Management of Unicystic Plexiform Ameloblastoma in a 12 year Old Female: Report of a Case

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Abstract

Ameloblastoma is a true neoplasm of odontogenic origin and benign in nature. But it has a high percentage of local recurrence with possible malignant development when treated inadequately. The treatment planning in ameloblastomas is still not clear. We describe a case report of unicystic plexiform ameloblastoma in a young girl who was treated with conservative surgical management (enucleation & chemical cauterization) in our unit. The patient remains well without disease after 5 years of postoperative follow-up and is still being followed up.

Introduction

Ameloblastoma is a benign epithelial odontogenic tumor but is often aggressive and destructive, with the capacity to attain great size, erode bone and invade adjacent structures [1]. The World Health Organization (1991) defined ameloblastoma as a benign but locally aggressive tumour with a high tendency to recur, consisting of proliferating odontogenic epithelium lying in a fibrous stroma [2].

The term "ameloblastoma" was suggested by Churchill (1934), because the old term ("adamantinoma", coined by Malassez in 1885) erroneously implied the formation of hard tissue [3]. It represents about 1% of all oral ectodermal tumors and 9% of odontogenic tumors. 80% of ameloblastomas occur in mandible and 20% of ameloblastomas occur in the maxilla. In maxilla they are localized most often in the canine and antral regions, where as in mandible 70% are located in the area of the molars or the ascending ramus, 20% in the premolar region, and 10% in the anterior region. Men are affected slightly more often than women. Ameloblastoma appears most commonly in the third to fifth decades but the lesion can be found in any age group including children [4].

There are 6 histologic subtypes of ameloblastoma: follicular, plexiform, acanthomatous, granular, basal cell and desmoplastic. They can be found combined or isolated and that are not related to prognosis of the tumor. There are also three different macroscopic subtypes: solid or multicystic, un cystic and peripheral. This classification may have a prognostic value [6].

Case Report

A 12-year-old girl was referred to the department of oral and maxillofacial surgery, with a diffuse swelling in the right side of mandible [Figure 1]. The swelling was painful and had been slowly increasing in size for 6 months. Extra oral examination revealed the swelling measuring about 4 cm x 4 cm in size on the right mandible extending antero-posteriorly to about 4 cm from the symphysis region to 1 cm in front of the ear lobule, and supero-inferiorly it was 2 cm from the cheek prominence to 1 cm below the lower border of the mandible. The swelling was firm in consistency, tender on palpation with a mild rise in temperature.

Intraorally, the swelling extended from the mesial surface of right first molar to the retromolar region, obliterating the buccal vestibule [Figure 2]. The orthopantomograph revealed a well-defined unilocular radiolucency extending from first molar region towards the ramus of mandible involving the coronoid process on the involved side [Figure 3]. It was found that second molar was completely impacted with radiolucency around its crown portion and the roots of first molar were displaced. Paranasal sinus radiograph revealed a well-defined radiolucency involving the ramus of mandible with its bucco-lingual extensions [Figure 4].

Fine needle aspiration cytology was performed which did not give any conclusive evidence. Therefore, an extensive enucleation of the said lesion was planned under general anesthesia. A Modified Wards incision was placed extending anteriorly up to the region of canine. After reflecting the mucoperiosteal flap, the expanded cortical plate was identified and separated from the mucoperiosteum. Subperiosteal dissection was carried out beginning from the sound bone near the canine region to posteriorly over the ramus of mandible [Figure 5]. The mental nerve was identified and preserved. The cystic lining was separated from the inferior border of the mandible taking care not to injure the inferior alveolar nerve. The impacted tooth bud was delivered out with its cystic lining. The retracted cystic mass was then sent for histopathological analysis. The histopathological processing of the tumor revealed a plexiform ameloblastoma, predominantly composed of...
epithelium arranged as a tangled network of anastomosing strands enclosing cysts of various size [Figure 6].

Discussions and Conclusion

Ameloblastoma is a true neoplasm of odontogenic epithelium. It is uncommon in children, in a review of 1,036 ameloblastomas of jaw, the average patient age is 38.9 years, with only 2.2% (19 of 858) were under 10 years and 8.7% (75 of 858) were between 10 and 19 years [6]. The unicystic ameloblastoma usually presents between 16 and 20 years of age, and the multicystic ameloblastoma after 30 years of age [7].

Typical ameloblastoma starts insidiously as a central bony lesion which is slowly destructive, however it tends to expand the bone instead of punching a hole through it. The tumor is rarely painful, unless infected and usually does not cause signs and symptoms of nerve involvement, even when large. Ackerman et al, in their study of unicystic ameloblastomas, defined three subgroups. Group I (42%): Luminal (tumor confined to the luminal surface of the cyst), Group II (9%): Intraluminal/plexiform (nodular proliferation into the lumen without infiltration of tumor cells into the connective tissue wall), Group III (49%): Mural (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium). Groups I and II (tumor confined to epithelium of the cyst) may be treated conservatively by enucleation, but lesions in Group III should be treated aggressively in exactly the same manner as solid or multicystic ameloblastomas [8].

Solid ameloblastoma is the most common form of the lesion (86%). It has a tendency to be more aggressive than the other types and has a higher incidence of recurrence [9]. The recurrence rate of solid ameloblastomas after curettage is about 90% for the mandible and 100% for the maxilla, as opposed to 13-15% after resection [10]. Unicystic ameloblastoma has a large cystic cavity with luminal, intraluminal or mural proliferation of ameloblastic tumor. It is a less aggressive variant and it has a low rate of recurrence, although lesions showing mural invasion are an exception and should be treated more aggressively. Peripheral ameloblastoma exists in soft tissue. Treatment of mandibular ameloblastoma still continues to be controversial.

Prior to choosing a treatment for ameloblastomas, the clinicoradiologic variant (solid/multicystic, unicystic, peripheral), anatomic location, clinical behavior, size of the tumor, and age of the patient should be assessed. Besides surgery, treatment may also include use of liquid nitrogen, cryo-radio and chemotherapy. Traditionally it has been considered radio-resistant. Ameloblastomas may become malignant and may even produce distance metastasis. If ever metastases were delivered by an ameloblastoma, the most common sites are lung (76.7%), followed by regional lymph nodes (37.8%), pleura (16.2%), vertebrae (13.5%), skull (10.8%), diaphragm (8.1%), liver and parotid (5.4%) and, even more rarely, the spleen and the kidney [11]. Although several mechanisms for the appearance of metastasis have been proposed, the most likely seem to be hematogenous and lymphangitic spread [12].

On microscopic examination, follicular, acanthomatous, granular cell, basal cell, and desmoplastic subtypes show considerable similarity when these characteristics are compared among the groups. Only the plexiform subtype shows significant variations from this core group of characteristics. Because the follicular subtype is the most commonly encountered variant, some pathologists believe that the acanthomatous, granular cell, basal cell, and desmoplastic variants are subsets of the follicular ameloblastoma [12].

The tumor found in our patient was an unicystic ameloblastoma of the plexiform type. The term “plexiform” refers to the appearance of anastomosing islands of odontogenic epithelium in contrast to a follicular pattern. Histologically, it presents cystic characteristics delimited by a layer of ameloblastic epithelium. There are three types of unicystic ameloblastomas: intraluminal, plexiform (where enucleation is considered the treatment of choice) and mural, that requires marginal resection because of aggressive behaviour and higher recurrence rate [14].

The histological patterns have no prognostic validity, except for unicystic subtypes, which is less aggressive variant and has a low rate of recurrence [15]. The recurrence rate for unicystic ameloblastomas is not zero but is reported to range from 10.7% to almost 25% when treated only by enucleation or curettage [16].

Treatment decisions for ameloblastoma are based on the individual patient situation and the best judgment of the surgeon. The surgical plan should be influenced strongly by the clinicoradiologic variant, anatomic location, clinical behavior, size of the tumor, and age of the patient. Radical treatment alters the facial aesthetics, maxilla-mandibular harmony and the social life of patient. Considering the various studies a conservative approach was used in this patient, rather than a radical approach. More such treated cases should be reported and provide us an insight to the biologic behavior and clinical course of such tumors,
which may help us in effective treatment plan.

References

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Illustrations

Illustration 1

FIGURE 1

Illustration 2

FIGURE 2
Illustration 3

FIGURE 3

Illustration 4

FIGURE 4
Illustration 5

FIGURE 5

Illustration 6

FIGURE 6
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