CASE REPORT

Orthokeratinised odontogenic cyst mimicking periapical cyst

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SUMMARY

Orthokeratinised odontogenic cyst (OOC) denotes the odontogenic cyst that microscopically has an orthokeratinised epithelial lining. OOC is characterised by a less-aggressive behaviour and a low rate of recurrence. This report describes a case of OOC involving posterior part of the mandible that mimicked periapical cyst in a 14-year-old boy. The initial clinical diagnosis was given as periapical cyst based on the clinical and radiographical features. Enucleation of the cyst was performed and the specimen was sent for histopathological examination. A definite diagnosis of OOC was made by histopathological examination of the biopsy specimen. This case emphasises on including OOC in the differential diagnosis of radiolucencies occurring in the periapical region of non-vital tooth.

BACKGROUND

Odontogenic keratocysts (OKCs) are developmental odontogenic cysts of epithelial origin, first identified and described in 1876 and further characterised by Philipsen in 1956.¹ In earlier literature, a keratocyst was described as a Cholesteatoma.² Now designated by the WHO as a keratocystic odontogenic tumour (KCOT), it is defined as “a benign uni- or multicystic, intra-osseous tumour of odontogenic origin, with a characteristic lining of parakeratinised stratified squamous epithelium and a potential for aggressive, infiltrative behaviour.” The WHO “recommends the term KCOT as it better reflects its neoplastic nature.”³ The OKC is known for its high recurrence rate, aggressive behaviour and its occasional association with the naevoid basal cell carcinoma syndrome. Three histological variants were recognised initially: a parakeratinised variant, an orthokeratinised variant, and combination of the two.^{4–5} The less-aggressive clinical behaviour and recurrence pattern of the orthokeratinised variant warrants the designation of the orthokeratinised variant as a separate entity, ‘orthokeratinised odontogenic cyst’ (OOC) comprises approximately 17% of all keratinising jaw cysts⁶ and was first clearly identified by Wright in 1981.^{7–8} The KCOT’s incidence is more than eight times that of the OOC, reflecting the 10% proportion of OOCs of the formerly known OKC.⁹

The purpose of this article is to present the clinical and histopathological profile of a young male patient with OOC that mimicked periapical cyst.

CASE PRESENTATION

A 14-year-old male patient reported to the department of paedodontics the swelling in the left side

of the lower jaw for the past 4 months (figure 1). It was initially small in size and has grown over the past 4 months to reach the present dimension. There was no pain associated with the swelling. He had no history of trauma or tooth extraction.

On extra-oral examination, a diffuse swelling was present in the left side of the mandible region, of about 3×3 cm in size, extending superiorly 2 cm below the buttress region to the lower border of mandible inferiorly, medially 1 cm away from the left commissure of lip to left posterior border of the ramus region laterally. The swelling was single, diffuse, non-tender and hard in consistency. The skin over the swelling was normal. There was no discharge from the swelling. Mouth opening was normal with interincisal distance of 42 mm.

On intra-oral examination, root stumps of 36 and 46 were present. A diffuse, mild swelling was present in relation to the left lower buccal vestibule region. Obliteration of lower left buccal sulcus was present. The swelling extended from 34 to 37 region and it measured approximately 4×4 cm in size. It was non-tender, non-fluctuant, hard in consistency and fixed to underlying structures. The buccal cortical bone expansion was present.

INVESTIGATIONS

An orthopantomograph of the patient revealed unilocular radiolucency with a sclerotic border measuring approximately 4.5×3.5 cm, involving the left lower border of the mandible, extending from 34 to 37 region. The radiolucency was associated with the root stump of 36 (figure 2). A haematological examination was performed and all the blood parameters were normal. A surgical enucleation of the

Figure 1 Swelling on the left posterior mandibular region.

To cite: Rajalakshmi R, Sreeja C, Vijayalakshmi D, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-200883

Figure 2 Pre-operative radiograph showing a unilocular radiolucency associated with the periapical region of 36.

cyst was performed and the specimen was sent for histopathological examination (figure 3). The H&E stained section under low-power showed a cystic lesion composed of a thin, stratified squamous lining epithelium that showed an orthokeratotic surface and a prominent stratum granulosum layer (figure 4). High-power view showed absence of basal cell palisading. The epithelial-connective tissue interface appeared flat with absence of rete ridges. The cystic lumen showed flakes of keratin and RBCs (figure 5). The connective tissue showed mild inflammatory cell infiltrate. Considering these features, the lesion was finally diagnosed as OOC.

DIFFERENTIAL DIAGNOSIS

The general differential diagnosis of periapical radiolucencies includes radicular cyst, residual cyst, fissural cyst, dentigerous cyst and tumour-like ameloblastoma. But in our case, there was no history of tooth extraction or previous surgery which excluded the possibility of a residual cyst. Location of the cyst and absence of impacted-tooth ruled out the other differential diagnosis like fissural cyst and dentigerous cyst, respectively. With regard to the size and location of the lesion ameloblastoma, OKC and radicular cyst were considered in the differential diagnosis.

TREATMENT

OOCs show little if any tendency to recur and may be treated conservatively by simple enucleation with or without curettage. In patients with developmental odontogenic cysts with secondary infection, treatment with antibiotics to eradicate the infection should be initiated prior to definitive surgical management of the cyst. Similarly, the present patient was subjected to

Figure 3 Surgical enucleation of the cyst.

Figure 4 Low-power view showing a cystic lesion lined by a thin, stratified squamous lining epithelium which showed an orthokeratotic surface and a prominent stratum granulosum layer.

intravenous antibiotic therapy and was treated by extraction of 36 followed by enucleation of the cyst. Intermaxillary fixation was carried out for 2 weeks to avoid iatrogenic fracture during the initial period of healing (figure 6).

DISCUSSION

The 2005 edition of WHO reclassified the parakeratotic-type as a KCOT and stated that 'Cystic jaw lesions' that are lined by orthokeratinising epithelium do not form part of the spectrum of KCOT.¹⁰ OOC is characterised by a less-aggressive behaviour and a low rate of recurrence.^{11 12} OOC can occur in an age range from infancy to old age with the largest proportion at their third decade. Males are more frequently affected for most decades, but in the second decade a female predominance is present, which may be associated with the menarche.⁹ They most commonly occur in the mandible especially in the posterior sextants.¹³

The present case is a male patient in his second decade of life with the lesion in the posterior sextant of mandible.

Most of the patients present with pain and swelling or it can be asymptomatic and identified as an incidental finding and associated with unerupted teeth.^{9 13 14} Approximately 75% of OOCs are associated with impacted teeth, clinically and radiographically mimicking a dentigerous cyst.¹⁵ OOC may present

Figure 5 ×40 View of the section showing a cystic cavity lined by a thin, stratified squamous lining epithelium, with an orthokeratotic surface, prominent stratum granulosum and absence of basal cell palisading. The epithelial-connective tissue interface appears flat, devoid of rete pegs. The cystic lumen shows flakes of keratin and RBCs.

Figure 6 Postoperative radiograph.

as a unilocular or multilocular radiolucency with most of the cases being unilocular. Ninety-three percent cases presented with unilocular radiolucency in the systematic review of OOC⁹ and the diameter of these cysts commonly ranges from 2 to 7 cm.^{11–13} OKC can be classified as follicular or extrafollicular-type based on its relationship to the associated tooth.^{16–17} Wright and Wysocki¹⁸ emphasised that OKC should also be included in the differential diagnosis of radiolucencies occurring in the periapical region of non-vital tooth. Onuki *et al*¹⁹ reported a case of OOC which was suspected to be a radicular cyst. Cynthia *et al*²⁰ highlighted the importance of including OOC in the differential diagnosis of developmental odontogenic cysts with secondary infection. Vuhahula *et al*¹³ found that reduced enamel epithelium that had completed its tooth-forming function had the capability to keratinise under appropriate stimuli. Our case also presented with a painless-swelling of the posterior mandible associated with infected teeth with a radiographic finding of unilocular radiolucency measuring 4.5×3.5 cm.

The higher recurrence rate, aggressive behaviour and neoplastic potential of OKCs highlight the importance to distinguish between OKCs and OOCs. The histogenesis of KCOTs and OOCs may vary and needs further investigation. Histologically,

Learning points

- ▶ Orthokeratinised odontogenic cyst (OOC) should also be included in the differential diagnosis of radiolucencies occurring in the periapical region of non-vital tooth.
- ▶ It commonly occurs in male in their third decade, with posterior mandible being the most frequent site.
- ▶ Radiological and histopathological features aid in differentiation of OOC from KCOT and former is usually unilocular whereas the later is usually multilocular.
- ▶ OOC is characterised by a less-aggressive behaviour and a low rate of recurrence.

the basal cells of OOCs are not well-developed as those of OKCs. The cells tend to be cuboidal and show little tendency to polarise or palisade. OOCs have a luminal surface of orthokeratin and a well-developed granular layer.²⁰

The type of treatment for OOC depends on several factors such as patient's age, location and size of the lesion. The various surgical modalities includes curettage, enucleation alone, enucleation with Carnoy's solution placed in the cyst cavity before enucleation or in the bone cavity after enucleation, radical enucleation, marsupialisation, and resection.¹⁶

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Oda D, Rivera V, Ghanee N, *et al*. Odontogenic keratocyst: the northwestern USA experience. *J Contemp Dent Pract* 2000;1:60–74.
- 2 Rudagi BM, Kharkar VR, Kini Y. Multiple odontogenic keratocysts with diverse histologic features in a non-syndromic patient. *Pravara Med Rev* 2010;35–7.
- 3 Barnes L, Eveson JW, Reichart P, *et al*. Pathology and genetics of head and neck tumours. Lyon: IARC Press, 2005. WHO classification of tumours series.
- 4 Eryilmaz T, Ozmen S, Findikcioglu K, *et al*. Odontogenic keratocyst: an unusual location and review of the literature. *Ann Plast Surg* 2009;210–12.
- 5 Crowley TE, Kaugars GE, Gunsolley JC. Odontogenic keratocysts: a clinical and histologic comparison of the parakeratin and orthokeratin variants. *J Oral Maxillofac Surg* 1992;22–6.
- 6 Neville BW, D'Amico DD, Allen C, *et al*. *Oral and maxillofacial pathology*. 2nd edn. Philadelphia: Saunders, 2004:597–8.
- 7 Slootweg PJ. Lesions of the jaws. *Histopathology* 2009;54:401–18.
- 8 Wright JM. The odontogenic keratocyst: orthokeratinized variant. *Oral Surg Oral Med Oral Pathol* 1981;609–18.
- 9 MacDonald-Jankowski DS. Orthokeratinized odontogenic cyst: a systematic review. *Dentomaxillofac Radiol* 2010;455–67.
- 10 Philipsen HP. Keratocystic odontogenic tumour. In: Barnes L, Eveson J, Reichart P, Sidransky D, eds. *WHO classification of tumours. Pathology and genetics of tumours of the Head and Neck*. Lyon: International Agency for Research on Cancer (IARC), 2005:306–7.
- 11 Li TJ, Kitano M, Chen XM, *et al*. Orthokeratinized odontogenic cyst: a clinicopathological and immunocytochemical study of 15 cases. *Histopathology* 1998;242–51.
- 12 Dong Q, Pan S, Sun LS, *et al*. Orthokeratinized odontogenic cyst: a clinicopathologic study of 61 cases. *Arch Pathol Lab Med* 2010;271–5.
- 13 Vuhahula E, Nikai H, Ijuhin N, *et al*. Jaw cysts with orthokeratinization: analysis of 12 cases. *J Oral Pathol Med* 1993;35–40.
- 14 MacDonald-Jankowski DS, Li TK. Orthokeratinized odontogenic cyst in Hong Kong community: the clinical and radiological features. *Dentomaxillofac Radiol* 2010;238–43.
- 15 Yoshida H, Onizawa K, Yusa H. Squamous cell carcinoma arising in association with an orthokeratinized odontogenic keratocyst. Report of a case. *J Oral Maxillofac Surg* 1996;647–51.
- 16 Khalam SA, Zacariah RK. Under diagnosis of an odontogenic keratocyst—a controversial lesion IOSR. *J Dent Med Sci* 2012;2:37–40.
- 17 Browne RM. The odontogenic keratocyst. Histologic features and their correlation with clinical behaviour. *Br Dent J* 1971;131:248–59.
- 18 Wright BA, Wysocki GP. Odontogenic keratocyst presenting as periapical disease. *Oral Surg Oral Med Oral Pathol* 1983;56:425–9.
- 19 Onuki M, Saito A, Hosokawa S, *et al*. A case of orthokeratinized odontogenic cyst suspected to be a radicular cyst. *Bull Tokyo Dent Coll* 2009;50:31–5.
- 20 Carvalho CH, Aquino AR, Nonaka CF, *et al*. Infected orthokeratinized odontogenic cyst: a rare cause of facial cellulitis. *Braz Dent J* 2012;23:612–16.

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