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CASE REPORT

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Giant Keratocystic Odontogenic Tumor of the Mandible – A Case Report

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Authors' Contribution:
A Study Design
B Data Collection
C Statistical Analysis
D Data Interpretation
E Manuscript Preparation
F Literature Search
G Funds Collection

Summary

Background: The keratocystic odontogenic tumor (KCOT) is a relatively rare, benign neoplasm which develops in the maxilla or mandible, arising from the dental lamina or basal cells of the oral epithelium. It is often found incidentally and brings about late symptoms as it does not cause bone distension for a long time.

Case Report: The presented case is of a young woman with a giant keratocystic odontogenic tumor of the mandible.

Conclusions: Despite its rare occurrence, it must be taken into consideration in radiological and clinical diagnostics. Due to the frequent recurrence of KCOT, patients are recommended to be kept under long-term and close radiological supervision.

MeSH Keywords: Mandible • Odontogenic Cysts • Odontogenic Tumors

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Background

The keratocystic odontogenic tumor is a relatively rare, benign neoplasm which develops in the jaw or mandible, arising from the dental lamina or basal cells of the oral epithelium. It was first described by Phillipsen in 1956 and was initially named the odontogenic keratocyst (OKC). In 2005, the World Health Organization (WHO) re-classified OKC and labelled it keratocystic odontogenic tumor (KCOT). The following features of the tumor contributed to that decision: i) clinically: local malignancy with high recurrence rate (up to 60% [1–4]), ii) histopathologically: pullulation of the stratum basale of the epithelium into the connective tissue and absence of mitotic figures in the stratum suprabasale, iii) genetically: occurrence of mutations of the suppressor gene PATCHED 1 [1–13]. The tumor develops more often in the mandible than maxilla, mainly in the posterior part of its body and ramus area. It occurs less frequently as an extraosseous manifestation in the gingival area, and rare cases in the malar area have also been documented [1,3–6]. KCOT may be related to the unerupted third molar [6,13]. Keratocystic odontogenic tumors constitute between 2% and 11% of all mandibular neoplasms [2,4]. They are more frequent in males and have their peak incidence in the third decade of life [1,2,13]. They may develop sporadically or accompany Gorlin syndrome (nevoid basal cell carcinoma syndrome, NBCCS) [7].

KCOTs are often found incidentally in X-ray images, they are painless and cause tooth dislocation more often than resorption, tend to grow in the mesial-distal direction (where "mesial" means directed towards the anterior midline in a dental arch) rather than vestibular-lingual, and in the mandibular ramus areas, which delays the symptoms of bone distension [13].

Case Report

In March 2013, a 31-year-old female patient palpated a hard nodule in the area of the angle of her left mandible. She was pregnant at the time and decided that it was caused by changes taking place in her organism. In early December, after giving birth to her child, she went to her family doctor because the nodule grew in size instead of diminishing. The GP referred her to the emergency department of the 4th Military Hospital in Wrocław, where she was X-rayed in both posteroanterior and lateral views. The description was: "Features of reconstruction of the

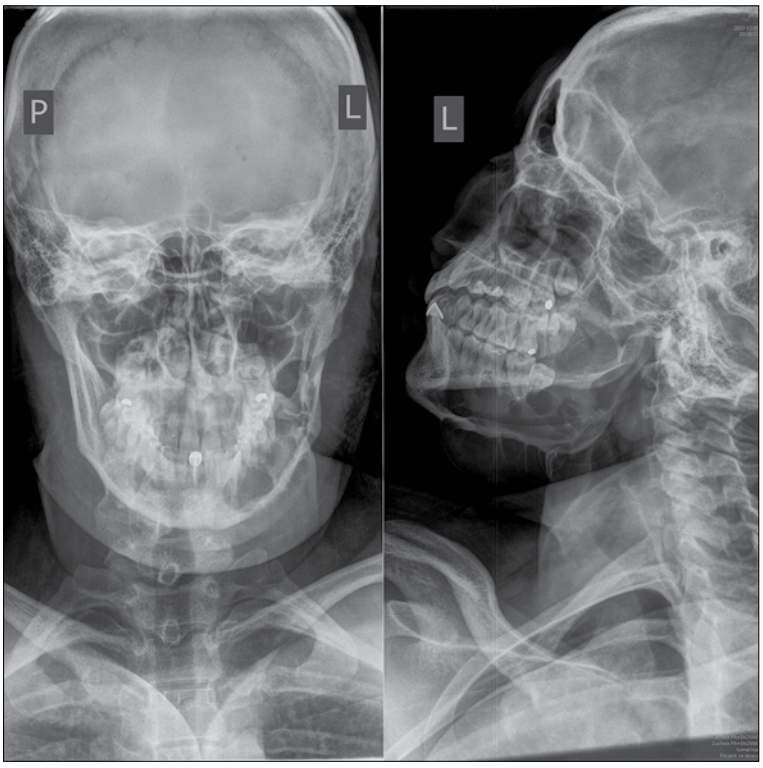

Figure 1. AP and lateral X-ray of the mandible. Features of reconstruction of bone structure and ramus area on the left side with considerable distension, presence of numerous cystic centres of rarefaction of bone structure and septum.

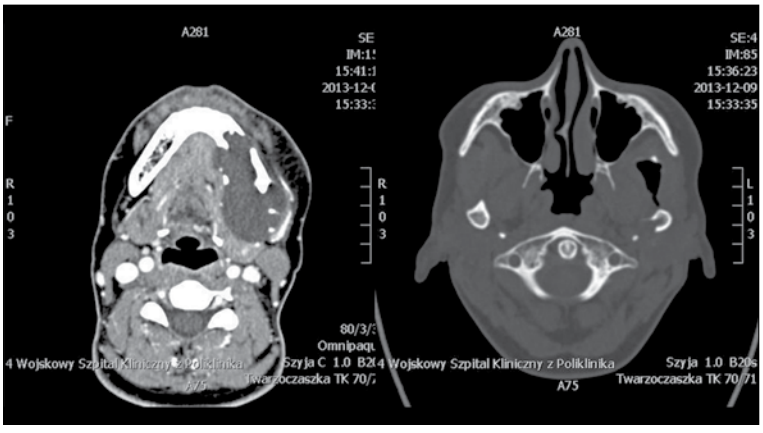

Figure 2. CT of the facial cranium with contrast medium. Extensive osteolytic decrease caused by the presence of a hypodense, polycyclic lesion of approximately 10×10×4 centimetres not undergoing a substantial enhancement on administering intravenous contrast medium.

bone structure in the mandibular body and ramus region on the left side with considerable distension, presence of numerous cystic centres of rarefaction of bone structure and septum" (Figure 1). The diagnostic process was extended to include computer tomography of the facial cranium. The examination revealed locally an extensive osteolytic decrease caused by the presence of a hypodense, polycyclic lesion of approximately 10×10×4 centimetres, which did not undergo substantial enhancement after administration of intravenous contrast medium. The tumor altered topographic relations of the left-side submandibular and neck areas, and its environs featured a developed network of blood vessels. Within the neck region no enlarged lymph glands were revealed (Figure 2).

The patient was referred to the Oral and Maxillofacial Surgery Clinic, where orthopantomography and decompression of the lesion were performed, with the resulting material examined histopathologically (Figure 3). The results of the histopathological examination revealed small fragments of connective tissue covered in epithelium paraepidermoidale with chronic inflammatory lymphoid infiltration in the stroma, without malignant lesions. The patient was scheduled for surgery and before that operation she underwent several decompression procedures. The results of the specimen cultures were always negative.

On admittance to hospital for tumor surgery, a clinical examination revealed extraorally a facial asymmetry and a painless, hard, immobile bulge covering the area of the body of the mandible and left-side submandibular area, with the skin unchanged. Intraorally, a distension of the body of the mandible was observed, extending from the mandibular ramus to the incisor teeth, as well as an active retromolar fistula discharging pus. The neighbouring mucosa was tender, and mild pain accompanied chewing. Pus sample cultures grew *Escherichia coli* and *Streptococcus oralis*.

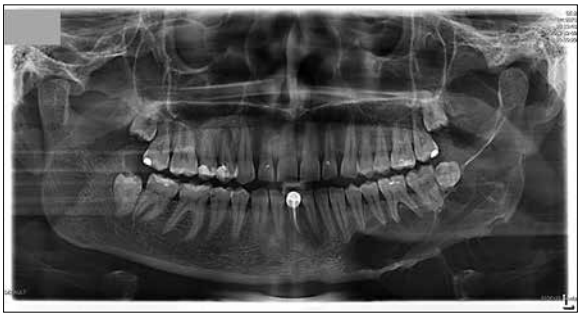

Figure 3. Orthopantomogram of the mandible. Features of reconstruction of bone structure and ramus area on the left side with considerable distension, presence of numerous cystic centres of rarefaction of bone structure and septum.

The patient underwent hemisection of the mandible with exarticulation of the mandibular condyle from the left temporomandibular joint, reconstruction of the left side of the mandible and left temporomandibular joint by means of autogenic bone grafting from the left ala of the ilium and a titanium implant (Figure 4).

The left neck lymphatic glands were examined histopathologically, as well as the lower left alveolar nerve, half of the mandible with the mandibular condyle and tumor, part of the joint capsule of the temporomandibular joint, and the bone of the stump of the mandible. The diagnosis was keratocystic odontogenic tumor of the mandible. The postsurgical process went without complications.

Discussion

The term "giant" in keratocystic odontogenic tumors is reserved for large-sized lesions which result in significant asymmetry and local dysfunction, therefore the case described here may be classified as giant – the dimensions of the tumor were 10×4×4 centimetres. The lesion is more aggressive than other odontogenic cysts. In the histopathological image it is e.g. lined with keratinized epithelium [2,9].

Immunohistochemical analyses revealed that the neoplasm had an increased expression of proteins responsible for cell proliferation, i.e. Ki67, COX-2 (cyclooxygenase 2), PCNA (proliferating cell nuclear antigen), p53, and p56 (epithelial stem cell enhancer factor), BCL2 (anti-apoptotic factor). Conversely, the expression of the BAC pro-apoptotic marker was decreased. The above-mentioned proteins may be used as biological markers for future KCOT diagnostics [6,10,11,14]. Other studies reported on an increased production of glycoprotein podoplanin by KCOT. Zhang et al. proved that after decompression (i.e. lowering the tumor-induced pressure) patients had a statistically significantly decreased level of podoplanin [8].

KCOT is a rare tumor which may also accompany Gorlin's syndrome (nevoid basal cell carcinoma syndrome, NBCCS). The gene responsible for the development of NBCCS is PTCH1, which codes the glycoprotein in cell membranes – TM2. In their research, Guo et al. described a close connection of TM2 mutation with the incidence of sporadic KCOT [7].

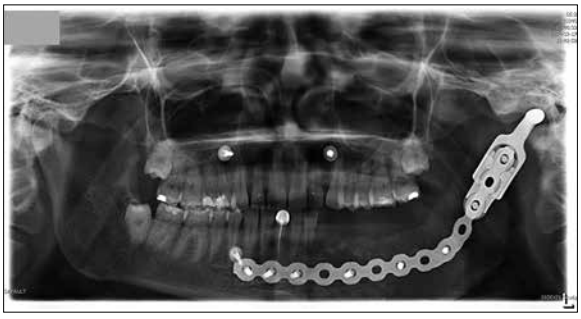

Figure 4. Condition after hemiresection of the mandible with exarticulation of the mandibular condyle from the left temporomandibular joint, reconstruction of the left side of the mandible and left temporomandibular joint by means of autogenic bone grafting from the left ala of the ilium and a titanium implant.

Some atypical manifestations of the keratocystic tumor are also worth mentioning here. It may occur in an extraosseous form as a peripheral keratocystic odontogenic tumor (PKCOT) – in gums, cheek tissue and the lateral facial deep region (LFDR) [3]. Several cases of the variety known as pigmented KCOT have been documented. It owes its name to the presence of melanocyte dendrite formations without atypical features within the squamous epithelium of the tumor. This kind of KCOT mainly appears in young females (average age 18) [9]. Shetty et al. described a case of KCOT with mural calcification [4], whereas Patil et al. – the occurrence of "spider-like bodies" in KCOT's inflammatory cells [5].

The tumor tends to be found incidentally because it shows no early symptoms and is painless. Its characteristic feature is growth in the mesial-distal rather than vestibular-lingual direction, and upwards in the mandibular ramus areas, which delays the symptoms of bone distension. Tooth resorption is rare, tooth dislocation is more common. Large tumors invade soft tissue after damaging the compact external bone lamella [13]. Keratocystic tumors are seldom large-sized – only one study in the quoted literature described three cases of giant (i.e. 7–10 centimetres) mandibular tumors [2]. The dimensions of our patient's lesion were 10×4×4 centimetres.

In radiological examinations, a KCOT may be manifested in various ways. Initially, it is shown as an oval or round centre characterized by a unilocular radiolucency. The radiolucency may be polycyclic, less often multi-locular, with an osteosclerotic rim. A third molar may be found within the tumor area. CT images show hypodense lesions reflecting keratin mass in the tumor. MR examinations reveal a heterogeneous signal – medium in T1-weighted images, high in T2-weighted images, contrast enhancement revealing only a thin perimeter of the lesion. CT and/or MR examinations may reveal infiltration of the maxillary sinus, orbit, pterygopalatine fossa and infratemporal fossa.

KCOT's differential diagnosis includes: follicular cyst when an unerupted third molar is impacted in the tumor area; ameloblastoma, odontogenic myxoma and *pericoronitis* of the impacted third molar [13].

Methods of treatment of KCOT include: decompression, marginal resection, *en bloc* resection and adjuvant therapy, i.e. cryotherapy, peripheral ostectomy and administering Carnoy's solution after surgery [1,2]. Adjuvant therapies are used in order to reduce the relapse rate. KCOT's recurrence ratio is high, up to 60% [1–4]. In their study, Campos, Telles et al. proved that marsupialisation before the final enucleation of the tumor causes its fibrous capsule to thicken and modifies its epithelial lining, thus greatly facilitating the enucleation [1]. The size of our patient's tumor as well as the lack of unambiguous histopathological diagnosis before surgery influenced the choice of therapy – hemimandibulectomy with simultaneous reconstruction by means of an autologous graft from the ala of the ilium and a titanium implant were performed. The patient will now undergo regular control examinations, including radiological examination, in the Oral and Maxillofacial Surgery Clinic.

Conclusions

Keratocystic odontogenic tumors (KCOTs) are rare tumors which most frequently develop in the mandible. They are often found incidentally. Untreated, they may lead to dysfunctions of the organ and significant asymmetry of the face. Despite their rare occurrence, they must be taken into consideration in radiological and clinical diagnostics. Due to scarcity of relevant Polish publications, we decided to present the issue. Due to the frequent recurrence of KCOT, also after radical surgery, patients are recommended to be kept under long-term and close radiological supervision.

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