FOLLICULAR AMELOBLASTOMA: A CASE REPORT WITH BRIEF REVIEW ON MANAGEMENT

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Abstract

Ameloblastoma is one of the infrequent benign neoplasms of the odontogenic epithelium affecting the posterior most areas of the jaws. It occurs in 1% of all the oral tumors and 11% of the odontogenic tumors. Despite being a slow growing neoplasm, the locally invasive nature makes it more aggressive. It is more commonly encountered in mandible (80%) than in the maxilla. High rate of recurrence result in some cases following the routine conservative treatment such as marsupialization and enucleation. In such cases, it is more important to consider the size, location and histopathologic subtypes while planning the treatment procedure. This case report elaborates an extensive mandibular follicular type ameloblastoma of a 60 years old female patient and provides a brief overview of current developments in the treatment approach.

Keywords: Ameloblstoma, case report, surgical management, recurrence

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1. Introduction

Ameloblastoma is a rare benign odontogenic tumor developing from the remnants of odontogenic ectoderm. Mandible (80%) is more commonly involved than the maxilla. It usually occurs as a painless swelling, affecting people in their 3rd to 6th decade of lives with no sex predilection. Lesion may be associated with pain in some cases along with tooth displacement and root resorption. Size of the lesion may vary from 5-115 mm in diameter. Young patients present with unilocular pattern while their elder counterparts exhibit multilocular appearance (1).

Robinson described ameloblastoma as a benign tumor that is "Usually, unicentric nonfunctional, intermittent in growth, anatomically Benign and Clinically Persistent." Ameloblastoma is a benign but locally aggressive tumour with a high propensity to recur which consists of proliferating odontogenic epithelium lying in a fibrous stroma, according to the World Health Organization (WHO,1991) (2).

Conventional radiographs give unilocular / multilocular corticated appearance. The bony septae gives rise to "honey comb / soap bubble" appearance in multilocular lesions. Root resorption may or may not be noted. It also mimics nonodontogenic neoplasms likesimple bone cyst, central giant cell granuloma and fibro osseous lesion, apart from other odontogenic tumors (3).

The benign ameloblastoma is classified by the WHO in 2005 as solid/multicystic, extraosseous/peripheral, desmoplastic, and unicystic forms. Based on the latest genetic studies the WHO classification of these tumors underwent a modification in 2017. It is categorized into three subgroups.1) Unicystic, 2) Conventional (Former name: Solid/Multicystic) and 3) Extraosseous/Peripheral type (4).

As no biologic significance was denoted by the prefix "Solid/Multicystic", it was removed to avoid the confusion with the unicystic variant. Based on the histopathological features, conventional ameloblastoma has been classified into follicular and plexiform type. The follicular type can be further divided into granular, acanthomatous, spindle and basal cell types. Follicular type (27.7%) is the most common histologic variant of ameloblastoma, which is followed by the plexiform type (21.1%) (5).

Recent studies have ruled out the connection between the development of ameloblastoma and some mutations encountered in cell signaling pathways such as mitogen-activated protein kinase (MAPK) and Sonic Hedgehog (SHH). Among the others, BRAFV600Eactivating mutation in MAPK pathway plays a pivotal role in tumor development. Accounting for 63% of ameloblastoma cases, this mutation seems to be occurring irrespective of the site and histological pattern (6).

Simple enucleation or enucleation combined with curettage, as well as the use of adjuvant therapies such as Carnoy's solution and cryotherapy are the conservative treatment modalities. Radical treatment procedure consisting of marginal or block resection, demands an immediate bone reconstruction. Large lesions necessitate facial reconstruction surgeries with grafts and microvascular flaps derived from iliac and fibular bone (7). Though the recent systems state that the prognosis is based upon the histologic growth pattern and early surgical approaches, a high rate of recurrence is noted in conventional ameloblastomas treated with enucleation or curettage comparing with the unicystic variants treated with same procedure(8).

Case report

A female patient, aged 60 years came to the clinic with the chief complaint of swelling in relation to right lower back tooth region. She complained of mild pain in association with the swelling for past 10 days. The patient told the swelling was slow growing in nature and took an year to attain the recent size. Patient had no relevant medical and family history.

Extraoral examination revealed a swelling on the right side of the face extending from the mandibular canine region to the molar region (figure 1). During intraoral examination, the body of the mandible, involving the 43–46 tooth region, showed evidence of buccal cortical expansion and 46 was absent. Overlying mucosa was coral pink in color and appears normal. Consistency of the lesion was firm and was tender on palpation. Radiographic features demonstrated a radiolucent area involving the lower border of the mandible extending from the mesial margin of the 43 anteriorly and mesial margin of the 47 posteriorly (figure 2). Based on all these clinical & radiographic findings and considering the location, initial diagnosis was given as unicystic ameloblastoma.

The exact location and margins of the lesion were determined using the Cone Beam Computed Tomography (CBCT) 3D image (figure 3). Under anaesthesia, a Right Segmental mandiblectomy incorporating the body of the mandible from 44 to 48 tooth region was performed. The resected margins were free of tumor invasion. Two split halves of rib auto grafts were approximated and then were positioned into the mandible with 2.5 mm reconstruction plate and 2.5x10mm screws (figure 4). The lesion was 2.5x2x2 cm in size and appeared to erode the body of the mandible and extending towards parasymphysis. The specimen was then sent for biopsy.

Histopathologic examination revealed islands of odontogenic epithelium with peripheral nuclear pallisading and stellate reticulum-like areas within the fibrous connective tissue (figure 5). Both follicular and plexiform patterns were noted. However, follicular patterns predominated the other type. These cells were infiltrating into the adjacent bony trabeculae. Few areas were showing cystic degeneration with mixed inflammatory cell infiltration and areas of hemorrhage. No mitosis or necrosis identified. Based on the histopathologic picture, diagnosis of Intraosseous follicular Ameloblastoma was given.

2. Discussion

Ameloblastoma is a rare benign odontogenic neoplasm of epithelial origin, exhibiting locally aggressive behavior. The mandible is more frequently affected (80%) than the maxilla. If left untreated, they can reach to large sizes, resulting in facial asymmetry and may associate with other functional problems. The rate of incidence is high in the third and sixth decades of life with no sexual predilection. Though the etiology is not clear, newer studies identified that recurrent mutations occuring in mitogen-activated protein kinase (MAPK) and Sonic Hedgehog (SHH) signaling pathways plays a pivotal role in tumor development. These pathways are crucial during the tooth development period and recent studies also claimed that they are associated with the development of both benign and malignant ameloblstomas (9).

While planning for surgical management, patient's age, size of the tumor, location and duration have to be considered. Based on the involvement of the cortical bone and adjacent soft tissue the treatment plan may be conservative or radical excision. However there still remains a debate regarding the extent of the tumor margin. In a study conducted by Carlson and Marx of 82 ameloblastoma cases, the histopathologic sections showed that the tumor margin extends approximately0.2 to 0.8cm beyond its radiographic margin (10). Hence it is recommended to excise the multicystic tumor with 1-2 cm margin to avoid high recurrence rate. A study by Milman(7) et al demonstrated a significant difference between the recurrence rate of patients treated with segmental surgery (6.1%) and those who underwent a marginal excision, enucleation and curettage (52 %).

Recent developments in molecular research have led to the development of tailored medicines that target the signalling pathways linked to the pathogenesis of ameloblastoma MAPK (Mitogen Activated Protein Kinases) inhibitors selectively inhibit the functions of BRAF (B-Raf proto-oncogene) to halt the uncontrolled proliferation and differentiation of ameloblastic cells. MAPK (Mitogen Activated Protein Kinases) specific medications selectively inhibit the functions of BRAF (B-Raf proto-oncogene) to halt the uncontrolled proliferation and differentiation of ameloblastic cells (6).

3. Conclusion

Given the high recurrence rate and malignant transformation of ameloblastoma, the patient was advised to come in for an annual check-up for the next ten years. One year following the treatment procedure, no signs of recurrence have been observed in the current case. The surgical resection of the tumor with wide margins and simultaneous reconstruction has made the treatment successful. The role of BRAFV600E mutation has to be considered before treating the patients with MAPK specific drugs

Figures

Figure 1. Side profile demonstrates an extra oral swelling in the posterior part of the right mandible

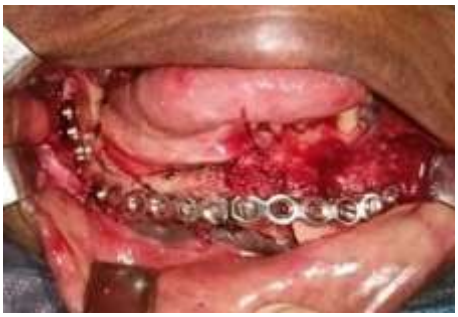

Figure 2. Radiograph illustrates a radiolucent lesion involving the lower border of the mandible

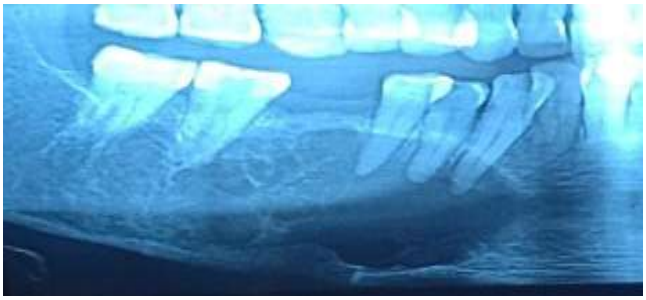

Figure 3. CBCT reveals an osteolytic lesion extending from 43 to 46 tooth region, involving the lower border of the mandible

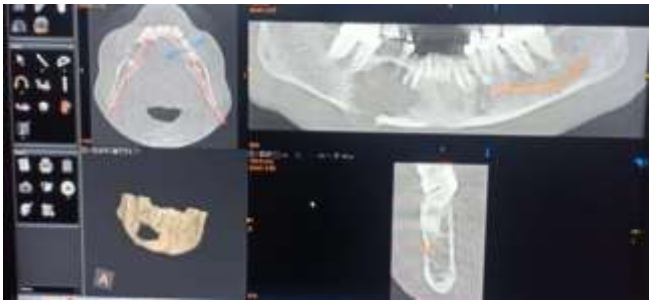

Figure 4.Autografts were approximated and positioned with reconstructions plate and screws

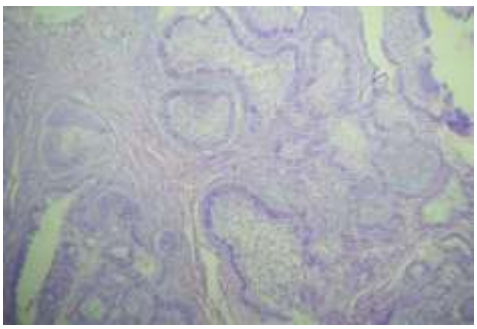

Figure 5.Peripheral nuclear pallisading and stellate reticulum-like areas are observed in the islands of odontogenic epithelium throughout the fibrous connective tissue.

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