

Case Report

Radiological and clinical features of peripheral keratocystic odontogenic tumor

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Received November 21, 2013; Accepted December 19, 2013; Epub January 15, 2014; Published January 30, 2014

Abstract: Peripheral Keratocystic odontogenic tumor (PKCOT) is rare and has not been reported with imaging findings. Aim: The aim of this study was to explore CT and MR imaging characteristics of two PKCOTs in lateral facial deep region (LFDR), and to present a review of the literature regarding their anatomic locations and characteristics of epidemiology and Radiology. Methods and material: Eighteen PKCOTs, sixteen from the previous literatures and two new cases of the authors, were reviewed. Fifteen of them had original sites in the gingival, one in buccal space and two in LFDR. CT imaging features were based on one PKCOT in the buccal space and two in LFDR. MR imaging features on one PKCOT in LFDR, and conventional radiographic characteristics on eight PKCOTs were analyzed. Results: The subjects' ages ranged from 37 to 81 years, with a mean of 54.7 years. The male and female ratio was 1:1.14, with no predilection for either gender. Buccal space (6%) and LFDR (11%) were relatively rare original sites to PKCOTs, compared to gingival (83%). PKCOTs were clearly depicted on CT and MR imaging as they had cystic changes. Contents of the cysts were further analyzed by using different series of MRI. No radiological features were found on radiographs except one subject with minimal bone resorption in the alveolar crest. Conclusions: LFDR is rare original site for PKCOT. PKCOT should be included as one of differential diagnosis of a cystic lesion found in LFDR on CT and MR imaging.

Keywords: Peripheral keratocystic odontogenic tumor, keratocystic odontogenic tumor

Introduction

Odontogenic keratocyst (OKC) occurs mainly in the jaws, with a mandible/maxilla ratio of 2:1 [1]. The term OKC was first proposed by Philipsen [2] in 1956. In 1962, Pindborg et al [3] established the histopathologic criteria for diagnosis of OKC, in which parakeratinization was particularly emphasized. In 2005, the World Health Organization (WHO) [4] reclassified OKC to keratocystic odontogenic tumor (KCOT) based on its clinical behaviors including potential aggression, infiltrative growth, and a high rate of recurrence up to 62.5%.

On rare occasions, KCOT may occur extraosseously, mainly in the gingiva except one case in the buccal space [5] according to the literatures, as the peripheral counterparts, although occurring predominantly intraosseously. The term peripheral odontogenic keratocyst (POKC)

was first described by Dayan et al [6] in 1988, which was considered as a distinctive entity, different from gingival cyst of the adult (GCA). POKC was renamed as peripheral keratocystic odontogenic tumor (PKCOT) since OKC was renamed as KCOT. To our knowledge, only 17 cases of PKCOT were reported [5-7, 9-13, 15, 16] in the English literatures up to date, but without detailed analysis of imaging findings.

In this paper, we reported 2 cases of PKCOT localized in the lateral facial deep region (LFDR) and analyzed the clinical and imaging features of 17 cases documented in the literature.

Cases and methods

Case 1

A 44-year-old female patient was admitted with an asymptomatic nodule in the left soft palate and pharynx. Clinical examination revealed a

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Table 1. Summary of reported cases of peripheral keratocystic odontogenic tumor in the literatures

Author	Year	No of case reported	Age	Gender	Site	Imagiological Method	Imaging findings
Stoelinga et al [15]	1975	1	N.S	N.S.	maxillary gingiva	N.S.	N.S.
Buchner and Hansen [16]	1979	2	N.S.	N.S.	Buccal gingiva buccal gingiva	N.S. N.S.	N.S. N.S.
Dayan et al [6]	1988	1	42	M	Left maxillary gingiva	X-ray	N.C.
Chegade et al [10]	1994	5	37	M	Right mandibular gingiva	N.S.	N.S.
			66	F	Left maxillary gingiva	N.S.	N.S.
			70	M	Left mandibular gingiva	N.S.	N.S.
			57	F	Right maxillary gingiva	N.S.	N.S.
			42	M	Right posterior mandibular alveolar mucosa	N.S.	N.S.
Ide et al [7]	2002	2	38	F	Left mandibular gingiva	X-ray	N.C.
			46	F	Right mandibular gingiva	X-ray	N.C.
Chi et al [9]	2005	2	81	F	Left mandibular gingiva,	X-ray	N.C.
			64	F	Left maxillary gingiva	X-ray	N.C.
Rhonda et al [12]	2005	1	83	F	Left maxillary gingiva	X-ray	N.C.
Faustino et al [13]	2008	1	57	F	Left mandibular gingiva	X-ray	Superficial erosion or depression of the alveolar bone
Vij et al [11]	2011	1	56	M	Left maxillary gingiva	X-ray	N.C.
Grobe et al [5]	2012	1	52	M	Buccal space	X-ray	N.C.
						CT	Marginal contrasted, without bone involvement
						(enhanced)	
Present cases		2	69	M	Right LFDR	CT	Pre-C: abnormal lobular mass, depression of adjacent maxillary tuberosity. Post-C: slight marginal enhancement
					(involving buccal mucosa and pterygopalatine fossa)	(pre-and post-contrasted)	
					Left LFDR	CT	Pre-C: lobular, low density soft tissue mass, involving left lateral pterygoid plate, adjacent pharynx pushed inside. Post-C: slight marginal enhancement
						(pre-and post-contrasted)	
						MRI	Pre-C: low signal intensity on T1WI, high signal intensity on T2WI, homogeneous, well-defined
						(pre-and post-contrasted, fMRI)	Post-C: mild marginal enhancement
							Mean ADC: $1.29 \times 10^{-3} \text{ mm}^2/\text{s}$

F: female; M: male; N.S.: not stated; N.C.: no changes; Pre-C: pre-contrasted; Post-C: post-contrasted; fMRI: functional magnetic resonance imaging; ADC: apparent diffusion coefficient.

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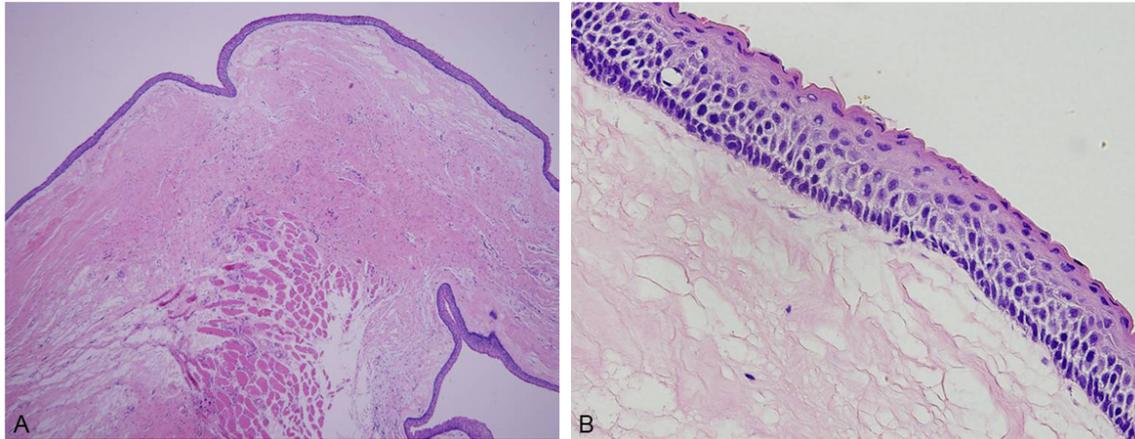


Figure 1. Histopathological findings. A. Photomicrography demonstrated a cyst with epithelial lining, the muscle was seen beneath the connective tissue, indicating extraosseous nature of the mass (HE, $\times 40$); B. Photomicrography of the cyst lining showed squamous epithelium with a parakeratotic layer (HE, $\times 400$).

solid non-mobile mass about 3 cm \times 4 cm in size, covered by normal oral mucosa. The mouth opening was unlimited, and there was no deviation of the mandible in habitual occlusion. Intraorally, the movement of tongue and secretion of salivary glands were normal. No enlarged lymph node was noted in the submandibular region and neck. Extensive resection of the mass in the left, left partial maxillectomy and reconstruction with submandibular gland flap were performed after CT scan, MR and routine lab examinations. Postoperative course was uneventful. The final histopathological diagnosis was KCOT of the left lateral facial deep region.

Case 2

A 69-year-old male was referred to an oral surgeon for painless swelling in the right cheek over 6 months. The swelling was initially noted accidentally without pain or numbness. The lesion was approximately 2 cm in diameter, solid, non-mobile, and covered by normal oral mucosa. Extensive resection of the mass in right skull base, reconstruction with sternocleidomastoid flap and buccal pad flap were performed after CT scan and routine lab examinations under general anaesthesia. Histopathological examination led to the diagnosis of KCOT of the right deep lateral face.

Bibliographic retrieval

Bibliography was retrieved in the PubMed database using the key words “peripheral”, “keratocystic”, “odontogenic”, “tumor”. Seventeen

cases of PKCOTs (**Table 1**), 16 in the gingiva and 1 in the buccal space, were found in the literature available. Eight cases in the gingiva had radiographic records, and 1 case in the buccal space had plain radiographic examination and CT scan.

Imaging methods

Imaging methods included plain radiography (periapical radiograph and pantomography), CT scan and MRI. The detail of CT scan in one case with KCOT in the buccal space was not provided. CT scan was performed using a 16-slice GE Lightspeed (General Electric Medical Systems, Milwaukee, WI, USA) in 2 cases with KCOT in LFDR. CT scans were acquired at 120 KV/250 mAs. Axial section of 5 mm thickness, reconstructed at 1.25 mm was conducted using a soft-tissue algorithm. A bolus intravenous injection of 50 ml (injection rate of 1.5 ml/s) of non-ionic contrast medium (300 mg/ml) was administered.

MRI scans were performed on a 1.5 Tesla GE Signa TwinSpeed system (General Electric Medical System, Milwaukee WI, USA), and a neurovascular array coil was used. Axial MR images were obtained by using T1-weighted spin echo sequence and T2-weighted fast spin echo sequence with or without fat suppression. Coronal MR images were acquired by using T2-weighted fast spin echo sequence with fat suppression. An intravenous injection of gadolinium contrast medium (0.1 mmol/kg body weight) was administered. In addition, function-



Figure 2. CT axial image showed an ill-defined marginal contrasted displaced mass in the left LFDR, and adjacent pharynx was pushed inward.



Figure 3. CT coronal image with bone window showed depression of the adjacent maxillary tuberosity near the PKCOT in the right LFDR (arrow).

al MRI series were performed including diffusion-weighted MR imaging (DWI).

Results

Nineteen cases were confirmed to be KCOTs by paraffin-embedded section pathology after operation (**Figure 1**). Clinical features of 19 subjects, including 2 cases reported in this study were as follows: the patients' age ranged from 37 to 83 years, with a mean of 56.5 years; the male to female ratio was 1:1.285, with no apparent predilection for either gender; the gingiva was definitely the more commonly site, accounting for 84.2%, and buccal space and LFDR were rare original sites for PKCOT, accounted for 5.2% and 10.5%, respectively.

Imaging features of gingival PKCOT

Eight out of 16 cases with gingival PKCOT had radiograph records. Seven of them showed no apparently radiographic changes and only one showed slight depression in the adjacent alveolar crest beneath the lesion.

Imaging features of PKCOT in the buccal space

One case with PKCOT in the buccal space had radiography and CT scan. No significant changes were found in the pantomography. CT scan showed a marginal contrasted displaced oval mass in the area of the right cheek without bony involvement.

Imaging features of PKCOT in LFDR

Two cases with PKCOT in LFDR underwent CT scanning, and MRI was also performed in one case. Axial soft-tissue window CT images showed two lesions in the right and left LFDR respectively. The mass was lobular, with linear-septa, and ill-defined boundary with surrounding soft tissues. PKCOTs presented as homogenous low density in pre-contrast CT images, and minimal marginal enhancement in post-contrast CT images, without enhancement in the vesicular space. The left pharynx was pushed inward and the airway became narrow (**Figure 2**). Depression of maxillary tuberosity was demonstrated in the bone window CT images (**Figure 3**) in the right LFDR, displacement and resorption of the pterygoid plate was found in the left one. MRI showed the lesion in the left LFDR was homogenous and hypointense on T1-weighted images, homogenous and hyperintense on T2-weighted fat-suppression images, and only slightly heterogeneous marginal enhanced after injection of gadolinium contrast medium (**Figure 4**). The result of DWI was, when $b=1000$, the mean apparent diffusion coefficient (ADC) was $1.29 \times 10^{-3} \text{ mm}^2/\text{s}$.

Discussion

The primary site of PKCOTs reported before 2012 was gingiva, and controversy existed in regarding to the terminology. Some authors [7,

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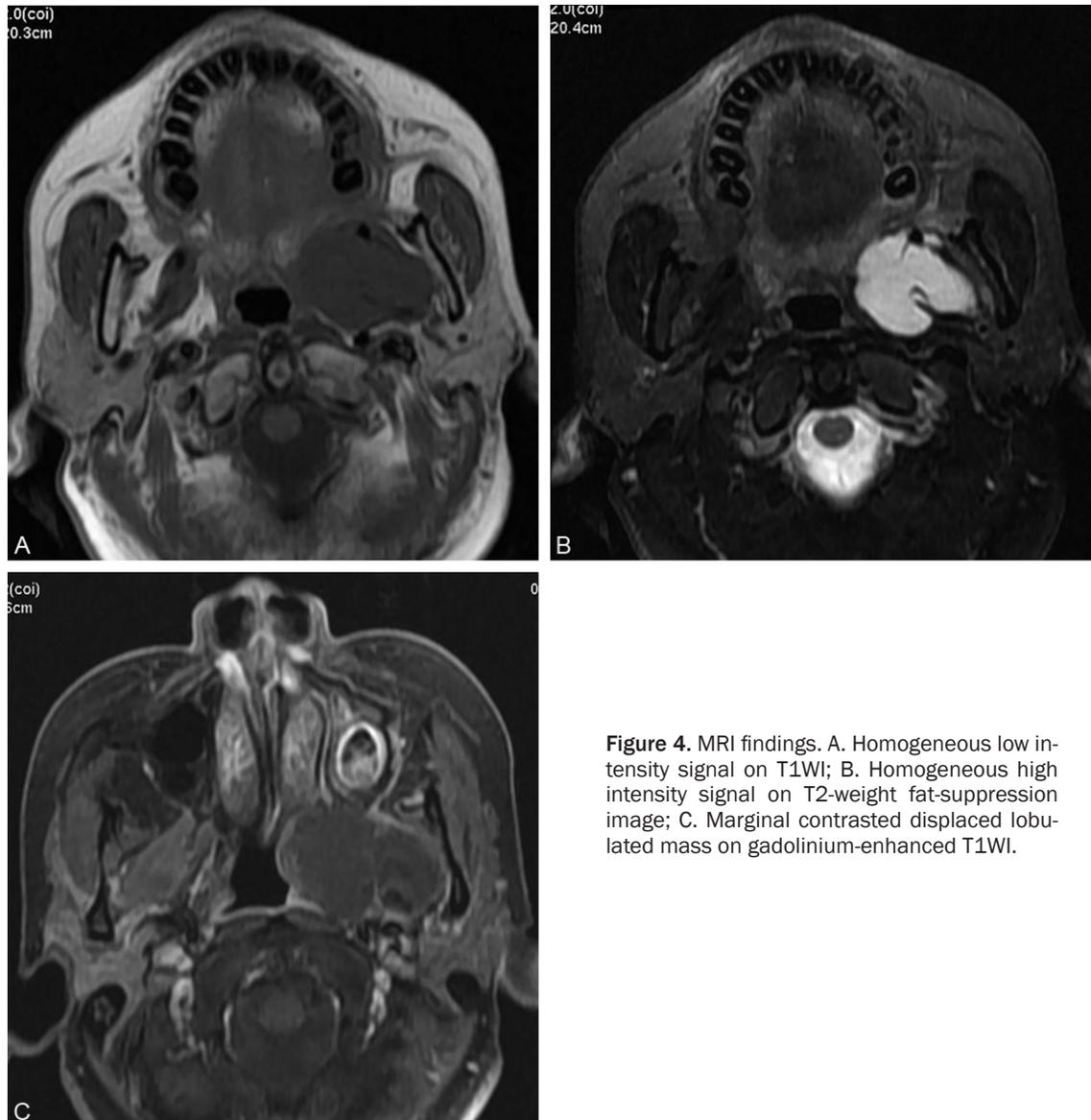


Figure 4. MRI findings. A. Homogeneous low intensity signal on T1WI; B. Homogeneous high intensity signal on T2-weight fat-suppression image; C. Marginal contrasted displaced lobulated mass on gadolinium-enhanced T1WI.

8] believed that PKCOT was the variant of GCA due to the same clinical behaviors, while many authors [9-12] believed that PKCOT was not a variant of GCA, but a counterpart of intraosseous KCOT, because of characteristics of parakeratinization, recurrence, and sometimes with nevoid basal cell carcinoma syndrome. The recently reported sites of PKCOT included the buccal space and LFDR away from gingival, supporting the concept that PKCOT is an isolated entity distinct from GCA.

Role of plain film in diagnosis of PKCOT

Periapical and panoramic radiographs are usually taken when PKCOTs located in the gingiva.

PKCOT is an extraosseous lesion within the soft tissues. Although slight saucer-shape superficial resorption of the bone underlying the PKCOT in the gingival can be noted during surgical excision, no apparent change was demonstrated in the plain radiograph. Only PA in 1/9 cases showed mild resorption of the alveolar crest beneath the lesion due to long-time compression. The positive rate of plain radiograph in diagnosis of PKCOT is only 11.1%.

Role of CT in diagnosis of PKCOT

Contrary to the low detection of plain radiograph, the positive rate of CT in diagnosis of PKCOT is 100%. CT has lower space resolution

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but is more sensitive in revealing bone shape than radiograph which can display trabecular bone pattern clearly. In PKCOT, it is more important to demonstrate the change of bone shape than the trabecular bone pattern change. CT can also reveal the relationship between the lesion and adjacent hard or soft tissues after multiplanar reconstruction. The lesion was represented as a slightly marginal enhanced mass with liquid radiodensity of 20-30HU (both for pre- and post-contrast) in CT images. By measuring the CT value, the lesion can be defined as cystic. Bone shape change in the early stage and relationship with the adjacent tissues can be found in CT images.

Role of MRI in diagnosis of PKCOT

MRI provides good contrast between different tissues and more detailed information than CT images, especially for the soft tissues. Many tissues, such as fatty tissue and ^1H -rich tissue (corpora vitreum, cerebrospinal fluid et al) have their unique features on MR image. Fatty tissue is of high signal intensity on T1 and T2 image and without change on T1 post-contrasted image, and ^1H -rich tissue is of high signal intensity on T2 weighted-image, low signal intensity on T1WI and T1WI after contrasting. The initial diagnosis of 2 cases presently reported led to a cyst based on the same MR image features as the ^1H -rich tissue and encapsulated displaying. Functional MR imaging including MR spectroscopy, dynamic contrast-enhanced MRI, diffusion-weighted MR imaging (MR-DWI) has progressed rapidly in recent years, offering more molecular biological evidences for diagnosis. Wang et al [14] reported the mean ADC of malignant lymphomas was significantly smaller than that of carcinomas, and the same between carcinomas and benign solid tumors, benign solid tumors and benign cystic lesions. When ADC was less than $1.22 \times 10^{-3} \text{ mm}^2/\text{s}$, malignant lesion should be suspected. The mean ADC of PKCOT in the left LFDR in our case was $1.29 \times 10^{-3} \text{ mm}^2/\text{s}$, suggesting a benign tumor, possibly cystic in nature. However, MRI is inferior to CT in demonstrating early bone changes, therefore, CT scan may be considered in some cases when bone changes was suspected.

Differential diagnosis of PKCOT

The differential diagnosis of PKCOT includes venous malformation, macrocystic lymphatic

malformation, epidermoid cyst, and mucoepidermoid carcinoma. Venous malformation and macrocystic lymphatic malformation are vascular anomalies, with irregular and serpentine shape. The presence of phleboliths is pathognomonic for low-flow venous malformation. Some venous malformations can present as a mild enhancement lesion in delayed phase. PKCOT presents as a circular (with liquid tension inside) slight marginal enhanced lesion. However, mucoepidermoid carcinoma may display as a cystic lesion similar to a cyst, but still have solid component which can be heterogeneous density in enhanced scan. Differential diagnosis can be difficult between PKCOT and cysts originating from ectopic minor salivary glands because of the similar imaging findings. In these circumstances, fine needle aspiration cytology may help to make diagnosis.

Conclusion

LFDR is rare original site for PKCOT. CT and MRI, especially the latter, can provide important information in diagnosis of PKCOT, which is usually presented as a cyst in soft tissues. Conventional radiographs have limited value in the diagnosis due to the location of the tumor. PKCOT should be considered when a cystic lesion was found in oral and maxillofacial soft tissues on CT and MR imaging.

Disclosure of conflict of interest

None.

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