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KERATOCYSTIC ODONTOGENIC TUMOUR: A CLINICO-RADIOGRAPHIC CORRELATION OF TWENTY CASES WITH SPECIAL EMPHASIS ON DIFFERENTIAL DIAGNOSIS

Sonam Khurana[1], Dhruv Khurana[2]
1.Consultant Oral & Maxillofacial Radiologist, Sonipat, Haryana, India
2.PhD Student, Department of Neuroeconomics, Claremont Graduate University, Claremont, CA, USA

ABSTRACT:

Objectives: The objective of this retrospective evaluation of twenty cases is to compare the clinical and radiographic features of Keratocystic Odontogenic Tumours and provide an appropriate differential diagnosis.

Study Design: Analysis of data related to patients' age, gender, clinical and radiographic features, differential diagnosis, and treatment was done.

Results: The mean age in males was 25.1 years and in females was 42 years. Twelve (60%) out of twenty KCOTs were located in the mandible and eight in the maxilla. Out of twenty, nineteen (95%) tumors showed well-demarcated margins. Out of these twenty cases, fifteen showed corticated borders and five showed non-corticated borders. Fourteen tumors showed unilocular appearance and six tumors showed multilocular appearance. Tooth displacement and impacted teeth were associated with eight tumors and root resorption with two tumors. The buccolingual expansion was seen in seven tumors and maxillary sinus was involved in three cases. Histologically, all tumors showed para keratinized stratified squamous epithelium.

Conclusions: KCOT more commonly seen in males in the second decade of life. Radiographic features in the majority of patients presented unilocular radiolucency with well-defined, corticated margins and expansion in anteroposterior direction with tooth displacement more than root resorption.

Keywords: Keratocystic Odontogenic Tumours (KCOT), Radiographic Features, Differential Diagnosis.

INTRODUCTION:

The term Keratocystic Odontogenic Tumour (KCOT) was proposed by WHO in 2005 and also included in the WHO's histological classification (2005) of odontogenic benign tumours.[1] It is defined as "benign uni- or multicystic, intraosseous tumour of odontogenic origin, with a characteristic lining of para keratinized stratified squamous epithelium and has potential aggressive, infiltrative behaviour." Keratocystic odontogenic tumor (KCOT) has bimodal age occurrence with the first peak at second and third decades and second occurrence in the fifth decade of life. KCOT has predominance in males and more common in blacks than whites.[2] The tumor most commonly involves mandible than the maxilla. The site most commonly involves is the angle of the mandible. It may be solitary or multiple.[1] In numerous cases, patients tend to observe symptoms very late because KCOT expands in the anteroposterior direction in medullary cavity without cortical expansion.[3]

*Corresponding Author Address: Dr Sonam Khurana. E-mail: khurana.sonam@gmail.com

According to Woolgar and co-workers, multiple KCOTs should always be considered as part of NBCC syndrome irrespective of the presence of other features of syndrome because sometimes other features are so mild that diagnostic criteria couldn't meet.[5, 6,] KCOT has the proclivity to recur which attributes to small satellite cyst or fragments of epithelium which remain after surgical removal of the cyst.[4]

Radiographic Features: - KCOT may exhibit small, round or ovoid unilocular radiolucent appearance on plain radiographs, although larger lesions with scalloped and multilocular appearance may also occur. The radiolucencies are well demarcated with thick sclerotic borders, but may be diffused in parts.[1]

CT Findings: - On bony window settings of Conventional CT, KCOT presents scalloping of the margins, internal spiculations, and small crevices involving the peripheral cortex of the lesions and appears hypodense to isodense to muscle on soft tissue window settings. CT attenuation values (in a study by L J van Rensburg and co-workers) [7] vary between -8.2 HU and 60 HU. It is difficult to delineate large expanding lesions with thin cortex from surrounding soft tissues.[7]

MRI Findings: On T1 weighed images, KCOT shows inhomogenous, hypointense to isotense signal to muscle with no enhancement. In case of infected cyst with focal area of inflammation and ulcerative changes of epithelial lining, cyst typically shows signal enhancement after administration of gadolinium. The KCOT lesions characteristically show high signal intensity with signal inhomogeneity or ''signal drop out'' on T2 images. The signal drop out is due to the amorphous, keratinaceous debris which is neutral intensity material that has been described histologically. The variable T1 contrast uptake and mixed T2 signal intensity and with the identification of daughter cysts characteristically allows diagnosis of KCOT with the help of MRI. [7]

Histopathology: The KCOTs are typically lined by para keratinized stratified squamous epithelium, usually about 6-10 cell layers thick and without rete ridges. It often has palisaded, basal layer of columnar or cuboidal cells with nuclei oriented away from the basement membrane. The luminal parakeratotic layers often have a corrugated surface. The cystic cavity of KCOTs may contain keratinaceous material. Mitotic figures are found frequently in the suprabasal layers.[2]

Treatment: Marsupialisation had been used for KCOTs in the past, although marsupialising the neoplastic tissue is not justified. Other surgical options include enucleation, curettage, peripheral ostectomy, removal of overlying mucosa in cases of cortical perforation and osseous resection including marginal or segmental osteotomies and enucleation with Carnoy's solution.[8, 9]

The main aim of this article is to evaluate clinical and radiographic features of keratocystic odontogenic tumor in twenty patients and give differential diagnosis after evaluation.

MATERIALS AND METHODS:

The study was conducted on twenty cases of keratocystic odontogenic tumor and cases were confirmed clinically, radiographically and histologically. The patients were treated radically or conservatively depending upon the individual case. All the specimens were sent for histopathologic examination post surgically.

RESULTS:

The total 20 cases of KCOTs were seen in 15 patients. The four cases out of fifteen were associated with Gorlin Goltz syndrome and showed multiple KCOTs. The case no.1, 2 &3 of Gorlin Goltz syndrome showed 2 cases each and case no. 4 showed 3 cases. Out of 15 patients, six were females and nine were males. The youngest patient was 18-year-old, while the eldest was 59 years of age. The mean age in males was 25.1 years, while that for females was 42 years [Table 1].

Location: Twelve out of twenty cases were seen in the mandible and eight in maxilla. Out of twelve mandible cases, four cases were extending in anterior-posterior direction, eight were seen in posterior region with these eight posterior cases seven involved ramus of the mandible. Out of eight maxillary cases, four involved anterior region, two involved Posterior region and two were extending in anterior-posterior direction.

Clinical presentation: In all 15 patients, the initial chief complaint was the swelling of the lower jaw and/or face. Only six patients experienced pain along with swelling. None of the patients experienced paraesthesia.

Radiographic features:The radiographic appearance of 20 cases of KCOT is depicted in Figure 1-.8. Total 14 cases out of 20 appeared unilocular and 6 cases appeared multilocular with scalloped margins. (Figure 1, 2) Total 19 lesions showed well-defined margins and only one showed ill-defined margin (Figure 3). In 6 multilocular lesions, we found both curved and straight septae and 4 lesions showed incomplete septation (false septation) and 2 lesions showed complete septation with scalloped margins (Figure 4).

Effects on surrounding structures: (See Tabel no. 1)

Histopathological features: All 20 cases showed Parakeratinized Stratified Squamous epithelium with 6 to 10 cell layers thickness without rete pegs.

Treatment: Out of 12 mandibular cases 4 were enucleated followed by application of Carnoy's solution, 2 were enucleated with peripheral ostectomy, 3 were treated by segmental resection and 3 were treated by marginal resection. In case of 8 maxillary lesions, the 4 maxillary lesions were treated conservatively by enucleation followed

by peripheral Osetectomy. In 3 cases partial maxillectomy was done and in 1 case Hemi maxillectomy was done. The follow up of these patients are ranged from 12 months to 72 months until date. Four patients showed recurrence of the lesion and were treated radically. Other patients showed satisfactory healing.

DISCUSSION:

The Keratocystic Odontogenic tumor has a wide range of occurrence from the first decade of life to the ninth decade. The first peak evidence occurs at second and third decades and the second peak at fifth decade of life or later. The peak incidence usually occurs a decade earlier in females than males.[3] In our case series, the mean age of male (25.1 years) and females (42 years) was in accordance with the previous studies by Mervyn Shear [3] and White and Pharoah[4]. According to Shears M [2, 3] KCOT is more common in mandibular molar ramus area than maxilla in non-syndromic patients. In syndromic patient, maxillary third molar area is most commonly involved site. In present study, 12/20 cases were seen in mandibular region and 8/12 cases in mandibular molar region. Out of 8 maxillary lesions, 4 were seen in syndromic patients. Hence results were in accordance with the literature. According to white and Pharoah, [4] KCOT usually presents no symptoms, but occasional swelling may occur. Pain occurs only after infection. In contrary to the literature, all patients in our case series presented with swelling. The pain was presented by six patients. The unilocular or multilocular lesions occupying a large portion of the jaw with out cortical expansion is the main radiographic feature of KCOT explained by Darpan Bhargava and co-authors.[10] In the present case series, 70% cases appeared unilocular and 30% multilocular. The periphery of the lesion is corticated unless infected with a smooth round or oval shape. [4] We noticed 95% well defined and 5% ill-defined lesion in our case series. 70% lesions with thick corticated borders, 5% lesion with thin corticated border and 25% non-corticated lesions. The most important features of KCOTs are to grow in anterior-posterior direction, with minimal expansion except in the region of upper ramus and coronoid process. [4] but in our case series, we have noticed expansion in 35% cases (20% in buccal direction and 15% in lingual direction). This can be attributed to their late detection and hence allow the lesion to expand. The other effects on surrounding tissues are resorption and impaction of the teeth but to a lesser degree than dentigerous cyst.[4] We found root resorption in 10% cases, tooth displacement in 40% cases and tooth impaction due to cyst in 40% cases.

Key points to differentially diagnose KCOTs on the basis of radiographic features: [4, 11]

If the lesion shows well defined unilocular radiolucency then the differential diagnosis could be:	
1. Lateral (Developmental) periodontal	***KCOTs***

cyst:- a) Shows small round, well-defined radiolucency. b) Located midway between cervical & apical portion of the root. c) Most commonly located in premolar-canine area d) Causes buccal cortical expansion. e) Little tendency to move adjacent teeth.	a) *Could present as* small round, well-defined radiolucency or large multilocular lesion. b) *Usually located apical to CEJ.* c) *Most commonly involve mandibular third molar-ramus area.* d) *Causes minimal expansion.* e) *Greater tendency to move adjacent teeth.*
2. Radicular cyst:- a) Round radiolucency surrounded by narrow radiopaque margins. b) Extend from lamina dura of involved teeth. c) Associated tooth is non- vital	***KCOTs*** a) Could present as round radiolucency surrounded by narrow radiopaque margins. b) Do not extend from the lamina dura of involved teeth. c) Associated teeth could or could not be non-vital.
3. Residual Cyst:- a) Well defined radiolucency with distinct sclerotic margins. b) On occlusal radiograph, expanded cortex shows hydraulic effect.	***KCOTs*** a) Could show Well defined radiolucency with distinct sclerotic margins. b) Doesn't show hydraulic growth effect.
4. Globulomaxillary Cyst:- a) Present in Maxillary Anterior Region. b) Inverted pear like radiolucency. c) It is a clinical term given to any cyst which occurs in Globulomaxillary area. It can be Keratocyst, Radicular Cyst & Lateral Periodontal Cyst.	***KCOTs*** a) Globulomaxillary Cyst is a clinical term and histologically it can be a Keratocystic odontogenic tumor.
5. Simple Bony Cyst a) Mostly occurs in posterior mandible and ramus area. b) Margins vary from well-defined delicate cortex to ill-defined borders with wide zone of transition. c) The shape is mostly oval with scalloped borders. d) Minimal or no effects on surrounding tooth structures. e) Minimal or no expansion f) Mostly associated with cemento-osseous dysplasia and fibrous dysplasia.	***KCOTs*** a) Location is same as that of simple bony cyst. b) Margins are well defined with thick sclerotic borders and hence narrow zone of transition. c) The shape is mostly oval with scalloped borders. d) Cause tooth displacement and impaction of developing teeth. e) Minimal or no expansion f) Multiple KCOTs are associated with basal cell nevus syndrome.

• ***If the lesion shows multilocular radiolucency then the differential diagnosis could be:***	
1. **Ameloblastoma :-** a) Buccal as well as lingual cortical expansion. b) Knife edge resorption of adjacent roots. c) Honey comb or soap bubble pattern is seen in the lesion.	***KCOTs*** a) Minimal or no cortical expansion b) Root resorption is not a typical feature of KCOT but can occur in some cases. c) Show scalloped margins with thickest condensed cortication with few false septae gives it multilocular appearance.
2. **Odontogenic Myxoma:-** a) Mild Expansion b) Septa forms elongated straight lines intersecting at right angles (tennis racket or stepladder-like appearance.	***KCOTs*** a) Mild Expansion. b) We found both curved and straight septae in our study and usually there in no specific pattern.
3. **Central Giant Cell Granuloma (CGCG):-** a) Difficult to differentiate from keratocyst when thin septae and scalloped variant of CGCG are present. b) Typically shows triangular crenations at the borders with poorly mineralized sclerotic margins. c) Expansion of both buccal, as well as lingual cortical plates was seen. d) CGCG typically envelops the roots, causing the roots to protrude into the lesion and aggressive cases cause root resorption in multiplanar direction**.** e) Usually, occurs anterior to mandibular first molar.	***KCOTs*** a) Difficult to differentiate from scalloped variant of CGCG. b) Typically shows scalloped margins with thick condensed cortication. c) Mild or rare expansion seen. d) Usually, cause tooth displacement or impaction. Root resorption rarely occurs. e) Usually, it occurs posterior to mandibular first molar.
• **If associated with impacted tooth:- (Pericoronal variant)**	
1. **Adenomatoid Odontogenic Tumour** a) Site: Anterior maxilla (Canine is the most common site) b) Well-corticated margins seen with significant expansion. c) Two-third of cases show radiopaque flecks.	***KCOTs*** a) Can occur in anterior maxillary region but posterior mandibular region is the most common site. b) Well, corticated margins with minimal or no expansion. c) Don't show radiopacity.
2. **Dentigerous Cyst** a) It arises cervical to CEJ. b) Well, corticated, thin & smoothly curved	***KCOTs*** a) KCOT mostly present apical to CEJ.

with little tendency towards scalloping. c) The buccal Cortical expansion is more commonly seen. d) Knife edge resorption of roots is a typical feature.	b) Well corticated with scalloped margins. c) Mild or no expansion. d) Root resorption is not a typical feature of KCOTs but can occur in rare cases.

CONCLUSION:

The key features of KCOTs in our study are not different from the literature. The characteristic features of KCOTs are male predominance, with bimodal age occurrence with the peak at second and fifth decade of life. The posterior mandibular region is the most common site with enlargement at the expense of medullary bone. The radiographic features of KCOTs range from well-defined unilocular radiolucencies with sclerotic margins to scalloped margins and multilocular patterns. The maxillary KCOTs are usually smaller in size than mandibular lesions. It has tendency to displace adjacent teeth and can cause impaction of developing teeth but rarely causes root resorption. Histologically, it is lined by para keratinized epithelium six to ten layers thick with presence of "Daughter cyst." The treatment of KCOTs should be aimed at complete removal of the cystic wall and include resection, curettage, and marsupialization. Post treatment periodic check-up is also necessary to lessen the chances of recurrence. The patient should be checked both clinically and radiographically post treatment. Recurrence is common within 5 years post treatment but can be delayed up to 10 years.

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TABLE:

Table 1: Effects on surrounding structures

Cortical expansion			Sinus Involvement (Figure 5)	Impacted teeth (Figure 6)	Tooth displacement (Figure 7)	Root resorption (Figure 8)
Buccal	Lingual	Inferior cortex				
3/20	4/20	3/20	3/20	8/20	8/20	2/20

FIGURES:

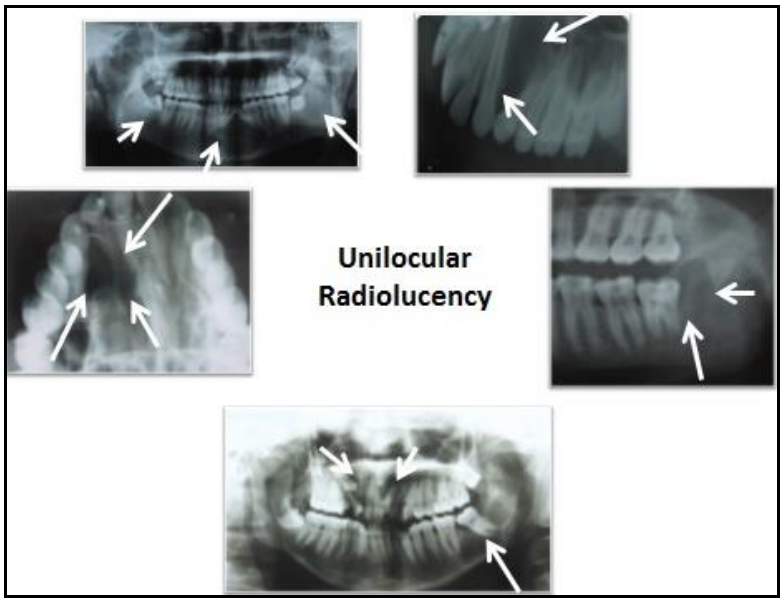

Figure 1: Images of Unilocular Radiolucencies (White

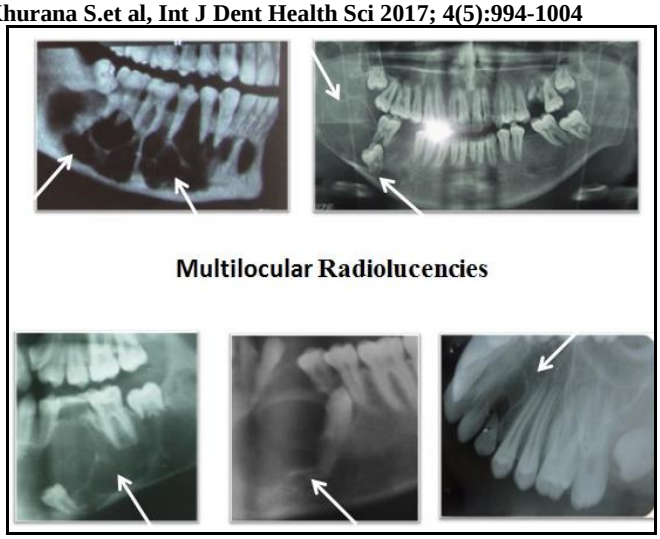

Figure 2: Images of Multilocular Radiolucencies (White arrow)

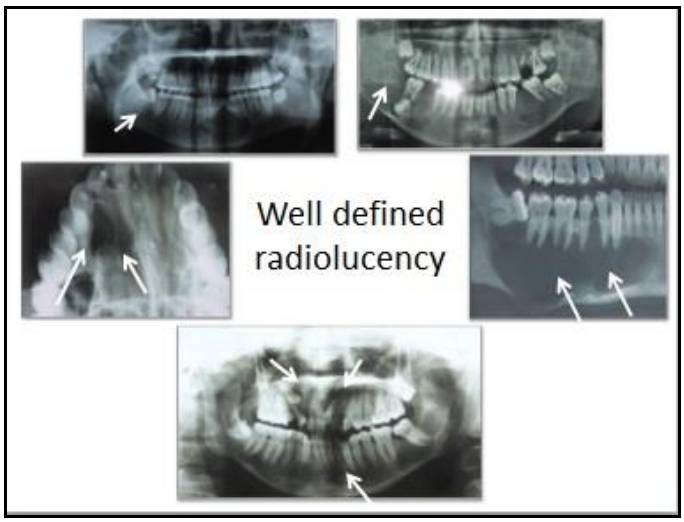

Figure 3: Images of Well Defined radiolucencies (White arrow)

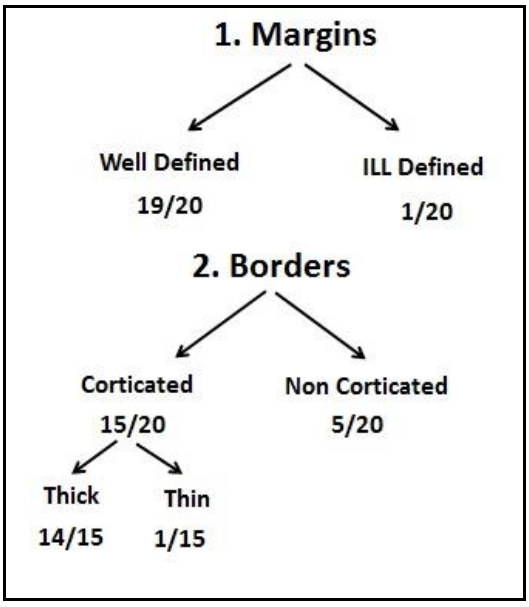

Figure 4: Flow chart depicting Margins and Borders in 20 cases of KCOTs

Figure 5: Axial View showing Complete Haziness of Right Maxillary Sinus with Deviated Nasal Septum

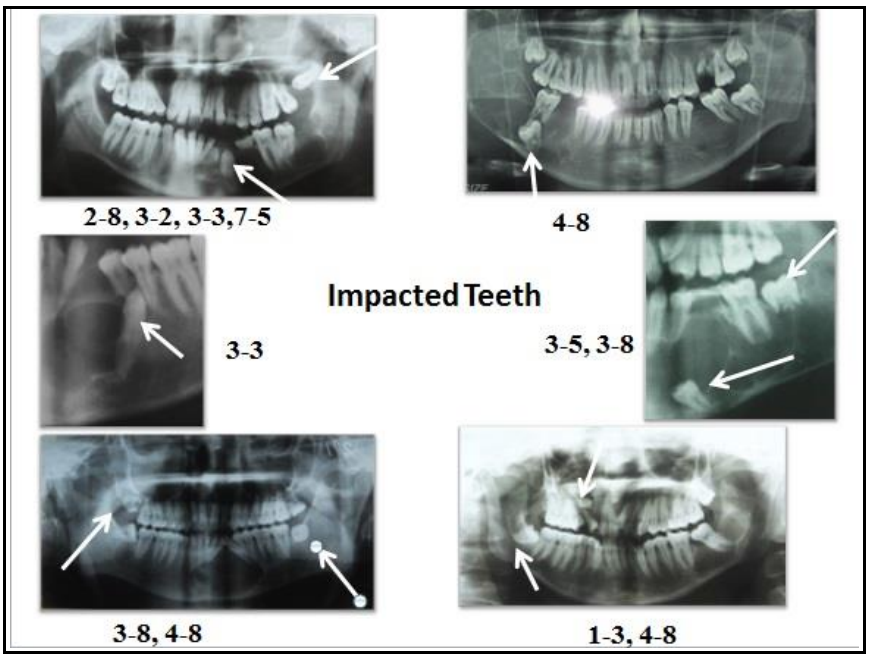

Figure 6: Images of Impacted Teeth associated with KCOT

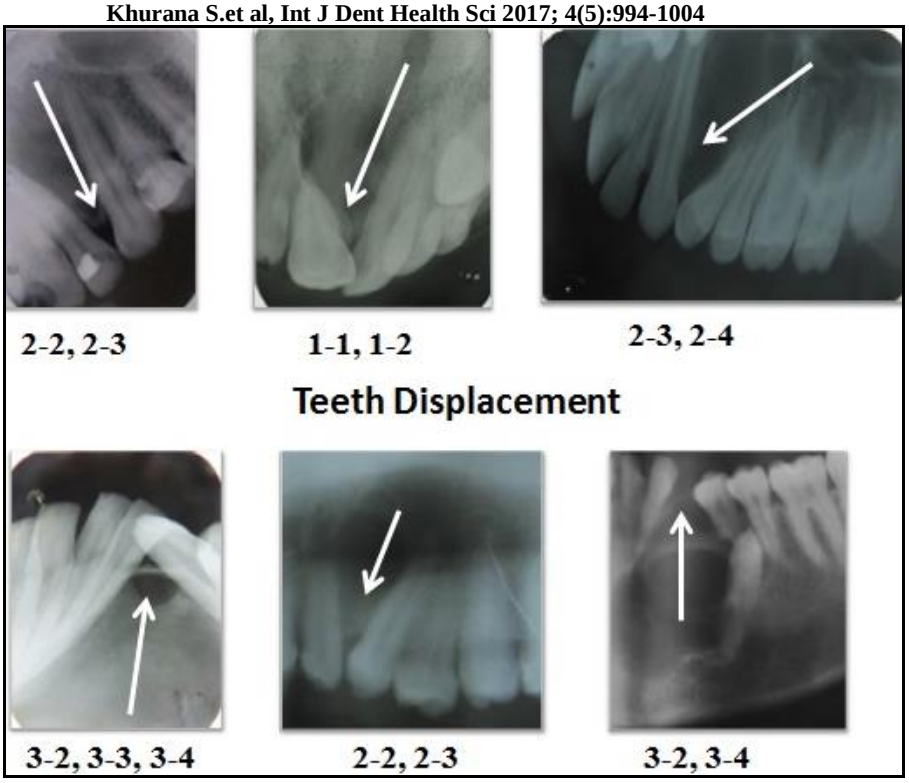

Figure 7: Images of Displaced Teeth due to KCOTs (White arrow)

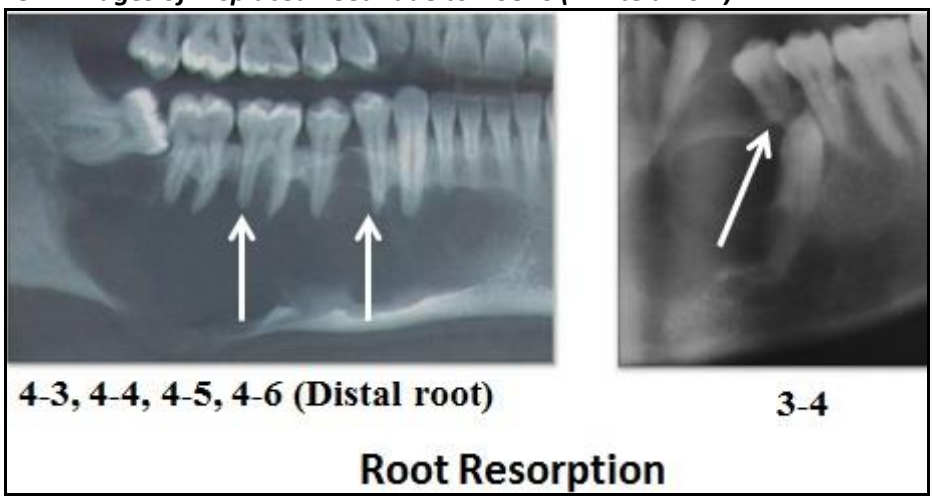

Figure 8: Root resorption due to KCOTs (White arrow)