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Odontogenic Keratocyst- A Review

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ABSTRACT

An odontogenic keratocyst (OKC) is a benign yet aggressive tumor that develops from the remnants of the tooth germ or dental lamina. Despite its benign nature, OKC has a high recurrence rate and potential to become cancerous. Early-stage OKC often presents without noticeable symptoms, with swelling being the most common clinical manifestation. Therefore, accurate diagnosis is crucial, necessitating a combination of differential diagnosis and auxiliary examination methods. Recent advancements in understanding the molecular basis of OKC have improved its diagnosis and prognosis, enabling targeted therapies and better patient outcomes. This study aims to provide a comprehensive overview of OKC's clinical features, diagnosis, treatment, offering valuable insights for dental practitioners and researchers.

Keywords: Cyst, Syndrome, Keratin, Epithelium.

INTRODUCTION

Cysts are common pathological conditions that affect individuals of all ages and can occur anywhere on the body. Odontogenic cysts, which originate from tissues involved in tooth development, are a significant category within this group. One notable odontogenic cyst is the odontogenic keratocyst (OKC), which comprises approximately 10% of odontogenic cysts. OKC is a locally aggressive cystic lesion that primarily affects the maxilla or mandible, and has the potential for significant bone destruction. The odontogenic keratocyst has long been a subject of debate and evolving classification.[1]

Initially described by Philipsen in 1956, OKC was classified as a cyst. However, in 2005, the World Health Organization (WHO) reclassified OKC as a tumor, renaming it the keratocystic odontogenic tumor (KCOT) to reflect its more aggressive nature and tendency to recur. In 2017, the WHO retracted this classification and returned to the original term, "odontogenic keratocyst," reflecting ongoing research and observations on the disease's behavior and characteristics. Despite this change, OKC remains a critical topic in oral pathology due to its high recurrence rate and its potential for aggressive behavior. It often presents with expansion into adjacent tissues and can cause significant damage to surrounding bone structures.[2]

CLINICAL FEATURES

OKC primarily affects adults between the ages of 30 and 50, representing around 14.3% of all odontogenic tumors. In its early stages, OKC typically shows no clear symptoms, and even large lesions may not cause noticeable jaw enlargement or facial changes. However, some patients may experience swelling, pain, abnormal sensations, discharge of pus, or loose teeth, with swelling being the most common sign. As the cyst grows, the bone around it may expand, causing facial deformity. The affected bone becomes thin and fragile, sometimes giving a "ping-pong" sensation or a parchment-like sound when touched

In severe cases, bone fractures or complete absorption may occur, resulting in a wave-like motion when palpated. OKC can also spread to the nasal cavity or maxillary sinus, potentially affecting vision and causing diplopia. If the cyst is near the teeth, it can cause displacement, loosening, or tilting of the teeth, potentially leading to tooth loss. Caution is needed when extracting loose teeth in OKC patients, as it may rupture the cyst and release sebum-like substances. If the cyst becomes infected, additional symptoms such as pain, fever, and discomfort may occur. Swelling in other areas of the body should prompt a thorough review of the patient's medical history to avoid misdiagnosis. For instance, a 41-year-old woman presented with painless, progressive swelling in her left temporal region. She had previously undergone two OKC surgeries at ages 25 and 38. A physical exam revealed a mass approximately 3.5 cm x 3.0 cm, soft, well-defined, non-tender, and located deeply beneath the temporal muscle.[3]

SYNDROMS ASSOSIATED WITH OKC

Gorlin-Goltz syndrome

MarFan syndrome

Ehler's Danlos syndrome

Noonan's syndrome

DIFFERENTIAL DIAGNOSIS

Ameloblastoma

Primordial cyst

Residual cyst

Traumatic cyst

RADIOGRAPHIC FEATURES

Odontogenic keratocysts (OKCs) are most commonly found in the mandible, particularly posterior to the canines, with over 50% occurring at the angle of the mandible. Radiographically, OKCs appear as undulating borders with a cloudy interior, suggesting multilocularity, and can reach up to 5 cm or more in diameter. The shape of the cyst is typically oval, extending along the body of the mandible with minimal

mediolateral expansion, and often exhibits hyperostotic margins with tooth displacement. While most lesions are unilocular with smooth borders, larger lesions may have irregular borders, and the radiolucency appears hazy due to the keratin-filled cavity, surrounded by a thin sclerotic rim indicating reactive osteocytes.[4]

HISTOPATHOLOGY

Odontogenic keratocysts (OKCs) are distinguished by a unique histopathological profile. Key features include:

Epithelial Lining

- Thin, parakeratinized stratified squamous epithelium

- Typically 6-10 cells thick

- Flat interface between epithelium and connective tissue

Basal Layer

- Well-defined basal layer of columnar or cuboidal cells in palisade

Additional Features

- Presence of keratin in the lumen or satellite cysts

- Inflammation in the connective tissue wall (75% of reported cases)

- Satellite cysts and odontogenic epithelial islands in connective tissue

- Dystrophic calcification

- Epithelial dysplasia (reported in some cases)[5]

DIAGNOSIS OF OKC

OKC (odontogenic keratocyst) can be diagnosed through medical history, clinical symptoms, and diagnostic methods. Puncture is a reliable technique, as yellow or white keratin-like (sebum-like) substances are often found at the site. Keratin staining of the extracted material can confirm the diagnosis of OKC. X-ray imaging is also valuable, revealing a round or oval transparent shadow with well-defined edges, sometimes with irregular borders, and an obvious white bone reaction line around the cyst. B-scan ultrasonography may serve as an auxiliary diagnostic tool. In the case presented, a CT scan showed a low-density mass with a small high-density lesion in the left temporal region above the zygomatic arch, with a clear edge and disappearance of the left mandibular ramus, though the metal joint head remained in place. Ultrasonography also revealed a cystic mass in this area. [6]

A subsequent CT scan showed that the OKC had grown larger and affected the coracoid apex. Given the patient's medical history and specialist examination, recurrence of the OKC in the temporal region was suspected. However, diagnosing OKC from the temporal mass alone was challenging, as conditions like hemangioma and lipoma had to be considered. Hemangiomas are congenital vascular malformations, often positive for the movement test, while lipomas are soft masses that may cause an “orange peel”

appearance when pressed. CT scans of lipomas show lipoid density due to the lipid content. After ruling out other potential tumors based on the patient's history and clinical features, recurrent OKC in the left temporal region was diagnosed. This diagnosis was further confirmed by keratin in the histological staining of the resected tumor. The final diagnosis was confirmed through pathological section analysis, which identified OKC.[7]

TREATMENT OF OKC

Decompression and marsupialization are two frequently employed techniques for treating cysts, particularly odontogenic cysts, to reduce their size and alleviate symptoms. Decompression involves making an opening in the cyst to relieve internal pressure, preventing further expansion. This is typically done by inserting a drain to maintain the opening. Marsupialization, however, involves transforming the cyst into a pouch by creating an opening and suturing the cyst lining to the surrounding mucosa, allowing the cyst to stay open and shrink over time.[8]

Marsupialization is considered a more definitive approach than decompression because it exposes the cyst lining to the oral environment, which may help resolve the lesion. For mandibular cysts, the procedure is usually performed by creating an opening into the oral cavity, while maxillary cysts can be marsupialized into the maxillary sinus, nasal cavity, or oral cavity. This technique was first introduced by Partsch in the late 19th century and is sometimes called the "Partsch I procedure."[9]

The effectiveness of decompression and marsupialization for treating keratocystic odontogenic tumors (KCOTs) is a subject of debate. Some researchers argue that these methods leave the cyst lining in place, which could lead to recurrence. However, other studies suggest that decompression and marsupialization reduce the cyst size and preserve important structures, such as teeth and nerves, ultimately allowing for enucleation. Some studies even report that marsupialization can lead to the complete resolution of KCOTs without requiring additional surgery.[10]

During marsupialization, an opening of at least 1 cm is created, and the cyst lining is sutured to the surrounding mucosa. For maxillary cysts, the cavity is often packed with iodoform gauze and bacitracin ointment to prevent closure. In the mandible, there is a higher likelihood of spontaneous closure of the fistula, but a nasopharyngeal tube may be used as a stent to keep the cyst open, requiring regular irrigation to prevent food buildup and closure of the fistula. Histologically, marsupialization has been shown to thicken the cyst lining, which may increase the likelihood of successful enucleation. The cyst lining eventually resembles normal oral mucosa. While some studies report recurrence, others indicate that decompression and/or marsupialization can be just as effective as more invasive treatments, with lower morbidity and better preservation of vital structures. [10]

CONCULISION

Due to their high recurrence rates and potential for malignant transformation, odontogenic keratocysts (OKCs) require careful consideration in terms of diagnosis, treatment, and management. To minimize the risk of recurrence and malignant transformation, it is essential to thoroughly evaluate preoperative imaging data, ensuring precise operative planning and complete tumor removal. [1]

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