

# CLINICAL AND RADIOLOGICAL BEHAVIOUR OF SPORADIC ODONTOGENIC KERATOCYST – A STUDY

<sup>1</sup>MUSLIM KHAN, BDS, FCPS (Oral Surgery)

<sup>2</sup>QIAM UD DIN, BDS, (Gold Medalist), MSc (Oral Surgery) (UK)

<sup>3</sup>ATA UR REHMAN, BDS, FCPS (Oral Surgery)

## ABSTRACT

*Odontogenic Keratocyst (OKC) is the most aggressive of the jaw cysts because of their greater tendency to recur. They are relatively common developmental cysts of the jaws. The present study (A descriptive case series) is about the clinical, features i.e. age, sex, site distribution and radiological features of sporadic odontogenic keratocyst. The present study was performed on 30 biopsy confirmed cases over a period of 5 years. This cystic lesion is common in males (63%) as compared to females (37%). The most common age group is the third (36.6%) and second decade (30%) of life. The most common site according to the present study was the posterior mandible i.e., the body and ramus of the mandible (40%), followed by the anterior mandible crossing the midline (20%). Anterior and posterior maxilla is not frequently involved by this cyst. Only in 9.9% of the cases the upper jaw was involved. Swelling has been the most common chief complaint i.e. (61.702%). In 16.6% of patients, Odontogenic keratocyst was a chance radiographic finding. Radiologically most of the cysts were multilocular radiolucencies (83%).*

**Key words:** Odontogenic Keratocyst, OKC, Dental Lamina, Khyber College of Dentistry

## INTRODUCTION

Odontogenic keratocysts (OKCs) are unusual cystic lesions that arise from the dental laminal epithelium and usually occur in the mandible or the maxilla.<sup>1</sup> Odontogenic keratocysts can arise sporadically or in association with the nevoid basal cell carcinoma syndrome (NBCCS or Gorlin syndrome), in which patients present with multiple abnormalities, especially basal cell carcinomas, OKCs, skeletal changes, and palmar pits.<sup>2</sup> Because of distinctive clinical features, aggressive clinical behavior, high recurrence rate, and association with basal cell carcinoma syndrome (BCCS), the odontogenic keratocyst (OKC) is unusual in comparison to other cysts affecting maxillofacial region.<sup>3</sup>

The odontogenic keratocyst (OKC), first described by Philipsen in 1956,<sup>4</sup> was recently designated by the World Health Organization as a keratocystic odontogenic tumor and was 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

behaviour.”<sup>5</sup> All aspects of the Odontogenic keratocyst OKC from its biological behavior to its optimal treatment, have been debated since its first description by Philipsen in 1956.<sup>4</sup>

In 1960, Gorlin and Goltz initially described the simultaneous existence of multiple basal cell carcinomas, multiple OKCs of the mandible and the maxilla, bifid ribs, and other variable manifestations that comprise the basal cell nevus bifid rib syndrome.<sup>6</sup> The first clinical investigation to reveal a high propensity for recurrence was by Pinborg and Hansen in 1963.<sup>7</sup>

The histologic features of the OKCs include a thin parakeratinized or orthokeratinized stratified squamous epithelium (usually five to eight cells in thickness) well defined basal cell layer generally without rete peg formation, an un-inflamed fibrous capsule and a lumen containing desquamated keratin.<sup>8</sup> The parakeratin lining predominates in upto 97% of OKCs. The Connective tissue wall often contains small islands of epithelium and small satellite or daughter cysts.<sup>9</sup>

<sup>1</sup> Assistant Professor Oral and Maxillofacial Surgery, Khyber College of Dentistry, Peshawar

<sup>2</sup> Professor, Head, Supervisor, Oral and Maxillofacial Surgery, Principal/Dean, Khyber College of Dentistry, Peshawar

<sup>3</sup> Demonstrator Oral and Maxillofacial Surgery, Khyber College of Dentistry, Peshawar

**Correspondence:** Dr Muslim Khan, Asst Prof Oral Surgery, Khyber College of Dentistry, Peshawar University Campus, Cell: 0300-5846906, Email: muslim177@hotmail.com

In the past two decades the benign neoplasm like behaviour of OKCs of the jaw has been a burning issue. The purpose of the present study is to evaluate the clinical and radiographic behaviour of this troublesome cystic lesion of the maxillofacial region. The study will help in the treatment planning of odontogenic keratocyst patients.

**METHODOLOGY**

The present study was carried out on biopsy proven cases of Odontogenic Keratocysts at oral and maxillofacial surgery unit of Khyber College of Dentistry, Peshawar between January 2004 to January 2009 over a (period of five years). The purpose of the study was to determine the clinical (age, sex and site distribution) and radiological features of Odontogenic Keratocysts. For this purpose a proforma was designed to evaluate these variables. The various radiographs taken were peri-apical view, occlusal view (both for maxilla and mandible), postero-anterior view of face, lateral oblique view mandible, paranasal sinus (PNS) view and Orthopantomograms (OPGs). Various surgical procedures performed under local or general anesthesia were enucleation, marsupialization, marsupialization followed by enucleation and enucleation with peripheral ostectomy depending upon the indications of these procedures. The cystic linings were sent for biopsy and confirmed as Odontogenic Keratocyst. The recurrent OKCs and syndromic patients Nevroid basal cell carcinoma syndrome (Gorlin syndrome) were excluded from the study.

**RESULTS**

The age range was from 4 years to 70 years with the mean age of 31.83 years SD  $\pm$ 17.87 and the peak incidence is in the third (36.6%) and second (30%) decades of life. Table 1. Odontogenic Keratocyst were more common in males i.e.19 (63%) as compared to females 11 (37%) with the male to female ratio of 1.72:1. Fig 1. The most common site according to the present study was the posterior mandible i.e., the body and ramus of the mandible (40%), followed by the anterior

TABLE 2: ANATOMICAL SITE DISTRIBUTION OF ODONTOGENIC KERATOCYST

Anatomical site	No of patients	Percent-age
Anterior Maxilla	2	6.6%
Posterior Maxilla	1	3.3%
Anterior body of the mandible crossing Midline	6	20%
Mandibular condyle	1	3.3%
Mandibular coronoid process	2	6.6%
Ramus and body of the mandible	12	40%
Body of the mandible	2	6.6%
Ramus of the Mandible	1	3.3%
Body and Angle of the Mandible	3	10%
Total	30	100%

TABLE 3: PRESENTING COMPLAINT OF DENTIGEROUS CYST PATIENT

Presenting complaint	No of patients	Percent-age
Swelling	11	36.6%
Fluid discharge	7	23.3%
Altered taste	2	6.6%
Missing mal-aligned teeth	1	3.3%
Chance radiographic finding	5	16.6%
Limited mouth openin	3	10%
Pain	1	3.3%
Total	30	100%

TABLE 1: AGE DISTRIBUTION OF KERATOCYST PATIENTS

Age in years	No. of patients	Percent-age
0-10	1	3.3%
1-20	9	30%
21-30	11	36.6%
31-40	1	3.3%
41-50	5	16.6%
51-60	2	6.6%
61-70	1	3.3%
Total	30	100%

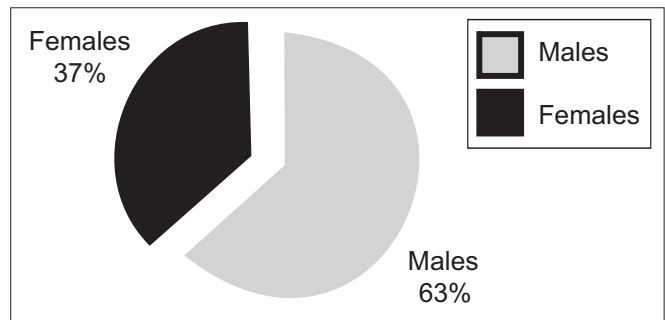


Fig 1. Gender Distribution of OKC Patients

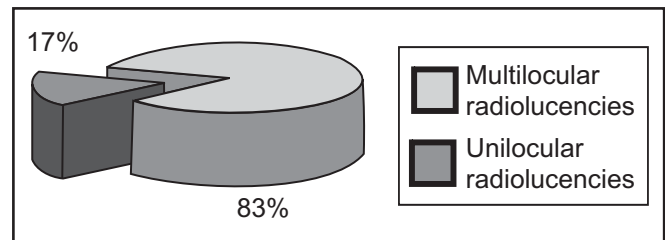


Fig 2. Radiological features of Odontogenic Keratocyst



Fig 3. Multilocular radiolucency crossing midline of the mandible is typical of keratocyst

mandible crossing the midline. Maxillary bone is not frequently involved by Odontogenic Keratocyst i.e. in 9.9% of cases Table 2. Swelling and fluid discharge has been the most common presenting complaint (Table 3) of the OKC patients i.e., 36.6% and 23.3% of the patients respectively. Three of the patients reported with limited mouth opening which is very un-usual for the jaw cysts. Radiologically most of the OKCs were predominantly multilocular i.e., 83% only 17% of the cystic lesions were unilocular. Fig 2

## DISCUSSION

Odontogenic Keratocyst cannot really be considered a malignancy; but some of its characteristics make this pathology extremely serious and aggressive. In fact, keratocysts tend to recur, can reach considerable dimensions, and can arise close to delicate and important anatomical structures (i.e. alveolar nerve). All these characteristics had suggested, in the past, that an aggressive surgical approach should be followed in order to eradicate these cysts completely. Such radical behavior might result in severe mutilation, so, for this reason, it is not always advisable<sup>9</sup> further more, its clinical and radiographic features rarely allow the making of a correct diagnosis, thus being a challenge for the oral surgeon.<sup>10</sup> The purpose of the study was to evaluate the clinical and radiological features of odontogenic keratocysts in the region North West Frontier Province of Pakistan. The study is first of its kind in the region which will be an addition to literature review of OKCs. Khyber College of Dentistry is the only tertiary care centre for the management of maxillofacial problems in the region.

Frequency data for any particular lesion are nor reliable indicators of their incidence and are often merely pointers that vary from institution to institution, reflecting the range of material seen in them. According to a study from Shear M of the 2616 cysts of the jaws 292 (11.2%) were Odontogenic keratocysts that occurred in 255 patients.<sup>11</sup> Unfortunately such studies on the incidence of odontogenic keratocyst are lacking in this region.

Keratocysts are found more frequently in men than women and this gender predilection was more pronounced in blacks than white patients.<sup>12, 13</sup> This study is consistent with the M Shear's<sup>12</sup> study and Wooglar's<sup>13</sup> findings when it comes to age. According to the present study Odontogenic Keratocyst is more common in males (63%) than females (37%), with male to female ratio of 1.72. Some studies show an equal gender distribution of the OKCs such as that of Rensburg study.<sup>14</sup>

Keratocysts occur over a wide age range, and cases have been reported as early as first decade and as late as the 9<sup>th</sup> decade of life. In most series there has been a pronounced peak frequency in the second and third decade of life with figure ranging from 40% to 60%. Many workers have demonstrated a bimodal age distribution with a second peak in the fifth decade of life or even later.<sup>15,16</sup> While the age range according to our study is from 4 years to 70 years with the mean age of 31.83 years SD  $\pm$ 17.87 and the peak incidence is in the 3<sup>rd</sup> (36.6%) and 2<sup>nd</sup> (30%) decades of life. The second peak is also in the fifth decade of life in our series of patients, i.e. 16.6%.

According to the present study 80% of the OKCs occur in the mandible excluding mandibular condyle and mandibular coronoid process which are quite unusual sites, as far as their site distribution is concerned. The present study is consistent with the Shear M<sup>1</sup> series of patients in which the frequency of mandibular involvement ranges from 69 to 83%. Only in 10% of the cases reported with maxillary involvement.

Approximately one half of the keratocysts occur at the angle of the mandible and extend for varying distance in the ascending ramus and forward into the body due to its antero-posterior growth pattern. As to the site distribution of other cases, reports of several studies indicated that they can occur anywhere in the jaws including the midline of the mandible and the maxilla and the globulomaxillary area in the maxilla.<sup>8,17,18,19</sup> Studies have also shown that the keratocyst occurred with much higher frequency in the maxilla after the age of 50 years.<sup>20</sup>

Swelling and fluid discharge has been the most common presenting complaint of the OKC patients i.e., 36.6% and 23.3% of the patients respectively. Three of the patients reported with limited mouth opening which is very un-usual for the jaw cysts. That was because of unusual site involvement i.e. coronoid and condylar process involvement.

Radiographically OKCs demonstrate a well defined unilocular or multilocular radiolucency Fig 3. When unilocular radiolucent OKC is encountered, it is difficult to distinguish it from other odontogenic or non-odontogenic cysts; when the multilocular variant is

present, it is difficult to differentiate it from other odontogenic or non-odontogenic neoplasms (eg, ameloblastoma, myxoma). One radiographic feature that may suggest the diagnosis of OKC is that, OKCs tend to grow in an anterior-posterior direction within the medullary cavity of the bone without causing obvious bone expansion. However, this feature is difficult to see in maxillary cysts. These lesions expand at the expanse of the medullary space. In maxillary lesions perforation of the floor of the maxillary sinus or the nasal cavity and the buccal cortex may occur.<sup>14</sup> The present case series shows that 83% of the odontogenic keratocysts were multilocular and only 17% were unilocular with well defined borders.

Odontogenic keratocysts are known as the “great mimickers” of many odontogenic jaw cysts. A study by Myoung et al<sup>21</sup> reported that the radiographic diagnosis concord with the histological findings in only 25.2% of 256 OKCs cases when diagnosis was made by oral surgeons. Although OKCs display characteristic T2 weighted image features, they may resemble dentigerous cyst, traumatic bone cysts, radicular cysts, ameloblastoma, lateral periodontal cysts, and enlarged dental follicles.<sup>21</sup> one of the author’s case resembles nasopalatine duct cyst.<sup>22</sup> OKCs may arise in other jaw cysts such as dentigerous cyst, residual cysts and lateral periodontal cysts<sup>23</sup>

A number of clinicians favor “conservative” therapy, while others advocate more “aggressive” forms of treatment. Meiselman et.al<sup>24</sup> considers conservative therapies to include “enucleation, curettage, and marsupialization.” Williams et al<sup>25</sup> define aggressive treatment as “that which is used in addition to enucleation, and includes curettage (mechanical, physical, and/or chemical) and/or resection with or without loss of jaw continuity.

**Acknowledgement:** The authors are very thankful to the staff members of Oral and Maxillofacial Surgery unit of Khyber College of Dentistry, Peshawar for their help and co-operation in the completion of the study.

## REFERENCES

- 1 Shear M. The aggressive nature of the odontogenic keratocyst: is it a benign cystic neoplasm? Part 1: clinical and early experimental evidence of aggressive behaviour. *Oral Oncol.* 2002; 38:219–26.
- 2 Shear M. The aggressive nature of the odontogenic keratocyst: is it a benign cystic neoplasm? Part 2: proliferation and genetic studies. *Oral Oncol.* 2002; 38: 323–31.
- 3 Richard CK. Histology and ultrastructural features of odontogenic keratocyst. *Oral and Maxillofacial surg Clin N Am* 2003;15: 325-33.
- 4 Philipsen HP. Om keratocyster (kolestetomer) i kaeberne. *Tandlaegebledet* 1956; 60:963-81.
- 5 Philipsen HP. Keratocystic odontogenic tumor. In: Barnes L, Eveson JW, Reichart PA, Sidransky D, World Health Organiza-
- 6 Gorlin RJ, Goltz RW. Multiple nevoid basal cell epithelioma, jaw cysts and bifid rib: a syndrome. *N Engl J Med* 1960; 262: 908-12
- 7 Pinborg JJ, Hansen J. Studies in odontogenic cyst epithelium. Clinical and roentgenologic aspects of odontogenic keratocysts. *Acts Pathol Microbiol Scand* 1963; 58:283-94.
- 8 Brannon RB. The odontogenic keratocyst. A clinicopathologic study of 312 cases. Part II. Histologic features. *Oral Surg Oral Med Oral Pathol* 1977; 43: 233-55.
- 9 Manfredi M, Vescovi P, Bonanini M, Porter S: Nevoid basal cell carcinoma syndrome: A review of the literature. In *J Oral and Maxillofac Surg.* 2004; 33: 117-24.
- 10 El-Hajj G, Anneroth G: Odontogenic keratocyst—A retrospective clinical and histologic study. *Int J Oral Maxillofac Surg* 1996; (24):124-8
- 11 Stoelinga PJ: Long-term follow-up on keratocysts treated according to a defined protocol. *Int J Oral Maxillofac Surg* 2001; (30):14-9
- 12 Shear M. Odontogenic Keratocysts: Clinical Features. *Oral and Maxillofacial Surg Clin N Am* 2003; (15): 335-45.
- 13 Wooglar JA, Rippin JW, Browne RM. The odontogenic keratocyst and its occurrence in nevoid basal cell carcinoma syndrome. *Oral Surg Oral Med Oral Pathol* 1987; (64): 727-30
- 14 Renseberg V L, Paquette M, Morkel A J, Norttje J C. Correlative MRI and CT imaging of the odontogenic keratocyst: a review of twenty-one cases *Oral and Maxillofacial Surg Clin N Am* 2003; (15): 363-82
- 15 Toller PA, Origin and growth of the cysts of the jaws. *Ann R Coll Surg Engl* 1967; (40): 306-36.
- 16 Rippin JW, Wooglar JA. The odontogenic keratocyst in BCNS and non-syndrome patients. In Browne RM, editor. *Investigative pathology of the odontogenic cysts.* 1<sup>st</sup> Edn. Boca Raton (FL) CRC Press: 1991. 221-32
- 17 Vedtofte P, Praetorius F. Recurrence of odontogenic keratocysts of the jaws in relation to clinical and histological features: A 20 years follow up study of 72 patients. *Int J Oral Surg* 1979; (8):412-20
- 18 Forsell K. the Primordial cyst: a clinical and radiological study. *Proc Finn Dent Soc* 1980; 76:129-74
- 19 Browne RM. The odontogenic keratocysts: histologic features and their correlations with clinical behaviour: *Br Dent J* 1971; (131):249-59
- 20 Wooglar JA, Rappin JW. Browne RM. A comparative study of clinical and features of recurrent and nonrecurrent odontogenic keratocyst. *J oral Pathol* 1987; (16): 124-8
- 21 Myoung H, Hong SP, Hong SD et al. Odontogenic keratocyst: review of 256 cases for recurrence and clinicopathological parameters. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2001; 91(3): 328-33
- 22 Yih,W Y . Krump, L J. Odontogenic Keratocyst in the Nasopalatine Duct Associated With Mural Cartilaginous Metaplasia. *J Oral Maxillofac Surg* 2005; (63):1382-84.
- 23 Minami M, Kaneda T, Ozawa K, et al. cystic lesions of the maxillomandibular region: MR imaging distinction of the odontogenic keratocyst and ameloblastoma from other cysts. *Am J Roentgenol* 1996; 166: (4): 943-9
- 24 Meiselman F: Surgical management of the odontogenic keratocyst: Conservative approach. *J Oral Maxillofac Surg* 1994; (52):960-3
- 25 Williams TP, Connor FA: Surgical management of the odontogenic keratocyst. *J Oral Maxillofac Surg* 1994; (52):964-7