RESEARCH

Orthokeratinized odontogenic cyst in a Hong Kong community: the clinical and radiological features

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Objectives: The aim was to evaluate the principal clinical and conventional radiological features of a consecutive series of cases of orthokeratinized odontogenic cyst (OOC) affecting a Hong Kong Chinese community and to determine the outcome by follow-up.

Methods: All cases were accompanied by appropriate radiography and were confirmed by histopathology.

Results: The clinical and conventional radiological presentations, differential diagnoses and outcomes of follow-up of five consecutive OOCs were reviewed. There were two males and three females. All affected the posterior sextant. The mean age at first presentation was 33.5 years. The mean of their period of prior awareness was 0.11 years. Swelling was the most frequent presenting symptom. All presented as well-defined corticated radiolucencies; three were unilocular and two were multilocular and all displayed expansion. This resulted in displacement and erosion of the lower border of the mandible in one case and the downward displacement past the lower border of a lateral cortex in two others. The inferior dental canal in each mandibular case exhibited both displacement and absence. The antrum was affected in a sole maxillary case. Four patients were followed up for a mean of 8.5 years. The fifth patient discharged himself shortly after surgery. No lesions recurred.

Conclusions: OOCs in this community displayed an expansile character, but did not recur after moderately long follow-up. The time between the prior awareness of their disease and their presentation for diagnosis and treatment was, so far, the shortest for any lesion affecting the jaw in this Hong Kong Chinese community.

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Introduction

The orthokeratinized odontogenic cyst (OOC)¹ was first clearly identified as a distinct entity by Wright in 1981.² It was also called the orthokeratinized variant of the odontogenic keratocyst². Due to its different histopathology and reduced likelihood to recur, Wright held it to be distinct from the parakeratotic type. Both the first two editions of the World Health Organization's (WHO) histological classification of odontogenic tumours recognized that "cases with orthokeratosis are seen".³ The WHO's 2005 edition,⁴ having reclassified the parakeratotic type as the keratocystic odontogenic tumour (KCOT), stated, "Cystic jaw lesions that are lined by orthokeratinizing epithelium do not form part of the spectrum of a ... KCOT."⁴

Aims and research questions

The aim of this study was to report a consecutive series of OOCs in the Hong Kong Chinese to determine their clinical and conventional radiological presentations. This is important as the Hong Kong Chinese have already displayed features in three benign odontogenic neoplasms^{5–9} that differ from other communities. Therefore, a review of the newly recognized odontogenic neoplasm affecting this community is merited.

The ethnic and environmental background of the Hong Kong Chinese and some of the pathology affecting their jaws has already been outlined.¹⁰

The research question for this study was "What are the clinical and conventional radiological characteristics of OOCs observed in a largely Southern Chinese Hong Kong community?" This required a detailed analysis of the clinical and conventional radiological features observed in a continuous series of cases of OOC lesions in a Hong Kong Chinese community admitted to Prince

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Philip Dental Hospital (PPDH), which also houses The University of Hong Kong's Faculty of Dentistry.

Materials and methods

The histopathological files of PPDH between 1988 and 2004 were reviewed for OOCs. The clinical notes and radiographs of each case were retrospectively reviewed. In order to diminish the effects of "expectation bias", which is intrinsic to a retrospective review of cases, the radiographs were read prior to the clinical notes. Each patient's sex, age, clinical history and findings on examination, and the differential or provisional diagnosis were obtained from the clinical records. The definitive diagnosis of an OOC was made on the basis of the histopathological report. Each OOC was radiographed in two planes. Panoramic and periapical radiographs were available for all cases. Every mandibular case was accompanied by a true occlusal radiograph. Periapical or occlusal radiographs were available for all lesions in the anterior region in both jaws. Occipitontal and lateral sinus views had been obtained for those cases involving the maxillary antrum. The radiographs were viewed on a standard illuminated screen under reduced ambient lighting. The influence of the OOC on adjacent structures, such as the teeth, the buccal and lingual cortices, lower border of the mandible and the maxillary antrum, was noted.

Definition of parameters

The criterion used to determine, objectively, the degree of definition of the boundary of the lesion was that established by Slootweg and Muller.¹¹ A lesion was considered to be well-defined when its radiodensity changed markedly within a distance of 1 mm when passing from the lesion to the adjacent normal bone.

The lower border of the mandibular alveolus is set by the inferior dental (mandibular) canal and the upper border of the maxillary alveolus is set by the image of the hard palate on panoramic radiographs or lateral cephalographs.¹²

The jaws were further divided into posterior (molar and premolar) and anterior (incisors and canines) sextants. The demarcation point between them was a vertical line passing just distal to the distal surface of the canine.

Results

All lesions recorded as odontogenic keratocyst (OKC) and Gorlin's syndrome in the Hong Kong Chinese were identified (see Table 1). Only those cases that had pre-biopsy clinical notes and radiographs were included. 33 non-syndromic cases were reported as the parakeratotic type (now KCOT) and 2 were the mixed parakeratotic and orthokeratotic type. Six were syndromic (naevoid basal cell carcinoma syndrome) and excluded. Four were considered OKCs on the basis of clinical and

radiological features, but the epithelium was so inflamed that histopathology could not confirm this; they were, therefore, excluded.

Finally, five cases of OOC were found. An outline of the histopathological context within which they were found is set out in Table 1. Details of the five patients' gender, age, clinical presentation or complaint, size and distribution, and radiological features are displayed in Table 2. The provisional and differential diagnoses are set out in Table 3. The mean age of the patients was 33.5 SD 13.06 years. 2 were male (mean age 22.00 SD 9.90 years) and all were female (mean age 42.33 SD 11.06 years). Four lesions affected the mandible and one the maxilla. The two female patients with mandibular lesions had a much older mean age (43.00 SD 15.57 years) at first presentation than the males (22.00 SD 9.90 years); this was not significant.

All five patients were, at least, aware of their lesions for a period of time before presentation (0.11 SD 0.09 years). The patient with the maxillary case presented longer after the first awareness (0.16 years) than patients with cases affecting the mandible (0.10 SD 0.01 years). Three of the patients with OOCs, which presented in the fourth decade or later, presented later after their first awareness of the lesions (0.16 SD 0.08 years) than the two patients who presented in the first three decades (0.03 SD 0.01 years).

Four patients first presented with swelling, two with pain and two with a purulent discharge. All five lesions affected solely a posterior sextant.

All five OOCs appeared radiographically as well-defined radiolucencies; three were unilocular (Figures 1 and 2) and two were multilocular (Figure 3). All five displayed cortication and buccolingual expansion (Figure 2). Three mandibular lesions reached the lower border of the mandible; however, only one displaced and eroded it. Two others expanded a lateral cortex down past the undisplaced lower border of the mandible (Figures 1 and 3).

Reduction in canal diameter, displacement or absence of the inferior dental canal (IDC) are denoted in that order by the triplets of Y(Yes) or N(No) in Table 2. Displacement (Figure 1) and absence (Figures 1 and 3) of the IDC occurred in all four mandibular OOCs. A reduction in the diameter of the canal was apparent in only the two youngest patients.

Although all cases displayed tooth displacement, not one was associated with root resorption. Three cases were associated with unerupted third molars (Figure 1).

Table 1 The histopathological context within which the orthokeratinized odontogenic cysts were found

	<i>Number of cases</i>
Naevoid basal cell carcinoma syndrome	6
Inflamed clinically diagnosed keratocysts which could not be histopathologically confirmed.	4
Mixed parakeratotic-orthokeratotic type	2
Keratocystic odontogenic tumours	33
Orthokeratinized odontogenic cysts	5
Total	50

Table 2 Orthokeratinized odontogenic cystsCase number & presenting complaint &
period of prior awareness

Gender (M/F)

& age (years)

& age (years)																							Assoc. teeth
			R	I	G	H	T					L	E	F	T								
Maxilla			Retro-molar	8	7	6	5	4	3	2	1	1	2	3	4	5	6	7	8	Retro-molar			
1. SPD	0.16	F 41																					
Mandible			Vertical ramus	8	7	6	5	4	3	2	1	1	2	3	4	5	6	7	8	Vertical ramus			
1. S	0.02	M 15	I-Rm-Uni(E, RLB(E&D)YYY-I																	8			
2. SP	0.04	M 29																		8			
3. D	0.08	F 32	I-Uni (E,Past,YYY&D)I																				
4. S	0.25	F 54	R-----Multi ----(E, RLB, NYY)-----I																	None			
			I-----Multi---(E,PastNYY,D)-----SN																	None			

S, swelling; P, pain; I, incidental finding; D, discharge; M/F, male/female; Anatomical parameters: A, angle; C, condyle; Cr, coronoid; MF, mandibular foramen; R, ramus (vertical); SN = sigmoid notch.

Radiology data codes for each case: Uni, unilocular; Multi, multilocular; All multilocular lesions have a soap-bubble appearance. The presence "E" means buccolingual expansion (when "E" is absent then there is no buccolingual expansion); Lower border of the mandible: RLB, reaches lower border (presence of "E" and/or "D" means "erosion" and "displacement" respectively; Past, displaces a lateral cortex down past the lower border; Inferior dental canal triplet (N, no; Y, Yes) 1st N or Y means narrowing of canal; 2nd N or Y means displacement of canal; 3rd N or Y means that at least part of the canal is absent or not seen; D, tooth displacement; Assoc.Teeth, associated unerupted teeth.

Table 3 Differential and provisional diagnoses

<i>Histology</i>	<i>No. of</i>	<i>Differential diagnosis</i>																
<i>OOC</i>	<i>cases</i>	<i>Cysts</i>					<i>Benign neoplasms</i>							<i>Malignant neoplasms</i>			<i>'Odontogenic keratocyst'</i>	
		<i>OKC</i>	<i>Dent.</i>	<i>Rad.</i>	<i>TBC</i>	<i>ABC</i>	<i>A.bla</i>	<i>Myx</i>	<i>CEOT</i>	<i>GC</i>	<i>COT</i>	<i>AOT</i>	<i>FOLs</i>	<i>Ca</i>	<i>Sar</i>	<i>Malignant</i>	<i>Alone</i>	<i>Not Cited</i>
Total	5	5	3	1	0	1	3	0	0	0	0	0	0	0	0	0	0	0

ABC, aneurysmal bone cyst; A.bla, ameloblastoma; AOT, adenoid odontogenic tumour; Ca, carcinoma; CEOT, calcified epithelial odontogenic tumour; COT, calcified odontogenic tumour; Dent, dentigerous cyst; GC, giant cell lesion; Myx, myxoma; OOC, orthokeratinized odontogenic cyst; Rad, radicular cyst; Sar, sarcoma.

Figure 1 Part of a panoramic radiograph of mandibular case 2. This displays a well-defined unilocular radiolucency with a corticated margin. The entire image of an unerupted third molar appears within the radiolucency. It extends from the mesial aspect of the first molar almost to the mandibular foramen. It also expands a lateral cortex down past the eroded, but still undisplaced, lower border of the mandible. The inferior dental canal has been displaced downwards towards the lower border of the mandible, where it is no longer apparent.

A provisional diagnosis was available for all five cases (Table 3) and all five cases included an “odontogenic keratocyst” in the differential diagnosis. It was not unexpected that the differential diagnosis of those lesions associated with unerupted teeth should include the dentigerous cyst.

Four cases were primarily treated by enucleation and Carnoy’s solution. The fifth was treated by enucleation alone. 4 had been followed-up for 8.5 years, 2 at the time of the study had already been followed up for 10 years (mandibular cases 2 and 4) and 1 other (maxillary case 2) for at least 8 years. Mandibular case 3 had already been discharged after 6 years’ follow-up. Mandibular case 1 discharged himself shortly after surgery. No cases recurred.

Discussion

A main feature that persuaded Wright² to consider OOC as a separate entity, other than the histopathology, was the reduced rate of recurrence. The OOC was also almost absent from cases of “naevoid basal cell carcinoma syndrome”; only Bolbaron and co-authors have reported one such case.^{13,14}

An important clinical feature of OOCs affecting the Hong Kong Chinese was the short period of awareness of the lesions before they present for diagnosis and treatment. The patient’s prior awareness is crucially

Figure 2 Part of a posteroanterior projection of mandibular case 2. Although it displays substantial buccolingual expansion the unerupted third molar appears to be sited within the line of the dental arch and, therefore, centrally placed within the lesion. The lateral cortex is displaced down past the lower border of the mandible in Figure 1. The expanded buccal cortex is obvious at that point.

important in disease management. The earlier the patient reports his or her disease, the earlier treatment can be provided and the more successful the final result. This is achieved by operating on smaller lesions and thus reducing the morbidity and recurrence. The mean time of prior awareness of OOC was 0.11 years in comparison to a mean of 0.46 years for odontogenic myxomas,⁵ 0.67 years for ameloblastomas,⁶ 0.77 years for KCOT,⁹ 3.60 years for fibrous dysplasia¹⁰ and 3.65 years for OF,⁸ all affecting the same Hong Kong Chinese community contemporaneously. The difference between the prior awareness of OOC and of KCOT was significant (see Table 4). A reason for any delay in seeking help from Western medical practitioners may be that the Hong Kong Chinese may have used “Traditional Chinese Medicine” in the first instance.⁵ The reason for the earlier presentation of OOC in comparison to the others can only be surmised. The

Figure 3 Part of a panoramic radiograph of mandibular case 4. This exhibits a well-defined multilocular radiolucency with a corticated margin. It extends from the distal aspect of the second premolar back to the base of the sigmoid notch. It expands the anterior margin of the vertical ramus and displaces a lateral cortex down past the lower border of the mandible.

most likely reason was that the onset of OOC symptoms must have been sudden and intense enough for the patient to acknowledge them. They must also have persisted at the same intensity or worsened in order for the patient to seek assistance so soon afterwards.

Although four OOCs displaced adjacent teeth, none exhibited root resorption. Therefore, root resorption was not a feature of the OOC and this differs markedly from KCOT (see Table 4).

An association between OOC and unerupted teeth had been made by both Vuhahula et al.¹⁵ and Crowley et al.¹⁶ The association, particularly of the crown, appears to lead, not surprisingly, to the inclusion of a dentigerous cyst within the differential diagnoses of the OOCs in this report.

Although all radiological features of OOCs and KCOT did not differ statistically (Table 4), the most visually striking radiological feature of these Hong Kong Chinese OOCs was their buccolingual expansion. Although the pattern of expansion was almost identical to that of the ameloblastoma affecting the same community (compare Figure 2 with MacDonald-Jankowski et al Figure 4⁷) it differed, with regards to degree, to that of the KCOT (compare Figure 2 with MacDonald-Jankowski et al Figure 2⁹). This also included the downward displacement of a lateral cortex past the largely undisplaced lower border of the mandible in two cases. So far this feature has only been reported in ameloblastomas affecting the same community (compare Figures 1 and 3 with MacDonald-Jankowski et al Figure 1⁷). It has not been observed in any of the KCOTs affecting the same community.⁹ Both the ameloblastoma⁷ and KCOT in this community⁹ frequently displayed root resorption, which was not a feature of these OOCs.

The treatment and management of OOC were similar to that of KCOT, reflecting the fact that both were considered variants of the same lesion, the odontogenic keratocyst, at the time of their presentation. This is clear in Table 4, in which all or the majority of both lesions cited “odontogenic keratocyst” in their differential diagnosis.

The compliance of this, albeit, small number of Hong Kong Chinese patients with follow-up is noteworthy. Follow-up of those lesions with a propensity to recur (ameloblastoma⁶ and odontogenic myxoma⁵) or become reactivated (fibrous dysplasia¹⁰) appeared to attract almost full compliance, whereas those which display a minimal tendency to recur, such as ossifying fibroma,⁸ may not. Although OOC has the reputation

Table 4 Comparison between the clinical and radiological features of the orthokeratinized odontogenic cyst (OKC) and the keratocystic odontogenic tumour

Feature	Orthokeratinized odontogenic cyst	Keratocystic odontogenic tumour	Statistical analysis
Gender, males:females	2:3	18:15	$\chi^2 = 0.33$; 1df: $P > 0.05$
Age, mean and SD (in years) (n)	34.20 SD 14.48 years (5)	30.58 SD 16.33 years (33)	$t = 0.50$; 36df: $P > 0.05$
Jaw, maxilla:mandible	2:4	20:13	$\chi^2 = 3.03$; 1df: $P > 0.05$
Period of prior awareness	0.11 SD 0.09 years (5)	0.77 SD 1.28 years (26)	$t = 2.66$; 29df: $0.05 > P > 0.01$
Swelling, yes:no	4:1	20:13	$\chi^2 = 0.64$; 1df: $P > 0.05$
Pain, yes:no	2:3	12:21	$\chi^2 = 0.03$; 1df: $P > 0.05$
Incidental, yes:no	0:5	6:27	$\chi^2 = 1.09$; 1df: $P > 0.05$
Expansion, yes:no	5:0	27:6	$\chi^2 = 1.10$; 1df: $P > 0.05$
Unilocular, multilocular lesions:	3:2	16:17	$\chi^2 = 0.24$; 1df: $P > 0.05$
yes:no			
Root resorption, yes:no	0:5	13:19	$\chi^2 = 3.25$; 1df: $P > 0.05$
Tooth displacement, yes:no	4:1	22:10	$\chi^2 = 0.28$; 1df: $P > 0.05$
Unerupted teeth, yes:no	3:2	20:12	$\chi^2 = 0.00$; 1df: $P > 0.05$
Cases which cited OKC in their differential diagnosis, yes:no	5:0	24:8	$\chi^2 = 1.14$; 1df: $P > 0.05$
Follow-up, yes:no (lost to F.U.)	4:1	30:3	$\chi^2 = 0.63$; 1df: $P > 0.05$

of a minimal tendency to recur, during the time of its treatment and follow-up it was still considered a variant of the odontogenic keratocyst and managed according to the protocol for that lesion. Crowley et al¹⁶ reported that only 1 OOC recurred out of 44 OOCs or a 2.2% recurrence rate in contrast to 42.6% for KCOTs.

Conclusions

1. Although OOC presented many of the features observed in the ameloblastoma and the KCOT, such as a predilection for the posterior mandible and multilocularity, so far the OOC has not displayed root resorption, a feature observed significantly frequently in ameloblastoma and KCOT.

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2. Buccolingual expansion was frequently observed. This expanded the entire mesiodistal extent of the lesion also observed for the ameloblastoma, but not for KCOT.

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