# Odontogenic Tumors

# 20

Jan Godzinski, Roberto Bianchi, and Marco Guzzo

## Contents



Odontogenic tumors (OT) derive from epithelial, ectomesenchymal, and or mesenchymal elements that still are or have been part of the tooth-forming apparatus. Therefore, they are found exclusively within the maxillofacial skeleton or in the soft tissue overlying tooth-bearing areas. These tumors are classified according to WHO histological classification published in 2005 (Barnes et al. 2005). There are few reports on OT in children (Adebayo et al. 2002; Asamoa et al. 1990; Guerrisi et al. 2007; Ulmansky et al. 1999). The literature shows these neoplasms accounted for between 1.0 and 28.8% of the oral lesion (Ulmansky et al. 1999).

Comparative studies of OT in children are difficult because various authors use differing classifications; age groups and patients are of different racial origins. Basically, all the defined histotypes can occur in children although most of them and particularly the malignant counterpart are rare in such patients.

The most common OT in children are reported in Table 20.1 and Table 20.2. There appears to be a racial predilection for OT types.

## 20.1 Ameloblastoma

Ameloblastoma, or adamantinoma as this tumor was named before 1930, is a rare benign tumor originating from odontogenic epithelium with mature, fibrous stroma without odontogenic ectomesenchyma. It is one of the most frequent OT in all age group, but the peak incidence is in the third–forth decades. About 10–15% of them occurred in children. According to WHO classification, four types have been described as follows: a solid/multicystic (about 70% of all), an

J. Godzinski (✉)
Department of Pediatric Surgery, Marciniak Hospital and Chair of Emergency Medicine, Wroclaw Medical University, Wroclaw, Poland
e-mail: jgodzin@dilnet.wroc.pl

R. Bianchi • M. Guzzo
Otolaryngology Unit, Istituto Nazionale Tumori, Milan, Italy
e-mail: roberto.bianchi@istitutotumori.mi.it; marco.guzzo@istitutotumori.mi.it

D.T. Schneider et al. (eds.), *Rare Tumors In Children and Adolescents*, Pediatric Oncology,
DOI 10.1007/978-3-642-04197-6_20, 

**Table 20.1** Odontogenic tumors in children and adolescent. Review of the literature

| Authors | No. of cases | Geographic area | Histotype | % |
|---|---|---|---|---|
| Adebajo et al. (2002) | 78 | Nigeria | Ameloblastoma | 54 |
| | | | Myxoma | 19 |
| | | | Adenomatoid odontogenic tumor | 9 |
| | | | Ameloblastic fibroma | 8 |
| | | | Odontoma | 9 |
| | | | Others | |
| Guerrisi et al. (2007) | 153 | Argentina | Odontoma | 50 |
| | | | Ameloblastoma | 18.3 |
| | | | Myxoma | 8.5 |
| | | | Adenomatoid odontogenic tumor | 5.2 |
| | | | Others | |
| Jones and Franklin (2006) | 243 | United Kingdom | Odontoma | 79.4 |
| | | | Adenomatoid odontogenic tumor | 4 |
| | | | Ameloblastoma | 3.7 |
| | | | Ameloblastic fibroma | 3.3 |
| | | | Others | |
| Ulmansky et al. (1999) | 18 | Israel | Myxoma | 38.8 |
| | | | Ameloblastic fibroma | 22.2 |
| | | | Ameloblastic fibro-odontoma | 22.2 |
| | | | Ameloblastoma | 11 |
| | | | Others | |
| Sato et al. (1997) | 79 | Japan | Odontoma | 59.5 |
| | | | Ameloblastoma | 34 |
| | | | Others | |

**Table 20.2** Diagnostic workflow of odontogenic tumors

| | |
|---|---|
| Physical examination | In most benign tumors, signs and symptoms are scarce. Sometimes slow swelling of jaw and/or movement of teeth can be noted. Less frequently can occur as painless exophytic-like lesion of the alveolar mucosa. In contrast, pain is the most frequent symptoms followed by rapid swelling of the bone in malignant odontogenic tumors |
| Laboratory assessment | None |
| Radiological assessment | |
| – First assessment | More often discovered incidentally during the course of routine intraoral dental (plain) radiographs or panoramic radiography |
| – Preoperative assessment | Panoramic radiography, CT |
| – FU | Panoramic radiography |
| Pathological assessment | Most odontogenic tumors are benign, and the diagnosis can be only achieved by clinical and radiological evaluations. Histological confirmation is often necessary and should be always obtained before planning a nonconservative surgery |
| Staging system (odontogenic carcinomas) | Usually, it is not applied |
| Grading system (odontogenic carcinomas) | Malignant odontogenic tumors are exceedingly rare in children and are usually highly aggressive. Grading is still definited by histotype |
| General treatment guidelines | Need for multidisciplinary approach<br>Need for referral to center with expert physicians professionally dedicated to the management of this neoplasms in adults (or strict collaboration with them) |
| – Surgery | Keystone of treatment. Complete segmental bone resection, enucleation or curettage according to histotypes. Immediate bone graft is recommended in order to prevent facial deformity |
| – Radiotherapy | High-grade tumors are very rare |
| – Chemotherapy | Palliative intent |

extraosseous/peripheral (1.3–10% of all), a desmoplastic (rare), and a unicystic (5–15% of all) type (Barnes et al. 2005). Some studies reported that the unicystic type is the most common type in children (Ord et al. 2002), but the relevant literature shows the rate of solid type is higher than unicystic type (Zhang et al. 2010). Solid and unicystic types locate usually in the mandible (more than 90% of the cases). While the mandible-to-maxilla ratio is 1:1 and 2.4:1 for desmoplastic and extraosseous type, respectively. In the mandible, the tumor has a marked predilection for the posterior region except in African Blacks in whom the tumor occurs in the symphysis (Chidzonga 1996). Localizations in other bones are described but very rarely (Kessler and Dominiquez 1986). Solid/multicystic, unicystic, and desmoplastic ameloblastoma grow slowly but may markedly deform involved portions of the bone. Diagnosis based on clinical appearance may reveal swelling of the corresponding area of face and mandible or maxillary intraosseous mass. Pain or paresthesia is rare. Solid/multicystic types may be diagnosed as unilocular or multilocular (soap bubble-like) radiolucencies at panograph X-ray or CT scan. An unerupted tooth may be associated. The roots of the involved tooth may be eroded. Despite characteristic imaging, microscopic confirmation of diagnosis is recommended. Unicystic type radiographically present as a unilocular, often pericoronal radiolucency, that may also be associated with an unerupted tooth. Desmoplastic ameloblastoma shows a pronounced stromal component compressing the odontogenic epithelial component. Consequently, about 50% of them are radiographically present as a mottled, mixed radiolucency/radiopacity with ill-defined borders. Extraosseous ameloblastoma usually is located to the alveolar mucosa in edentulous area. This tumor grows as an exophytic-like lesion determining a superficial erosion of the bone crest due to pressure resorption (Stevenson and Austin 1990) (Figs. 20.1–20.7).

Once the diagnosis is established, the treatment may be planned. Surgery is the treatment of choice. In children, the decision for treatment strategy should be made according to the patient's age, tumor size, location, and whether it is a primary or a recurring tumor. Resection usually creates fewer difficulties than reconstruction of the mandible. Although ameloblastoma is not a malignant disease, surgeons must keep in mind that complete excision of this locally aggressive tumor is the mainstay of treatment. Enucleation or curettage leads to risk of

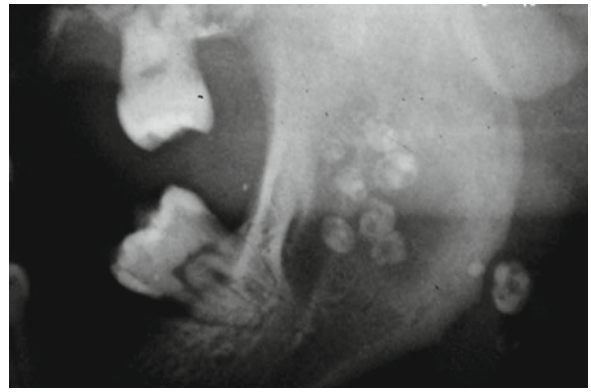

**Fig. 20.1** Odontoma compound type in an 18-year-old girl

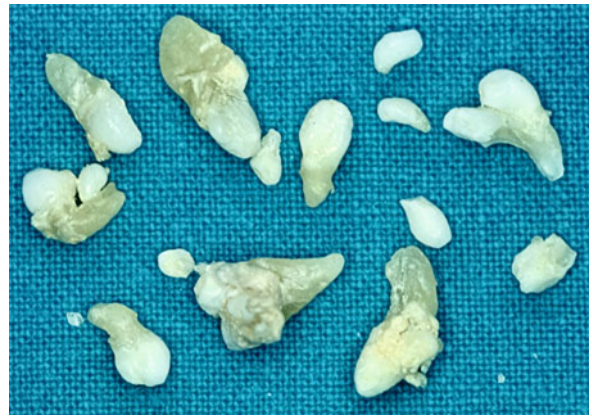

**Fig. 20.2** Macroscopic specimen of odontoma compound type in an 18-year-old girl

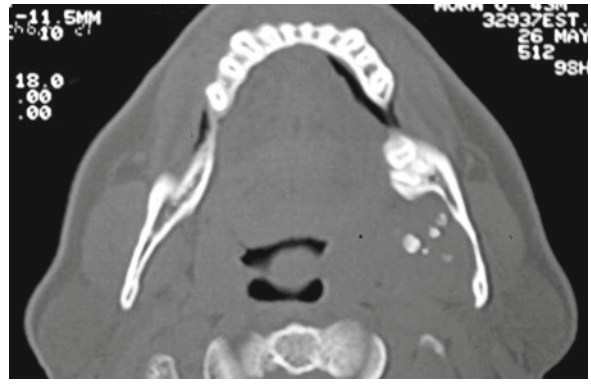


**Fig. 20.3** Computed tomography of odontoma compound type in an 18-year-old girl

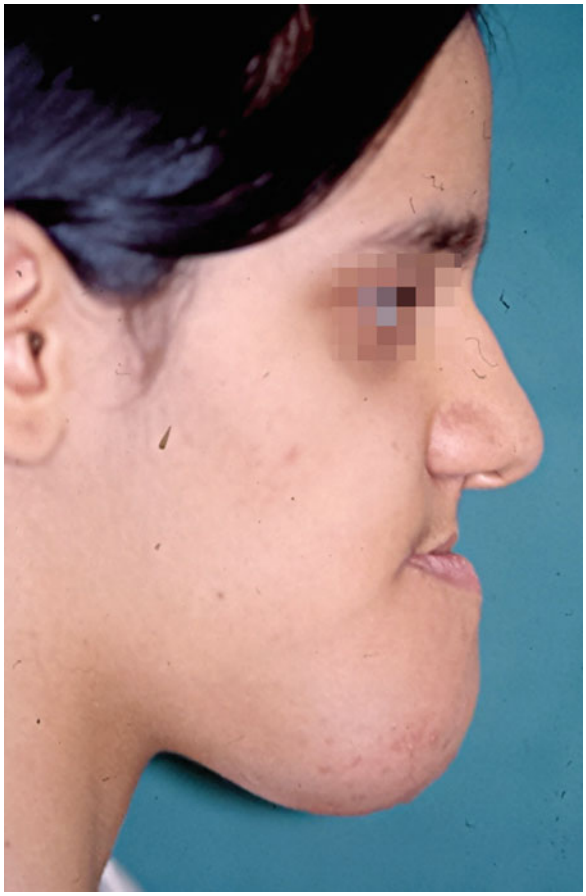

**Fig. 20.4** Cementoblastoma in a 17-year-old girl

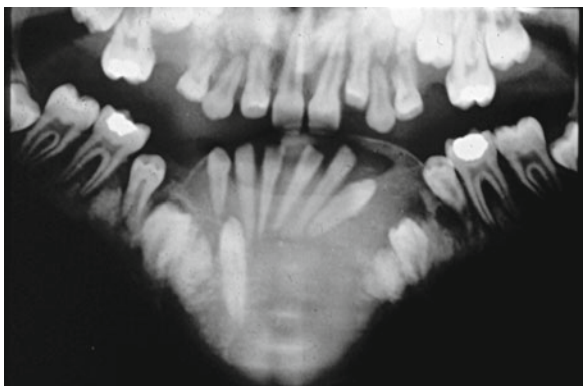

**Fig. 20.5** Ortho-panoramic X-ray of cementoblastoma in a 17-year-old girl

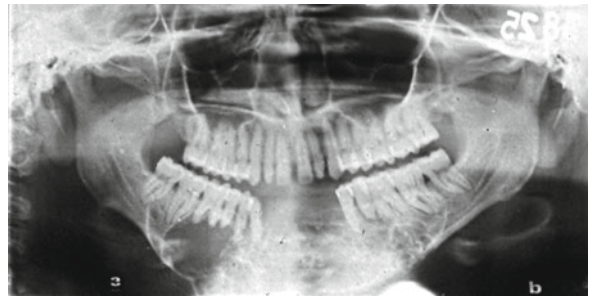

**Fig. 20.6** Ortho-panoramic X-ray of ameloblastoma of the mandible extending from 4.7 to 3.7

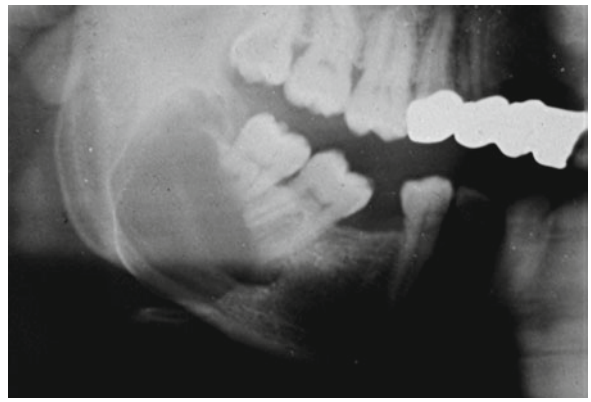

**Fig. 20.7** Ortho-panoramic X-ray of ameloblastoma of the right mandible. Unilocular radiolucency resembling cysts with scalloped borders

recurrence as high as 25–40%. The block resection of the tumor with subsequent reconstruction with bone graft is routinely recommended. In more advanced cases, the segmental resection of mandible followed by complete reconstruction may be necessary. Biopsy for histological confirmation is often necessary and should be always obtained before planning a nonconservative surgery (Zhang et al. 2010). Unicystic ameloblastoma is thought to recur less frequently than other subtypes, thus some authors advocate also less aggressive treatment in this form of tumor and reserve more aggressive surgery for any recurrence. However, a recent large review revealed that this histotype is aggressive and should be treated as solid ameloblastoma (Philipsen and Reichart 1998).

The tumor usually recurs in between 1 and 15 years of follow-up with a peak within 5 years from surgery (Ord et al. 2002). Consequently, a long-term postoperative follow-up seems reasonable.

The complementary treatments, as chemotherapy or radiotherapy, are of limited value. In spite of a benign histologic aspect, some ameloblastoma may spread distant metastases, mainly to the lungs (metastasizing ameloblastoma). Moreover, an ameloblastic

carcinoma may arise from a benign ameloblasoma either intraosseous or peripheral (Barnes et al. 2005).

## References


Adebayo ET, Ajike SO, Adekeye EO (2002) Odontogenic tumours in children and adolescents: a study of 78 Nigerian cases. J Craniomaxillofac Surg 30(5):267–272

Asamoa EA, Ayanlere AO, Olaitan AA, Adekeye EO (1990) Paediatric tumours of the jaws in northern Nigeria. Clinical presentation and treatment. J Craniomaxillofac Surg 18(3):130–135

Barnes L, Eveson JW, Reichart P, Sidransky D (2005) Pathology and genetics of head and neck tumours. IARC Press, Lyon

Chidzonga MM (1996) Ameloblastoma in children. The Zimbabwean experience. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 81(2):168–170

Guerrisi M, Piloni MJ, Keszler A (2007) Odontogenic tumors in children and adolescents. A 15-year retrospective study in Argentina. Med Oral Patol Oral Cir Bucal 12(3):E180–E185

Jones AV, Franklin CD (2006) An analysis of oral and maxillofacial pathology found in children over a 30-year period. Int J Paediatr Dent 16(1):19–30

Kessler A, Dominiquez FZ (1986) Ameloblastoma in childhood. J Oral Maxillofac Surg 44:609–611, Ref Type: Generic

Ord RA, Blanchaert RH Jr, Nikitakis NG, Sauk JJ (2002) Ameloblastoma in children. J Oral Maxillofac Surg 60(7):762–770

Philipsen HP, Reichart PA (1998) Unicystic ameloblastoma. A review of 193 cases from the literature. Oral Oncol 34(5):317–325

Sato M, Tanaka N, Sato T, Amagasa T (1997) Oral and maxillofacial tumours in children: a review. Br J Oral Maxillofac Surg 35(2):92–95

Stevenson AR, Austin BW (1990) A case of ameloblastoma presenting as an exophytic gingival lesion. J Periodontol 61(6):378–381

Ulmansky M, Lustmann J, Balkin N (1999) Tumors and tumor-like lesions of the oral cavity and related structures in Israeli children. Int J Oral Maxillofac Surg 28(4):291–294

Zhang J, Gu Z, Jiang L, Zhao J, Tian M, Zhou J et al (2010) Ameloblastoma in children and adolescents. Br J Oral Maxillofac Surg 48(7):549–554