

## Odontogenic Keratocyst in Jamaica: A Review of Five New Cases and Five Instances of Recurrence Together with Comparative Analyses of Four Treatment Modalities

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### ABSTRACT

**Aim:** Five new cases of odontogenic keratocyst (OKC) together with five instances of recurrence are reviewed with special emphasis on radiology and surgical management. A comparative analysis of four different treatment modalities used in the treatment of OKC in these patients (new and recurrent cases) is reported.

**Subjects and Methods:** The case notes and radiographs of patients who had histological confirmation of OKC at both the Cornwall Regional Hospital and Kingston Public Hospital in Jamaica were reviewed for demographics, radiological presentation, treatment modalities and outcome of treatment. Cases of recurrence were separated from new cases. This study was conducted for the period 1980 to 2004.

**Results:** Five new cases and five instances of recurrence were documented over the 25-year period. The new cases of OKC keratocyst accounted for 1.71% of the total jaw bone tumours and 12% of OKC keratocysts over the first 16 years. The posterior mandible appears to be the most favoured site. Of significance, one case of nevoid basal cell carcinoma syndrome (NBCCS) and a case of ameloblastomatous transformation in the wall of an OKC keratocyst were recorded. The age range of the new cases was 12 to 44 years.

**Conclusion:** The radiological finding from this review is similar to previous reports. However, the authors record a unique and historic case of ameloblastomatous transformation of OKC. A case of OKC in NBCCS is also documented. Of all four surgical treatment modalities compared, only cryosurgery was promising, so far, with no recurrence after a follow-up period of six years.

## Queratoquiste Odontogénico en Jamaica: Revisión de Cinco Casos Nuevos y Cinco Casos Reincidentes Acompañados de Análisis Comparativos de Cuatro Modalidades de Tratamiento

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### RESUMEN

**Objetivo:** Se examinan cinco casos nuevos de queratoquiste odontogénico (QOO) junto con cinco casos reincidentes, con énfasis especial en el tratamiento radiológico y quirúrgico. Se reporta un análisis comparativo de las cuatro diferentes modalidades de tratamiento usadas en el tratamiento del QOO en estos pacientes (los casos nuevos y los casos reincidentes).

**Sujetos y métodos:** A fin de conocer la demografía, la presentación radiológica, las modalidades de tratamiento y los resultados del tratamiento, se examinaron las radiografías y las notas de los casos

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que tuvieron confirmación histológica de QOO, tanto en el Cornwall Regional Hospital como en el Kingston Public Hospital de Jamaica. Los casos de reincidencia fueron separados de los casos nuevos. Este estudio abarcó el periodo comprendido de 1980 a 2004.

**Resultados:** Cinco nuevos casos y cinco casos reinicidentes fueron documentados en ese periodo de 25 años. Los nuevos casos de QOO dieron cuenta del 1.71% del total de tumores de mandíbula y del 12% de los queratoquistes odontogénicos en los primeros 16 años. La mandíbula posterior parece ser el sitio más favorecido. De significación particular fue la documentación de un caso de síndrome de carcinoma basal celular nevoide (SCCBN), y un caso de transformación ameloblastomatosa en la pared de un queratoquiste odontogénico. El rango de la edad de los nuevos casos fue de 12 a 44 años.

**Conclusión:** El hallazgo radiológico de este estudio, es similar al de reportes previos. Sin embargo, los autores documentan un caso único e histórico de transformación ameloblastomatosa del QOO. También se documenta un caso de QOO en SCCBN. De las cuatro modalidades de tratamiento comparadas, sólo la criocirugía ha sido promisorio hasta el momento, por cuanto no presentó reincidencias luego de un periodo de seguimiento de seis años.

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## INTRODUCTION

The term odontogenic keratocyst (OKC) was first used by Philipsen in 1956 (1). In 1963, Pindborg and Hansen (2) described the essential features of this cyst. The most significant characteristic of this cyst is the exceedingly high recurrence rate. Various modalities have been utilized in the management of OKC (3–9). These modalities include marsupialization, enucleation, and *en bloc* resection with or without immediate reconstruction. However, irrespective of the modality of treatment, the recurrence rate of OKC is known to be high (from 5%–62.5%). This high recurrence rate has been attributed to the presence of epithelial remnants of satellite cysts in the osseous margin.

The use of cryosurgery seems to be very promising as Schmidt and Pogrel documented a recurrence rate of only 11.5% in 36 patients (22 of the 36 patients had received previous treatment consisting of enucleation alone) treated with enucleation and liquid nitrogen cryotherapy (10).

## SUBJECTS AND METHODS

The case files of all patients who had histological confirmation of the diagnosis of odontogenic keratocyst from 1980–1995, at both the Cornwall Regional Hospital and Kingston Public Hospital are reviewed and studied for demographics, radiographic appearance and clinical behaviour. No record pertaining to a patient was recorded twice as instance of recurrence was documented separately up to 2004 (a period of 25 years). These records subsequently formed part of the records on a computer programme (tumour powerhouse) designed by one of the authors, which can be used both on PC and McIntosh computers for the continuous documentation of data relating to jawbone tumours in Jamaica.

Four treatment modalities (enucleation, cryosurgery, *en bloc* resection without reconstruction and radical resection with reconstruction, utilizing autogenous bone graft) were used. All these surgical modalities are compared based on

the presence or absence of recurrence after an appropriate follow-up period.

## RESULTS

Five new cases of OKC were documented during this period. Of these, four patients were female and one male. Odontogenic keratocyst accounted for 2.8% of jaw bone tumours and 12% of odontogenic cysts in Jamaica, during the period 1980–1995 (a 16-year period). Five instances of recurrence were also documented during this 16-year period and up to 2004. These are summarized in Tables 1 and 2.

The posterior mandible was the most favoured site except for *case 1* (Table 1) which occurred anteriorly and posteriorly on both sides (Fig. 1). *Case 3*, which was a patient with nevoid basal cell carcinoma syndrome (NBCCS), had lesions of OKC affecting all four quadrants of the jaw (Fig. 2a and 2b). Ameloblastomatous transformation in the walls of the lesions was the unique histological finding in *case 1*.

The mean age was 30 years with a range of 21–44 years. Table 3 shows a comparative analysis of four treatment modalities- (enucleation, cryosurgery, *en bloc* resection without autogenous cortico-cancellous bone graft and radical resection with reconstruction with autogenous bone graft). Of all these treatment modalities, only the case treated by cryosurgery did not recur up to seven years post surgery.

### Case report 1: Odontogenic keratocyst with ameloblastic transformation.

A 21-year old female Jamaican presented to the Cornwall Dental Centre in Jamaica on January 3, 1995, with a two-month history of a painless intra-oral swelling of the right mandible and a six-month history of numb feeling of the following teeth: # 31–33, 41– 43.

### Radiographic investigation

A dental panoramic tomogram revealed a well defined and moderately well corticated, extensive radiolucent lesion from

Table 1: Showing the cases of odontogenic keratocyst in Jamaica

Case	Sex	Age	Site	Radiological Appearance
1	F	21	Ant mandible (bilateral) with posterior extensions	Extensive radiolucency of R & L mandible with root resorption
2	F	34	Post mandible (R)	Extensive radiolucency of mandible from 4/ region up to ascendingramus ramus
3**	F	33	All four quadrants	Opacification of R & L antrum* and radiolucency of R & L mandible
4	M	22	Post mandible (L)	Large radiolucent area involving both vertical and horizontal ramus and incorporating UE / 8
5	F	44	Post mandible (L)	Radiolucent lesion of posterior mandible involving angle and ascending ramus

Table 2: Showing the cases of recurrent odontogenic keratocyst in Jamaica

Case	Sex	Age	Site	Radiological Appearance
1	F	33	Post mandible (R) after 10 years	Extensive area of bone destruction with evidence of pathologic fracture
2	F	34	Post mandible (L) after 1 year	Extensive area of bone destruction of posterior mandible
3	F	48	Post mandible (L)	Extensive radiolucency of mandible /678 region up to ramus



Fig. 1: Dental panoramic tomogram showing an extensive multilocular radiolucency which was subsequently diagnosed as an odontogenic keratocyst.

the region of # 45 (distal) crossing the midline to the region of #34. The lesion extends from the lower cortical bone to the apex of all the teeth in this region. The radiolucency was unilocular with no internal bony septa. Resorption of the roots of # 41–44 and #31–33 was also present (Fig. 2).

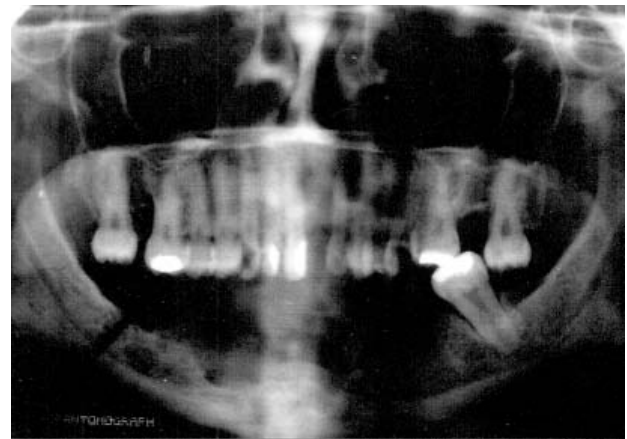


Fig. 2: Dental panoramic tomogram of patient shown in figure 1, 4-years post cryosurgery.

**Histopathological investigation**

The lesions were primarily an OKC exhibiting the typical parakeratotic squamous cell lining which was variably thick and easily detached from the underlying stroma. In one area, however, the epithelium showed transition to ameloblastic type epithelium and there were small buds which were present in the stroma. This is considered to represent ameloblastomatous transformation in the cyst wall; the so-called unicystic ameloblastoma.

**Treatment**

Approximately four years after diagnosis, the patient accepted the treatment of cryosurgery with preservation of the lower cortical bone of the mandible. The radiological follow-up for a period of six years (Fig. 3–5) showed no recurrence. Subsequently three Endopore® implants were placed six years after cryosurgery in solid bone which showed adequate bone volume.

Table 3: Showing the authors modality of treatment and outcome for four cases of odontogenic keratocyst

Case	Sex	Site	Appearance	Radiological Modality	Treatment/ Follow-up Status
Case 1	F	Post Mandible ®	Extensive radiolucency of mandible from 44 region to the ascending ramus	Resection with reconstruction with autogeneous bone from the iliac crest	Recurred within 2 years
*Case 2	F*	All 4 quadrants	Opacification of the R and L antrum and radiolucency of R and L mandible	Enucleation of mandibular lesion.	Recurred in less than 1 year.
Case 3	M	Post mandible ®	Evidence of recurrence within the grafted site after 13 years	Enucleation	Recurred within 2 years
Case 4	F	Posterior and bilateral mandible	Multilocular radiolucency	Cryosurgery alone	No recurrence up to 4 years of follow-up.

§ A Case of BCNS



Fig. 3: Dental panoramic tomogram of patient shown in figure 1, 5-years post cryosurgery.



Fig. 4: Posterior –anterior view of the jaws showing bilateral lesions of odontogenic keratocyst in the patient with NBCCS.

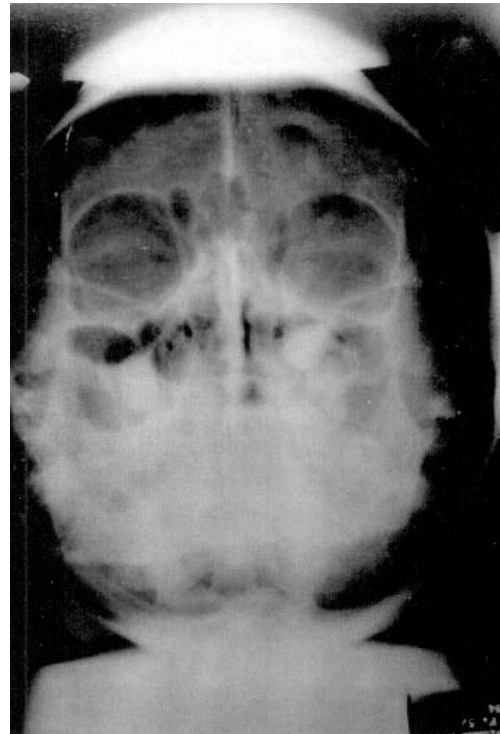


Fig. 5: Occipitomeatal view confirming pathologies in both right and left maxilla in the patient with NBCCS.

### Case Report 2: Multiple odontogenic keratocyst in nevoid basal cell carcinoma syndrome

A 33-year old female Jamaican presented with a history of swelling of her left cheek on February 9, 1994, following an alleged assault by her boyfriend. The radiological examination revealed the following: extensive radiolucent lesion of the left and right mandible in the posterior region with tooth #38 embedded within the lesion on the left side of the mandible (Fig. 8). There were cystic lesions of the right and

left maxillary sinus in relation to the misplaced molars on both sides, which are missing clinically from the dental arch (Fig. 9), and there was calcification of the falx cerebri. The patient's facial features, which included frontal bossing, broad nasal ridge, associated hypertelorism together with her low IQ were suggestive of NBCCS.

Her past dental history dates to 1981 when she presented to the department of oral and maxillofacial surgery at the Cornwall Regional Hospital with painful swelling of the right side of the cheek for two years. Her intra-oral examination then revealed that various molar teeth which were not previously extracted were missing. A radiographic examination revealed cystic lesions of all the four quadrants of the jaw and misplaced molars were found to be associated with all the four cystic lesions. These lesions were then suggestive of multiple OKC as seen in the NBCCS which was confirmed histologically.

Her medical examination revealed ophthalmological and cutaneous abnormalities clinically diagnosed as naevus.

Her uterus was unremarkable but salpingogram revealed bilateral hydrosalpinges. The lesion recurred on the right side of the mandible with evidence of pathological fracture and the lesion on the left side of the mandible, also recurred shortly after enucleation.

## DISCUSSION

Five new cases of OKC were documented during the study period. In this series of jawbone tumours in Jamaica, OKC accounted for 1.7% of the total jawbone tumour and 12% of odontogenic cysts.

The posterior mandible seems to be the most favoured site except for case one which occurred anteriorly and bilaterally (Fig. 1) and case three which was a patient with NBCCS with lesions of OKC affecting all four quadrants of the jaw. Ameloblastic transformation of the walls of the lesion was the unique histopathologic finding in case one. The age range of this series was 21 to 44 years with an average age of 30.8 years.

Radiologically all cases characteristically presented as an extensive well-defined area of bone destruction. The border of these lesions appears to be thinly sclerosed (Fig. 1) except for case one (Fig. 7) which presented as an almost total destruction of the posterior mandible, including the ramus with evidence of pathologic fracture of the right mandible.

Only *case 3* (NBCCS) had unerupted teeth associated with three of the four lesions (the unerupted tooth in the left mandible might have been removed during the initial surgery). Root resorption was another radiological finding seen distinctly in *case 1* which histologically had ameloblastic transformation of its wall. With regard to the cases of recurrent OKC, two were solitary OKC and two were recurrence in one patient who had OKC in association with NBCCS.

Recurrence is usually characterized radiologically by evidence of further destruction of the bone with or without the clinical evidence of infection. Radiological findings in this current series of OKC from Jamaica are similar to what has been previously reported in the literature. The clinical findings are similar to other reports (11–16). However, it is not possible in a series with five new cases and five instances of recurrence to deduce any statistically significant gender ratios. In this series, the mandible seemed to be the most favoured site.

Although the cyst may occur at any age, from the very young to the elderly, Brannon (15) found it to be exceedingly rare under the age of 10 years, as only two such patients were in his series of 233 cases (11–12). The peak incidence is in the second and third decades of life with a gradual decline thereafter. The data of Browne (12) with 104 patients and Forsell (11) with 119 patients are virtually identical. In all series, there is predilection for the male gender, ranging from 1.44: 1 in the study of Brannon (15) and 1.46: 1 in the study of Browne (12) to 1.79: 1 in a study by Forsell (11). As in this series, the mandible is the more favoured site than the maxilla, 65% vs 35% in Brannon's series and 78% vs 22% in that of Forsell (11). In these series, the ramus third molar area followed by the first and second molar areas are the most favoured sites.

Multiple OKC have been documented to occur with some frequency, but notably associated with the NBCCS. Ameloblastomatous transformations of the odontogenic keratocyst is very rare, but has been reported previously (14). As stated in a case recently reported by Ichalque and Rippin (16), the presence of an ameloblastomatous island does point in the direction of nevoid basal cell carcinoma syndrome since in previous studies such histological features were seen in NBCCS, however our only case of ameloblastomatous transformation did not fit into the criteria for the NBCCS, thus differing from previous documentations and findings.

The nevoid basal cell carcinoma syndrome was first reported in 1894 by Jarish (17), who described a patient with multiple basal cell carcinoma, scoliosis and learning disability. Howel and Caro in 1959 (18) were the first to associate the nevoid basal cell carcinoma syndrome with the cutaneous disorders and anomalies, while Gorlin and Goltz in 1960 (19) defined the condition as a syndrome comprising the triad of multiple basal cell naevi, jaw keratocyst and skeletal anomalies (20). A wide range of neurological, endocrine, ophthalmic and genital manifestations (20–22) are known to be variably associated with this syndrome. The prevalence of the NBCCS has been estimated to range from 1 in 57 000 (23) to 1 in 164 000 (24) but there is now a general agreement that the prevalence is about 1 in 60 000 (25). This syndrome probably arises in all ethnic groups but most reports have been in Caucasians. Male and female are equally affected (26, 27).

Since the description of the nature of the odontogenic keratocysts and its possible histological subtypes in 1960, there has been discussion regarding the most appropriate clinical management of OKC. It is generally stated that these cysts are more aggressive forms of odontogenic cysts because of the high recurrence rate due to the presence of epithelial remnants of satellite cysts in the osseous margin. It is believed that the parakeratinized variant (previously called primordial cyst) is thought to have a higher recurrence rate than the orthokeratinized variant. It is for this reason that a more aggressive treatment than simple enucleation has been advocated (3, 4). The recommended treatment is as follows: curettage with peripheral osteoectomy (5, 6), cryosurgery (curettage with liquid nitrogen therapy) (2), curettage pulse application of carnoy's solution (7), localized en bloc resection (8, 9) and occasionally, mandibular segmental resection (8).

Table 3 shows a comparative analysis of four modalities of treatment utilized by one of the authors in this current series from Jamaica. Of significance is the encouraging result following the use of liquid nitrogen in case one, which showed no recurrence up to date (six year radiological follow-up). Schmidt and Pogrel in 2001 (10) concluded that the combination of enucleation and liquid nitrogen cryotherapy may offer improved therapy in the management of OKC. We agree with this based on the comparative analysis of the four modalities of treatment of which cryosurgery lead to no recurrence (Table 3).

More recently Pogrel and Jordan (28) described their experience with marsupialization of lesions of OKC. They concluded that marsupialization was the definitive treatment for odontogenic keratocyst as all ten cases (6 males and 4 females) in their series resolved completely after marsupialization.

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