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Case Report

Pediatric case of squamous cell carcinoma arising from a keratocystic odontogenic tumor

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ABSTRACT

Keratocystic odontogenic tumors (KCOT) are exceptional in children and adolescents as they usually occur in the third decade. The present study reports the case of a 15 years old girl who was diagnosed with a KCOT that underwent malignant transformation. KCOT diagnostic was based on clinical, radiological, histopathological and immunohistochemical findings. A conservative treatment by enucleation was performed. Histopathological analysis of the surgical specimen concluded to a KCOT, with an infra-centimetric focus of well-differentiated squamous cell carcinoma. Owing to the well-differentiated character of the squamous cell carcinoma, a single clinical and MRI surveillance every 3 months was decided, without complementary treatment.

1. Introduction

Keratocystic odontogenic tumors (KCOTs) are exceptional in children and adolescents as they usually occur in the third decade of life [1]. It is defined as « a benign uni- or multicystic intraosseous tumor of odontogenic origin, with a potentially local aggressive and infiltrative behavior » [2]. Owing to its destructive nature and high recurrence rate, it was reclassified by the World Health Organization (WHO) in 2005 as a benign neoplasm of odontogenic origin, and not as a cyst [3].

KCOTs represent 14.5% of odontogenic lesions in the pediatric population [4], with a male preponderance, and a mean age of occurrence of 14.7 years old [5] in pediatric series. They can be sporadic, or syndromic, associated with nevoid basal cell carcinoma syndrome.

Various authors described cases of squamous cell carcinoma arising from a KCOT in adults [6–10], but the exact incidence of KCOT malignant transformation is not known, and believed to be rare [2].

The present study reports the case of a 15-year-old girl who was diagnosed with a KCOT that underwent malignant transformation, forming a well-differentiated squamous cell carcinoma of the mandible.

2. Case presentation

A 15-year-old female with no medical history was referred to the Head and Neck Surgery Department of the Léon Bérard Center in Lyon (France), for a cystic lesion of the right mandible developed over an included supernumerary tooth. The lesion was described by her parents as rapidly increasing in size over a period of two months, and was associated with pain and intermittent bleeding.

Clinical examination revealed a right irregular mandibular mass extending from teeth numbers 42 to 47 that was tender to the touch. Dental occlusion seemed to be modified. There was no lower lip numbness, and no significantly enlarged submandibular or cervical lymph nodes on palpation. A dental CT scan showed a cystic lesion with root resorption.

Although dental CT scan findings were in favor of a benign lesion, as a malignant tumor was suspected, owing to the assumed rapid growth of the tumor, clinical staging was completed through a **CT scan**, an **MRI**, and a **PET-CT scan**. The CT scan revealed a tissular mass 36 × 20 × 28mm in size that developed over an included supernumerary tooth, lysing the lingual cortex of the right posterior mandible, with heterogeneous enhancement after contrast material injection (Fig. 1A and B). The lesion was hyperintense on T2-weighted MRI, hypointense on T1-weighted MRI, and infiltrated the medulla of the right mandible over 4 cm (Fig. 2). It was intensely hypermetabolic on the PET-CT scan, without locoregional lymph node or distant metastatic fixation (Fig. 3).

Two **surgical biopsies** were carried out. **Microscopic examination** of the tissue revealed a cystic lesion lined by hyperplastic pseudo-

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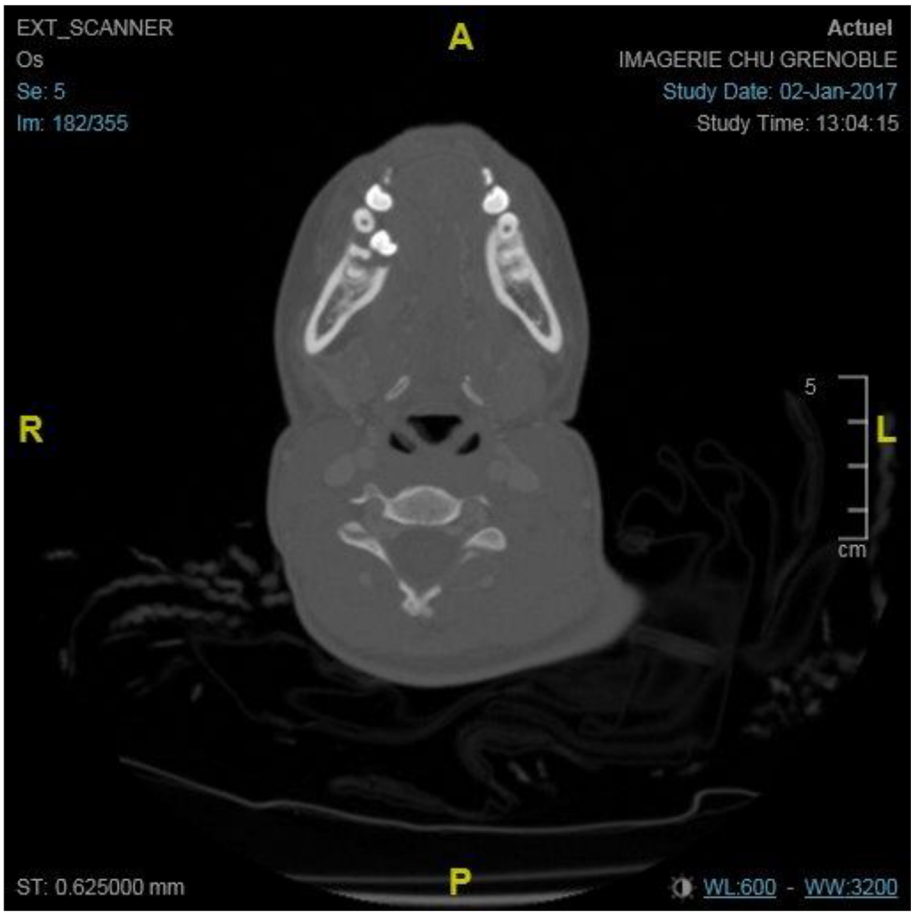

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Fig. 1. A) Included supernumerary tooth on a dental CT scan. **B)** Lysis of the lingual cortex of the right posterior mandible on a transverse CT scan.

epitheliomatous stratified squamous epithelium in a few areas, features suggestive of a form of dysplasia such as nuclear hyperchromatism and pleomorphism. **Immunohistochemical analysis** indicated the presence of Ki-67-positive cells in the basal and suprabasal cell layers of the tissue samples. 60% of epithelial cells presented a nuclear and cytoplasmic expression of p16, without significant p53 expression. In situ DNA hybridization for HPV oncogene detection was negative.

A diagnosis of **KCOT** was made. An **enucleation** was decided, along with the avulsion of the supernumerary tooth, and teeth 44, 45 and 46 (Fig. 4A and B). Histopathological analysis of the right mandible lesion indicated a KCOT, with an **infra-centimetric focus on well-differentiated squamous cell carcinoma in the middle of the cyst** (Fig. 5A–C).

Testing for the Gorlin-Goltz syndrome gave a negative result based on familial/clinical history and a blood sample. MRI was done one month post-operation, showing no sign of recurrence. Owing to the well-differentiated character of the squamous cell carcinoma, and its central character with large margins of cancer-free tissue, a single clinical and MRI surveillance every 3 months was decided, without complementary treatment. To date, no local or cervical recurrence of the squamous cell carcinoma was observed, with a retrospective time perspective set at 18 months after the surgery.

3. Discussion

The presumed rapid growth of the tumor led us to suspect a more