includes cores lined by well-differentiated squamous epithelium with varying degrees of dysplasia, and high mitotic activities, allowing distinction from SKCOT and KA.[9,12]

The distinguishing features of keratinizing lesions:

Keratinizing lesions	Stellate reticulum	reverse polarization of basal cell layer nuclei	palisading of the basal cell	corrugated parakeratin	Dysplasia
Solid odontogenic keratocyst	−	−	+	+	−
Keratoameloblastoma	+	+	+	−	−
Squamous cell carcinoma	−	−	−	−	+

Despite genetic and behavioral evidence, some pathologists refused to accept the neoplastic nature of OKC because of the traditional concept of non-neoplastic character of a cyst.[7] We are believed that the number of reported cases of SKCOT is scant to exact evaluation of histological, radiological, and biologic behavior of SKCOT. Accurate evaluation of these spectrum depends on report of further studies.

AUTHOR CONTRIBUTIONS

Pouyan Aminishakib: Data curation. **Azadeh Jafarai:** Conceptualization. **Tahereh Padeganeh:** Resources.

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None.

CONFLICT OF INTEREST STATEMENT

None declared.

DATA AVAILABILITY STATEMENT

The datasets supporting the conclusion of this article are included within the article.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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REFERENCES

1. Shuster A, Shlomi B, Reiser V, Kaplan I. Solid keratocystic odontogenic tumor—report of a nonaggressive case. *J Oral Maxillofac Surg*. 2012;70(4):865-870.
2. Vered M, Buchner A, Dayan D, Shteif M, Laurian A. Solid variant of odontogenic keratocyst. *J Oral Pathol Med*. 2004;33(2):125-128.
3. Kahraman D, Gunhan O, Celasun B. A series of 240 odontogenic keratocysts: should we continue to use the terminology of 'keratocystic odontogenic tumour' for the solid variant of odontogenic keratocyst? *J Cranio-Maxillofacial Surg*. 2018;46(6):942-946.
4. Ide F, Ito Y, Muramatsu T, Saito I, Abiko Y. Histogenetic relations between keratoameloblastoma and solid variant of odontogenic keratocyst. *Oral Surg Oral Med Oral Pathol Oral Radiol*. 2012;114(6):812-813.
5. Kawano K, Okamura K, Kashima K, et al. Solid variant of keratocystic odontogenic tumor of the mandible: report of a case with a clear cell component and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol*. 2013;116(5):e393-e398.
6. Geng N, Lv D, Chen Q, et al. Solid variant of keratocystic odontogenic tumor with ameloblastomatous transformation: a case report and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol*. 2012;114(2):223-229.
7. Daley TD, Multari J, Darling MR. A case report of a solid keratocystic odontogenic tumor: is it the missing link? *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 2006;103(4):512-515.

8. Ide F, Mishima K, Saito I. Solid-cystic tumor variant of odontogenic keratocyst: an aggressive but benign lesion simulating keratoameloblastoma. *Virchows Arch*. 2003;442:501-503.
9. Zhang R, Yang J, Zhang J, Hong Y, Xie X, Li T. Should the solid variant of odontogenic keratocyst and keratoameloblastoma be classified as the same entity? A clinicopathological analysis of nine cases and a review of the literature. *Pathology*. 2021;53(4):478-486.
10. Santana DCP, de Aquino Xavier FC, dos Santos JN, Henriques ÁCG. Is the solid variant of odontogenic keratocyst the neoplastic counterpart of the lesion? *Oral Dis*. 2022;28(8):2215-2218.
11. Said-Al-Naief NAH, Lumerman H, Ramer M, et al. Keratoameloblastoma of the maxilla: a case report and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endodontol*. 1997;84(5):535-539.
12. Robinson L, Smit C, Fonseca FP, et al. Keratoameloblastoma: a report of seven new cases and review of literature. *Head Neck Pathol*. 2022;16(4):1103-1113.

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