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Case report: multiple keratocystic odontogenic tumour in a non-syndromal pediatric patient

ABSTRACT

Background *Keratocystic odontogenic tumour (KCOT) is an odontogenic tumour which stems from the odontogenic organs mostly localised in the lower jaw, particularly posterior body and ascending ramus of the mandible. The majority of these tumours are single lesions. When detected in the jaw in multiple forms, these cysts are seen in association with Gorlin Goltz/Basal cell naevus syndrome. However a few cases of non-syndromal multiple keratocystic odontogenic tumour have been reported in the literature.*

Case report *We report a case of multiple keratocystic odontogenic tumour in a 13-year-old girl demonstrated by panoramic radiography and cone beam computed tomography (CBCT). The differential diagnosis, treatment and imaging modalities are also discussed.*

Keywords *Keratocystic odontogenic tumours; Non-syndromal.*

Introduction

The keratocystic odontogenic tumour is an epithelial developmental tumour of the jaws [Marx and Stern, 2003]. The lesion is commonly found in the maxilla and mandible, and can become quite large because of its potential for significant expansion, extension into adjacent tissues, and rapid growth [Scharffetter, 1989]. According to the World Health Organization's (WHO) 1992 classification [Kramer et al., 1992], odontogenic cysts are typed under developmental (dysembryogenetic) cysts. In 2005, a WHO working group categorised odontogenic keratocyst (OKC) as an odontogenic tumour and it is now designated as keratocystic odontogenic tumour (KCOT) [Barnes et al., 2005]. It is defined as "a benign uni- or multicystic, intraosseus tumour of odontogenic origin, with a characteristic lining of parakeratinised stratified squamous epithelium and potential for aggressive, infiltrative behaviour" [Borgonovo et al., 2011]. OKC has a multicentric, infiltrative growth pattern and is locally infiltrative, many studies report cases of cortical bone penetration with extension into adjacent soft tissues [Myoung et al. 2001; Shear and Speight, 2007]. Recurrence rates are variable, ranging from 2.5% to 62.5% [Browne, 1975; Forssell, 1980; Voorsmit et al., 1981; Forssell et al., 1988; Mendes 2010].

The majority of these tumours are single lesions. Multiple KCOTs are mostly seen with Gorlin Goltz/Basal cell naevus syndrome. However, though rarely, a few cases of non-syndromal multiple keratocystic odontogenic tumour have been reported in the literature [Myoung et al., 2001; Morgan et al., 2005; Tolstunov and Treasure, 2008; Iatrou et al., 2009; Bartake et al., 2011; Borgonovo et al., 2011]. Hence, it was considered worthwhile to report an unusual presentation of a multiple KCOT in a 13-year-old girl demonstrated by panoramic radiograph and cone beam computed tomography (CBCT); differential diagnosis, treatment and imaging modalities are also discussed.

Case report

A 13-year-old girl was referred to our outpatient clinic for assessment of her dental status. She presented with pain and swelling of the left and right molar region of the mandible. Her general medical history was unremarkable, without any syndromes. Her dental history revealed missing teeth from the age of 7.

After an intra-oral and radiographic examinations, it was discovered that the patient had multiple cystic lesions (Fig. 1). A biopsy was performed from the left side of the mandible and the result was subsequently reported as keratocystic lesions. A surgery was advised but the patient's parents refused at the time, agreeing to have her assessed annually. Panoramic images were obtained at the following visits which showed impacted left and right maxillary canines and displaced germs of mandibular and maxillary second molars (Fig. 2).

The follow-up radiographs showed dramatic

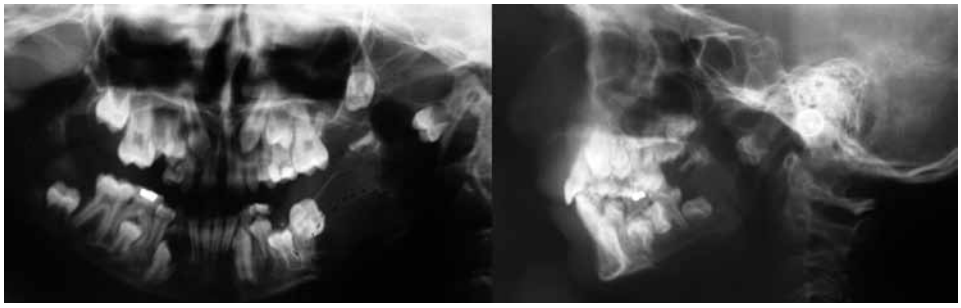


FIG. 1

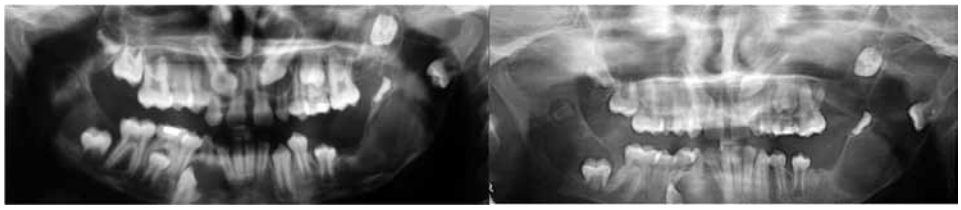


FIG. 2

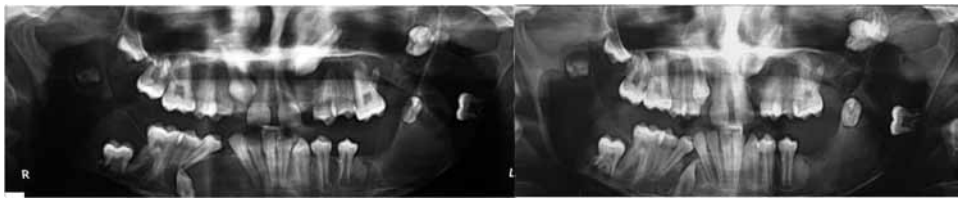


FIG. 3

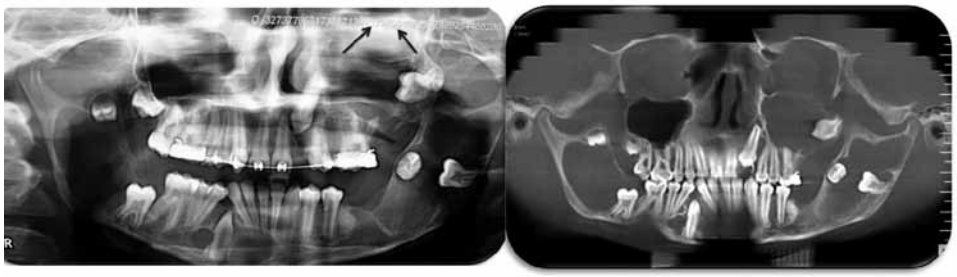
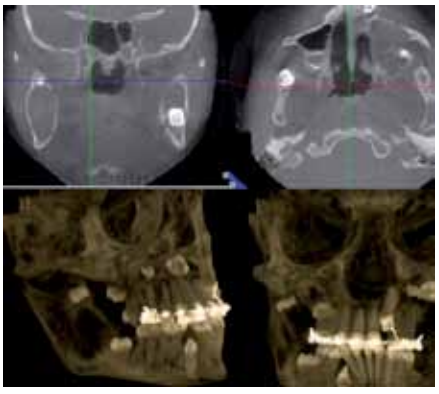


FIG. 4

expansion of the radiolucent lesions and similarly displaced germs of maxillary and mandibular third molars (Fig. 3).

In the fifth year of her assessment, the patient was referred for orthodontic consultation regarding eruption of the maxillary canines. Upon examination, the patient complained of the same symptoms as at the first visit, with pain and swelling of both left and right molar region in the mandible. The panoramic radiograph revealed multiple radiolucent lesions in maxillary and mandibular molar teeth and also the impacted left maxillary canine, and the right mandibular canine with large expansive lesions. Moreover, displaced teeth and germs were noticed both in left and right mandibular molar and left maxillary molar regions (Fig. 4). Consecutively, CBCT scans were performed to obtain a more precise location and definition of the pathologic features of the jaws. 3D imaging demonstrated multiple unilocular radiolucent lesions and the 3D surface rendering showed the exact location of these impacted teeth and lesions (Fig. 5). Surgical treatment was planned for the removal of the lesions, but the patient and her parents again refused treatment.

FIG. 5



When the patient visited our clinic after 2 years, moderate changes were seen. A new clinical and radiologic examination showed that the maxillary left canine tooth was erupted and the germs of left mandibular mmolar teeth were extracted. However, the large expansible radiolucent lesions were still seen on the panoramic radiograph, especially an enlarged area was detected on mandibular right molar region (Fig. 6).

This time the surgical treatment plan was accepted

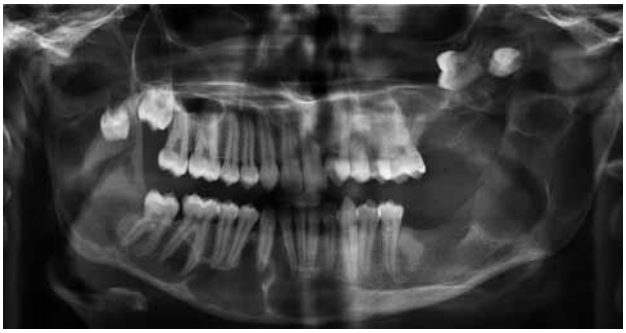


FIG. 6

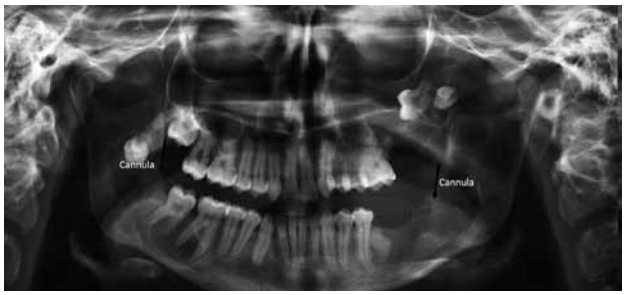


FIG. 7

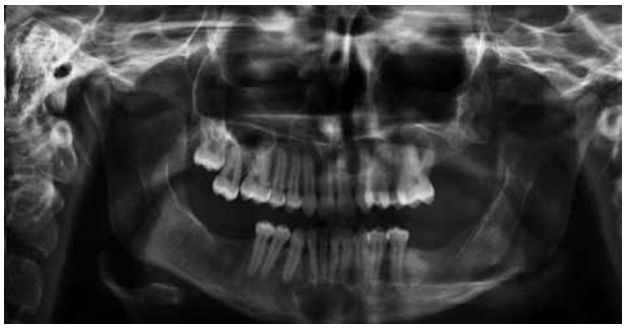


FIG. 8

by the patient. First, enucleation and curettage were performed in the mandibular anterior area. In order to avoid the risk of mandibular fracture or damaging important anatomical structures, we performed marsupialisation in the mandibular left and right ramus area. Three months later, a new marsupialisation was performed in the maxillary left molar area. During thi period, cannulas were used to keep the sites open on both sides. Follow-ups were made twice a week which involved clinical examination and irrigation only. Marsupialisation continued for almost 15 months until the size of the keratocysts were reduces (Fig. 7), followed by separate enucleation of all lesions with curettage under general anaesthesia, using nasotracheal intubation. Mandibular right first, second and third molar teeth and maxillary left second and third molars had to be extracted during this phase (Fig. 8). The histopathologic examination confirmed the diagnosis of keratocystic odontogenic tumours.

The patient was recalled periodically for clinical and radiographic examinations every 6 months. After 12 months, the lesions in the mandibular and maxillary anterior regions recurred and noted approximately to be 1-2 cm in diameter (Fig. 8). As these lesions are small and easily accessible, their enucleation under general anaesthesia has recently been performed.

Discussion

Cystic lesions of the jaws may be epithelial or non-epithelial, odontogenic or non-odontogenic, developmental or inflammatory in origin [Killey et al., 1977]. OKC is a developmental odontogenic cyst, first described by Mikulicz and introduced by Philipsen in 1956 [Philipsen, 1956], originating either from epithelial rests of the dental lamina or basal cell extensions from the overlying oral epithelium [Voorsmit et al., 1981; Browne, 1975; Shear and Speight, 2007]. The lesion is commonly found in the maxilla and mandible, and can become quite large because of its potential for significant expansion, extension into adjacent tissues, and rapid growth [Scharffetter, 1989].

Odontogenic keratocysts, reclassified by the WHO as keratocystic odontogenic tumours [Cottom et al., 2012]. This more accurately reflect its aggressive, often infiltrative behaviour and potential for recurrence [Philipsen, 2005; Mendes et al., 2010; Li, 2011]. The reclassification in terminology remains controversial, and as yet, no universal consensus has been reached regarding the cyst's name or its neoplastic characteristic [Shear and Speight, 2007; Vargas et al., 2007].

The KCOT has a multicentric and infiltrative growth pattern and is locally infiltrative with numerous studies documenting cases of cortical bone penetration and extension into adjacent soft tissues [Myoung et al., 2001; Shear and Speight, 2007]. Recurrence rates are variable and are reported to range from 2.5% to 62.5% [Browne, 1975; Forssell, 1980; Voorsmit et al., 1981; Forssell et al., 1988; Mendes et al., 2010]. Diagnosis of cystic lesions is based on clinical and radiologic findings [Iatrou et al., 2009]. Clinically, they may be asymptomatic or may have acute or chronic findings. In the latter case, pain and swelling may be the presenting symptom [Bodner et al., 1996]. Their size and location are evaluated by routine radiographs, computerised tomography (CT), and 3-dimensional CT [Bodner et al., 1996]. Fine-needle aspiration and biopsies may assist the diagnosis in selected cases [Pogrel, 2007]. Histological examination usually clarifies the type and origin of the lesion and helps the clinician to decide the best treatment for the patient [Iatrou et al., 2009].

KCOT occur three times more often in the mandible than the maxilla and is most frequently seen in the canine to premolar, mandibular retromolar, ramus areas, and the region of the maxillary second permanent molar,

respectively [Graham et al., 1968]. In children and adolescents the cysts may cause displacement of the developing teeth [Gorlin, 1987], and delayed dental development has been reported [Rosenblum, 1998] as in our case.

Some odontogenic tumours have radiological and histological features similar to KCOT which must be differentiated. Ameloblastoma is the most common tumour in this category, that can behave similarly to the dentigerous cyst, and its unicystic variant, that is most prevalent in adolescence and can be seen as early as the age of 5 years [Dunsche et al., 2003]. Dentigerous cysts are another type of lesions which are also known to occur in association with other pathologic odontogenic entities such as mucoepidermoid carcinoma, ameloblastoma and squamous cell carcinoma [Yasuoka et al., 2005].

The treatment of KCOT remains controversial. The treatments available are generally classified as conservative or aggressive. Conservative treatment generally includes simple enucleation, with or without curettage, using spoon curettes or marsupialisation. Aggressive treatment generally includes peripheral ostectomy, chemical curettage with Carnoy's solution, and resection [Marx and Stern, 2003; Morgan et al., 2005]. Following enucleation of these cysts, the recurrence rate is high; estimated to be 30–60% [Donatsky and Hjørting-Hansen, 1980].

Multiple KCOTs commonly associated with Nevoid basal cell carcinoma syndrome, also known as Gorlin-Goltz syndrome was defined in 1960 by Gorlin and Goltz [1960]. The syndrome affects multiple organ systems, which comprises as major findings, multiple basal cell carcinoma, odontogenic keratocysts, palmar/plantar pits, calcification of the falx cerebri, spine and rib anomalies [Ortega et al., 2008]. However, absence of a syndrome with multiple KCOTs is extremely rare as in our case.

Conclusion

In conclusion, this paper presents a case of KCOT which was treated by marsupialisation followed by enucleation in a paediatric patient. A multiple disciplinary approach should be considered while treating such cases. It should also be concluded that 3D CBCT imaging is beneficial for the diagnosing of multiple KCOTs with precise anatomical detail and 3D rendering.

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