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RETROSPECTIVE ANALYSIS OF KERATOCYSTIC ODONTOGENIC TUMOUR IN A COHORT OF SRI LANKAN PATIENTS AND REVIEW OF LITERATURE.

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ABSTRACT

Introduction: Keratocystic Odontogenic Tumour (KCOT) represent 4-10% of all odontogenic tumours and 5% of those cases are involved in nevoid basal cell carcinoma syndrome (NBCCS). **Objectives:** To investigate the patterns and behavior of KCOT and its association with Gorlin-goltz syndrome in a cohort of Sri Lankan patients. **Materials and Methods:** A retrospective study was conducted using the cases selected from the archives from 2003 to 2016 Orthokeratinized odontogenic cysts were excluded from the study. The data were presented as means and proportions. Associations were assessed with Fisher's exact test. **Results:** A significant male predilection was observed (P= 0.033). The male to female ratio is 1.32:1. Age ranged from 7-84 years and the mean age was 35.46 years. Mandible (77.1%) was significantly affected than maxilla (22.1%) (P= 0.001) and only 0.8% occurred in both jaws. **Conclusion:** The present cohort of KCOTs is the first study in Sri Lanka and some data in par with English literature. As our data indicated high rate of malignant transformation, it is necessary to follow these patients regularly.

KEYWORDS: KCOT, NBCCS, Sri Lanka.

INTRODUCTION

Keratinizing jaw cysts are relatively uncommon but very important because of its strong tendency of recurrence and aggressive behavior. They are mainly divided in to two histological subtypes, as parakertinized and orthokeratinzed. Parakeratinized cysts were previously named as odontogenic keratocysts that showed high recurrence rate (Bhargava et al., 2012). Odontogenickeratocyst, the 2nd most common developmental odontogenic cyst, was first described by Philipsen et al in 1956 and its clinical and histologic features were confirmed by Browne et al in 1971.(Abdullah, 2011, Barnes et al., 2005) At that time "Odontogenic Keratosis" and "Primodial Cyst" terms were used to denote the cysts with specific histopathological features and clinical behavior, which was believed to arise from dental lamina (Grasmuck and Nelson, 2010). It was believed that these are benign but potentially aggressive and recurring. However after wide range of researches it was identified as a tumor with the features of a benign uni or multicystic, intra osseous odontogenic origin, characteristic parakeratinized stratified squamous epithelial lining, potentially aggressive and infiltrative, high recurrence, intrinsic growth potential, involvement with mutation in PTCH, a tumor suppressor gene and associated with nevoid basal cell carcinoma syndrome (NBCCS). Accordingly, World Health Organization (WHO) classified and renamed this condition as Keratocystic Odontogenic Tumour (KCOT) in 2005 (Barnes et al., 2005).

KCOT represent 4-10% of all odontogenic tumours and 5% of those cases are involved in NBCCS (Grasmuck and Nelson, 2010). It may found in patients from infancy to old age, mostly occur in 2nd and 3rd decades. There is a slight male predilection. Generally it is intra-osseous but rarely peripheral cases have been reported. The mandible is mostly involved and can be unicystic or multicystic. It extends from anterior to posterior direction causing bone expansion. The occurrence in maxilla is rare (Abdullah, 2011, Barnes et al., 2005, Bhargava et al., 2012). The small KCOTs are usually asymptomatic but larger ones may be associated with pain, swelling or drainage. Histopathologically it shows parakeratinzed epithelium with basal cell palisading. Treatment of KCOT may vary from enucleation to peripheral ostectomy, application of carnoy's solution (60% ethanol, 30% chloroform and 10% glacial acetic acid), decompression and dredging or combined therapies (Abdullah, 2011, Grasmuck and Nelson, 2010).

The Gorlin-goltz syndrome, also known as NBCCS is an infrequent multisystemic disease inherited in an autosomal dominant way, which shows a high level of penetrance and variable

expressiveness (Joshi et al., 2012). It has a classic triad composed of KCOT in the jaw, multiple basal cell nevi carcinomas and skeletal abnormalities. This syndrome is infrequently diagnosed with routine radiographic exams of jaw bones for various dental procedures, since multiple KCOTs are usually one of the first manifestations of the syndrome(Jawa et al., 2009). This study aims to retrospectively investigate the patterns and behavior of KCOT and its association with Gorlin-goltz syndrome in a cohort of Sri Lankan patients.

MATERIALS AND METHODS

A retrospective cross sectional analytical study was conducted at the Departments of Oral and Maxillofacial Surgery and Oral Pathology, Faculty of Dental Sciences, University of Peradeniya. Ethical approval was obtained from the Ethics Review Committee, Faculty of Dental Sciences, University of Peradeniya. Patients consent was also taken before the surgery. Cases were selected from the archives from 2003 to 2016. Orthokeratinized odontogenic cysts were excluded from the study. Clinical data and diagnosis were collected from patients' records and pathology reports. The data were evaluated using SPSS 20.0 statistical software and presented as means and proportions. Associations were assessed with Fisher's exact test.

RESULTS

Out of 270 patients who has fulfilled inclusion criteria 154(57%) were male patients and 116(43%) were female patients. A significant male predilection was observed (Fisher's exact test: P= 0.033). The male to female ratio is 1.32:1. Age ranged from 7-84 years and the mean age was 35.46 years. Mandible (77.1%) was significantly affected than maxilla (22.1%) (Fisher's exact test: P= 0.001) and only 0.8% occurred in both jaws. Considering the anatomy, right side showed significant preponderance (48.3%) compared with left side (43.9%) (Fisher's exact test: P = 0.054).

Five cases were presented with multiple KCOT and out of them three cases were associated with Gorlin Goltz Syndrome (1.1% from total sample). Histopathologically eight cases showed morphological variations. Out of them, three KCOTs showed dysplastic changes ranging from mild to moderate epithelial dysplasia. Four cases showed squamous cell carcinoma developing from pre-existing KCOTs and one case has been transformed in to conventional ameloblastoma.

DISCUSSION

KCOTs accounts for 4%-10% of odontogenic tumours and show high recurrence rate. It shows an adult male preponderance with a peak in 2nd to 3rd decades of life and average age is 30.8 years (Bhargava et al., 2012, Grasmuck and Nelson, 2010, Jing et al., 2007). In our series there is a prominent male predilection and the mean age is also little higher. Mean age for multiple KCOTs and KCOTs associated with NBCCS is less than that for single non recurrent KCOTs. Mandible is affected more than the maxilla, with 50% affecting the angle of the mandible (Jawa et al., 2009). Cases reported here also showed the tumour mostly occurred in the mandible. Rare peripheral type of KCOT has been found in gingiva and alveolar mucosa of the canine and premolar regions (Jing et al., 2007). The tumour usually doesn't show any symptoms unless concomitant inflammation cause swelling, pain and sometimes even discharging (Grasmuck and Nelson, 2010).

Radiologically this tumour characterized by a well-circumscribed radiolucency with smooth radiopaque margins. It could also be either unilocular or multilocular with scalloped borders in which smaller lesions are unilocular. About 40% of cases showed dentigerous relationship[4]. Although root resorption and displacement of adjacent teeth are rare events, larger lesions might displace the teeth (Abdullah, 2011, Bhargava et al., 2012, Jing et al., 2007). Certain KCOTs can reach a significant size before being noticed by the patient and penetrate cortical bone to involve adjacent structures. This is due to antero-posterior extension along the medullary cavity without bucco-lingual expansion (Abdullah, 2011, Bhargava et al., 2012).

According to the literature 5% of KCOT can be associated with Gorlin-Goltz syndrome.(Jawa et al., 2009) Cases with multiple cysts/tumours should be investigated for NBCCS. Features of this syndrome includes multiple basal cell carcinomas, bifid ribs, calcification of falx cerebri, frontal bossing and macrocephaly. The definitive diagnosis mainly depends on its histopathological features (Joshi et al., 2012). The present series also have 5 cases with multiple lesions and out of them 3 cases were syndromic (1.1%) which was less compared with the literature. Histopathologically it shows a thin, friable wall, which is often difficult to enucleate from the bone in one piece. The epithelial lining is composed of a uniform; 6-8 cells thick parakeratinized stratified squamous epithelium (Jawa et al., 2009, Joshi et al., 2012).

The ideal treatment option for KCOT still remains a controversial. However due to high rate of recurrence the choice of treatment should be the most suitable one which depends on several factors such as age, size of the lesion, localization, cortical perforation and relationship with surrounding structures (Abdullah, 2011).

KCOT has a relatively high recurrence rate that may varies from 5%-62% (approximately 30%) particularly high in multiple cysts and those in the posterior body and ascending ramus. The reasons for this high recurrence rate have been identified as presence of thin, fragile linings, finger-like cyst extensions into cancellous bone and presence of satellite (daughter) cysts in the wall. Rapid proliferation of epithelium, formation of additional cysts from other dental lamina remnants and inferior standards of surgical treatment could also prone to have recurrences (Abdullah, 2011, Bhargava et al., 2012, Wushou et al., 2014) . Therefore post-operative follow up is essential for at least 5 years after surgery. Malignant transformation is rare (0.46%) and there are cases transform in to ameloblastoma (Morgan et al., 2005). This series also report 4 cases that transformed in to squamous cell carcinomas (1.5%) and one case in to conventional ameloblastoma. Some data of our study is in par with the literature and some showed higher preponderance such as site, mean age and rate of malignant transformation. However long term follow-up is very difficult in our society in order to assess accurate recurrence rate and data related to malignant transformation.

CONCLUSION

The present cohort of KCOTs is the first study in Sri Lanka and some data in par with English literature. As our data indicated high rate of malignant transformation, it is necessary to follow these patients regularly. Association of NBCCS also should be investigated if the patient presented with multiple lesions.

CONFLICT OF INTEREST

Authors declare that there are no conflict of interest or what so ever.

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