Extensive Type III Unicystic Ameloblastoma – A Case Report with Conservative Management

Astha Chaudhry, Manjunath M, Sridevi K , Ishita Gupta, Renu Tanward

Department of Oral Medicine and Radiology , SGT University, Budhera, Gurgaon

Abstract

Unicystic ameloblastoma is a distinct type of ameloblastoma that resembles clinically and radiographically a dentigerous cyst but on three dimensional evaluation shows tumorous behavior and histopathologically shows ameloblastic epithelium. As compared to conventional ameloblastoma it shows a low recurrence rate after conservative treatment. We report a case of extensive unicystic ameloblastoma of mandible in young male patient that showed tumorous characteristics on computed tomography (CT) and was treated conservatively by decompression. CT follow up after 3 months showed significant bone formation. The lesion was then enucleated. Follow up at 8 months showed tremendous bone healing.

Key Words: Ameloblastoma, Carnoy's solution, Computed tomography

Introduction

Ameloblastoma represents 13-54% of all jaw tumors [1]. It is a locally aggressive benign odontogenic neoplasm of epithelial origin. The source of epithelium may be enamel organ, remnants of dental lamina or Hertwig's epithelial root sheath, lining of odontogenic cysts or basal epithelial cells of oral mucosa.

Ameloblastoma may present as 3 biologic variants: solid/ multicystic, unicystic and peripheral [2]. The unicystic variant was described by Robinson and Martinez in 1977 [3]. It arises as a de novo neoplasm and not following secondary cystic changes in the solid counterpart [4]. The pathogenesis of completely cystic lesion is explained by epithelial dysadhesion i.e defective desmosomes or due to the intrinsic production of enzymes like metalloproteinase, serine proteinase that degrade the central zone of the enamel organ after tooth development [5].

Unicystic ameloblastoma radiographically appears similar to dentigerous cyst. But the two can be differentiated following three dimensional evaluation by CT scan. Also CT helps in delineating the exact extent into the hard and soft tissues, guiding the best treatment. Histopathologically, unicystic ameloblastoma is categorized into three subtypes with distinct prognostic and therapeutic implications [6].

This article presents a case of extensive unicystic ameloblastoma in the mandible of young male patient which on conventional radiography appeared to be a dentigerous cyst but on CT and histopathologic examination turned out to be unicystic ameloblastoma type III. Usually, type III lesions are treated aggressively but this case was treated conservatively by decompression, thus preventing esthetic and functional complications for the patient. The article also emphasizes on the importance of CT in the diagnosis and follow up of such lesions.

Case Report

A 27 years old male patient reported with a complaint of pain and swelling in the lower third of face on right side since 4 months. Swelling preceded the pain with associated history of occasional pus discharge from the mouth and reduction in mouth opening. No contributing medical history was reported.

On extraoral examination, diffuse swelling was noted in right middle and lower third of face with a smooth overlying surface, hard consistency, tender, with movable overlying skin. Mouth opening was 30mm. Buccal cortical plate expansion was noted in the region of 28,29,30,31. Pus discharge was noted from the distal pocket of 31. Retromolar mucosa was tender on palpation. However, no lingual cortical plate expansion was noted.

A clinical diagnosis of an infected dentigerous cyst in relation to 32 was made. The patient was subjected to panoramic radiograph (OPG), computed tomography (CT scan), fine needle aspiration cytology (FNAC) and routine blood investigations.

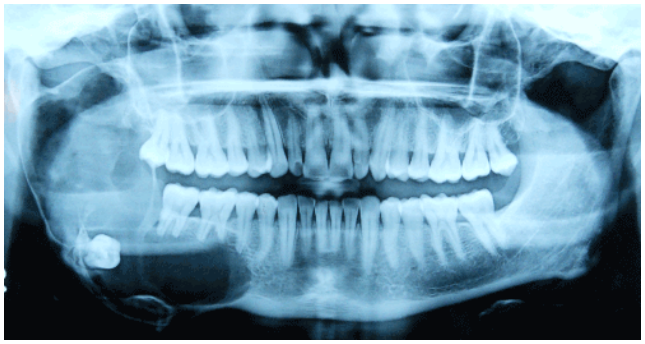

Figure 1: Orthopantomogram: Showing well defined unilocular radiolucency in the right side of mandible associated with impacted and displaced 32 and cortical expansion at the ramus, sigmoid notch, coronoid process and inferior border of mandible.

OPG revealed a well defined unilocular radiolucency surrounded by corticated borders associated with impacted 32 involving the right body, angle, ramus of the mandible that extended till the condylar process, sigmoid notch and coronoid process approximately measuring 6X9cm. 32 was displaced apically and posteriorly to the angle of mandible. Cortical expansion and thinning was seen at the inferior border of mandible, sigmoid notch, ramus and the anterior

Corresponding author: Dr. Astha Chaudhry, House no. 617, 1st floor, Sector -9, Gurgaon-122001, Haryana, India, Tel: 09711807733; Fax: - +91 124 2278151; Email: dr.asthac@gmail.com

border of ramus. External root resorption was seen in 29,30,31 (*Figure 1*).

CT scan axial section showed a unilocular lesion in association with impacted 32. The expansion of buccal as well as lingual cortex was appreciated. Cortical thinning and perforation was seen in the region of 30 *(Figure 2)*.

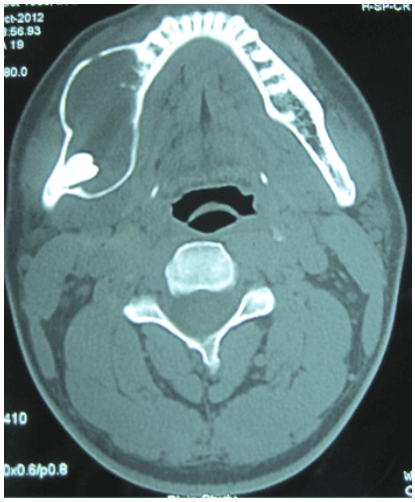

Figure 2: *CT (axial section): Showing well defined unilocular radiolucency with bicortical expansion and attachment of the lesion below the CEJ of 32.*

The sagittal and coronal section showed the extent of the lesion in the angle, ramus, sigmoid notch and coronoid process and expansion of the inferior border as well as cortical perforation in the inferior part. The apical and backward displacement of 32 was seen (*Figure 3*).

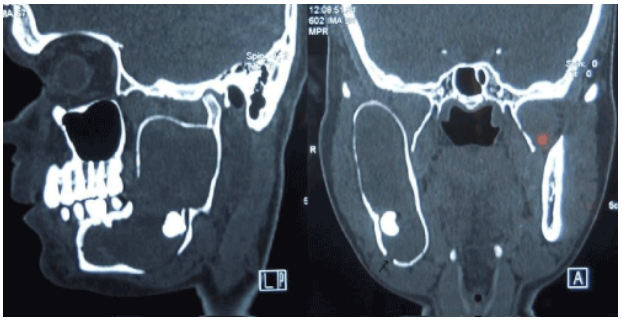

Figure 3: Sagittal and coronal section: Showing the extent of ameloblastoma, cortical expansion and perforation in the inferior part (arrow).

The blood investigations showed values within normal limits. Aspiration yielded a thick material and on microscopic examination showed abundant mixed inflammatory cells consisting predominantly of neutrophils and few epithelial squames. Cytopathological features were suggestive of infected odontogenic keratocyst (OKC) (*Figure 4*).

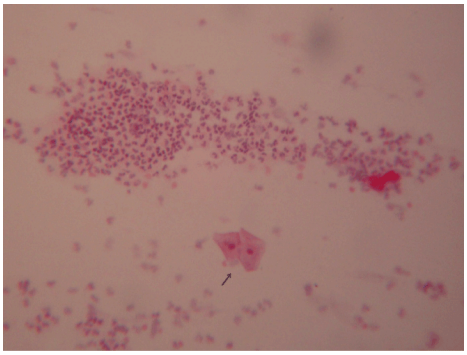

Figure 4*: Cytological smear : Showing inflammatory cells and epithelial squames (arrow).*

The patient was subjected to conservative management. Irrigation into the bony cavity *(Figure 5)* was done every alternate day under antibiotics and analgesics.

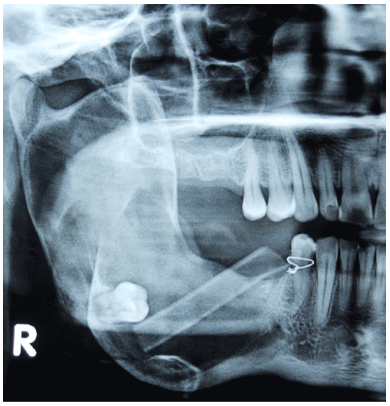

Figure 5: OPG showing the drain placed inside the lesion for decompression.

Three months follow up CT scan revealed significant bone formation at the inferior border of mandible, at the inner surfaces of cortices, above the impacted tooth and anteriorly at 28 region. The cortical perforations were repaired *(Figure 6)*.

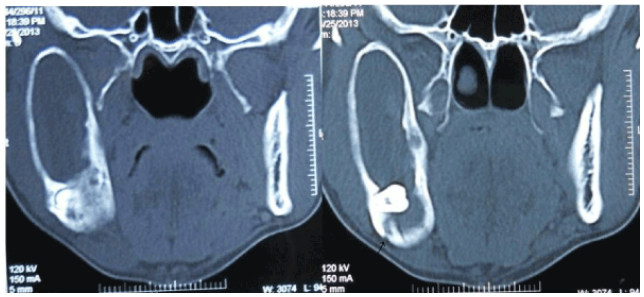

Figure 6: Follow up at 3rd month: Showing bone formation around the impacted tooth, thickening of the cortices and repair of perforation at the inferior border (arrow).

The lesion was then enucleated and carnoy's solution (Mixture of chloroform, absolute alcohol, glacial acetic acid,

ferric chloride) was applied for few minutes followed later by the irrigation of the bony cavity.

The enucleated specimen showed cystic epithelium comprising of tall columnar cells mimicking ameloblasts with hyperchromatic nuclei and stellate reticulum like cells. Focal areas showed luminal proliferation of the epithelium in plexiform pattern. Outer fibrous wall showed multiple ameloblastic islands with cystic degeneration, squamous metaplasia and areas of hyalinization. The histologic picture was diagnostic of unicystic ameloblastoma type III (*Figure 7*).

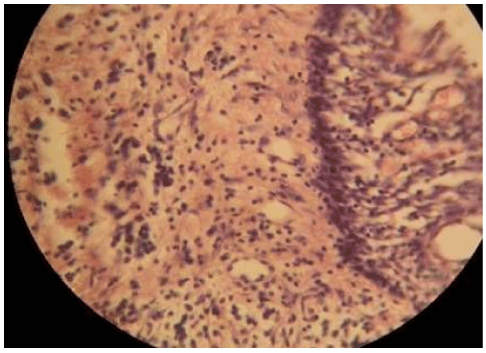

Figure 7: Histopathology: Showing ameloblastic epithelium with hyperchromatic nuclei and stellate reticulum like cells.

The follow up radiograph after 8 months showed significant bone formation anteriorly in the region of 29, at the angle and inferior border of mandible with significant reduction in the expansion of the inferior border (*Figure 8*). The patient is on continued follow up with continued irrigation of the bony cavity.

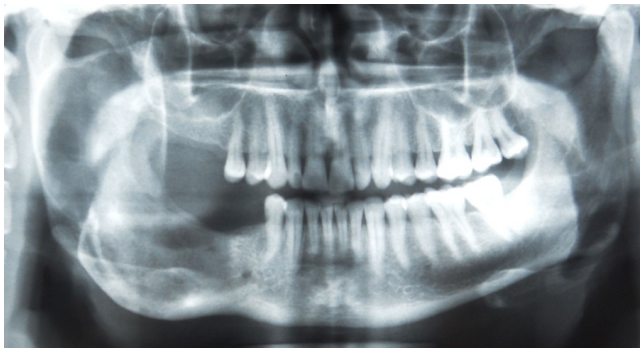

Figure 8: Follow up at 8th month: OPG showing significant bone formation and reduction in cortical expansion.

Discussion

Unicystic ameloblastoma accounts for about 10-15% of all ameloblastomas and presents as sharply demarcated monolocular radiolucency with a corticated border. Sometimes this monolocular radiolucency may show septae and is usually associated with an impacted tooth in a pericoronal position. This appearance is similar to dentigerous cyst, adenomatoid odontogenic tumor (AOT) and odontogenic keratocyst (OKC). In our case, same appearance was seen and provisionally it was thought to be dentigerous cyst associated with impacted 32. However, with the CT scan it was appreciated that the pericoronal radiolucency was not attached to the cementoenamel junction (CEJ) of the involved tooth and was extended much lower down. The buccal cortex was expanded in tumorous fashion as the expanded cortex was making a smooth obtuse angle with normal cortex anteriorly and an abrupt acute angle with the unexpanded cortex posteriorly [7]. The CT also showed bicortical expansion which ruled out the possibility of dentigerous cyst. OKC is classically known to run in antero-posterior direction without causing much cortical expansion and it rarely causes root resorption [7]. In our case, the FNAC report suggested the diagnosis of OKC due to the presence of epithelial squames. But that could be the result of squamous metaplasia seen in the cystic lining of ameloblastoma. AOT is uncommon in the posterior region.

The extent of the lesion and aspiration results in the present case demanded aggressive surgical treatment in the form of hemimandibulectomy. But such aggressive treatment would result in loss of teeth, esthetics and function, contributing to the reduced quality of life of patient. Considering these facts in addition to patient's request and young age, conservative treatment was planned. The treatment options included enucleation, enucleation followed by carnoy's solution, marsupialization followed by enucleation and marginal resection [8]. Since, the lesion size was extremely large, it was decided to reduce the lesion size by decompression followed later by enucleation.

The patient responded well and CT showed good bone formation at third month. The lesion was enucleated and final diagnosis of unicystic plexiform ameloblastoma type III was made. Type I lesions show unilocular cystic lesion with epithelium exhibiting features of ameloblastoma. Type II shows intraluminal extension of the epithelial nodules from cystic lining. In Type III, there is invasion of connective tissue wall of the cyst by islands of ameloblastomatous epithelium. Curettage is acceptable for luminal and intraluminal unicystic ameloblastoma as the tumor does not extend beyond the basement membrane of the cyst. Once the tumor invades into the wall of the cyst, then it has a solid component which needs to be treated aggressively [4].

In the present case, the type III lesion was treated conservatively by enucleation followed by carnoy's solution application for few minutes to minimize recurrence. Carnoy's solution (chloroform 3 ml, absolute alcohol 6 ml, glacial acetic acid1 ml, ferric chloride 1 g) was initially described as a sclerozing agent for the treatment of cysts and fistulae [9]. Stoelinga and Bronkhorst in 1987 advocated its use after conservative treatment of unicystic ameloblastoma [10]. Lee et al [11] have showed that use of carnoy's solution after enucleation of type III unicystic ameloblastoma results in lower recurrence rate.

The architectural pattern of ameloblastoma is such that the tumor extends into the cancellous bone beyond its apparent macroscopic surface and radiographic boundaries [12]. Marx et al [13] have showed that ameloblastoma cells penetrated about 2-8mm into the trabecular spaces of bone beyond the apparent radiographic margin. Carnoy's solution has been shown to penetrate cancellous bone to a depth of 1.5 mm [14]

thereby fixing the residual ameloblastomatous tissue after enucleation diminishing the risk of recurrence.

The prognosis of ameloblastoma is also dependent on the histologic type. The follicular, granular cell and acanthomatous types have been shown to have relatively high likelihood of recurrence, while the desmoplastic and plexiform types show a low potential for recurrence [15,16]. In our case, plexiform type was noted, thus favored conservative management.

Conclusion

Unicystic ameloblastoma, despite being a low grade variant of ameloblastoma may show aggressive behavior especially the type III. Hence, this lesion should always be included in the differential diagnosis of swellings of the maxillofacial region. A thorough clinical, radiological and histopathological evaluation must be performed to diagnose such lesions and plan the treatment accordingly. CT scan is indispensable in progressing towards the accurate diagnosis, to know the extension and subsequently adapting the best treatment plan. It also helps in monitoring of bone formation and early diagnosis of recurrences. The choice of correct treatment should include consideration of age, type, anatomic extent and histology of ameloblastoma. Conservative treatment should be the first choice in younger individuals for better quality of life. However, long term follow up should be done for early diagnosis of recurrences and their management

References

1. Reichart PA, Philipsen HP, Sonner S. Ameloblastoma: biological profile of 3677 cases.

2. *European Journal of Cancer Oral Oncology*. 1995; **31B:** 86-99.

3. Sivapathasundharam B, Rajendran R. *Shafer's textbook of Oral Pathology*. 5th edition. Elsevier, UK. 2006.

4. Robinson L, Martinez MG. Unicystic ameloblastoma: A prognostically distinct entity. *Cancer.* 1977; **40:** 2278.

5. Black CC, Addante R R,Mohila CA. Intraosseous ameloblastoma. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology*. 2010; **110:** 585-592.

6. Rosenstein T, Pogrel MA, Smith RA et al. Cystic ameloblastoma- Behaviour and treatment of 21 cases. *Journal of Oral and Maxillofacial Surgery*. 2001; **59:**1311-1316.

7. Ackermann GL, Altini M, Shear M. The unicystic ameloblastoma: a clincopathological study of 57 cases. *Journal of Oral Pathology and Medicine.* 1988; **17:** 541–546.

8. Langlais RP, Langland OE, Nortje CJ. Diagnostic imaging of the jaws. 2ndedition *Williams & Wilkins*, Baltimore.

9. Sampson DE, Pogrel MA. Management of mandibular ameloblastoma: the clinical basis for a treatment algorithm. *Journal of Oral Pathology and Medicine*. 1999; **579:**1074-1047.

10. Cutler EC, Zollinger R. Sclerosing solution in the treatment of cysts and fistulae. *The American Journal of Surgery*. 1933; **19:** 411–418.

11. Stoelinga PJW, Bronkhorst FB. The incidence, multiple presentations and recurrence of aggressive cysts of the jaws. *Journal of Cranio-Maxillo-Facial Surgery*. 1988; **16:**184-195.

12. Lee PK, Samman N, Ng IO. Unicystic ameloblastoma- Use of Carnoy's solution after enucleation. *International Journal of Oral and Maxillofacial Surgery*. 2004; **33:** 263-267.

13. Ghandi D, Ayoub AF, Pogrel MA et al. Ameloblastoma: a surgeon's dilemma. *Journal of Oral and Maxillofacial Surgery*. 2006; **64:** 1010-1014.

14. Marx RE, Smith BH, Smith BR. Swelling of the retromolar region and cheek associated with limited opening. *Journal of Oral and Maxillofacial Surgery*. 1993; **51:** 304–309.

15. Voorsmit RACA. The incredible keratocyst: a new approach to treatment. *Dtsch Zahnarztl Z* 1985; **40:** 641–644.

16. Hong J, Yun PY, Chung IH, et al. Long-term follow up on recurrence of 305 ameloblastoma cases. *International Journal of Oral and Maxillofacial Surgery*. 2007; **36:** 283–238.

17. Ueno S, Mushimoto K, Shirasu R. Prognostic evaluation of ameloblastoma based on histologic and radiographic typing. *Journal of Oral and Maxillofacial Surgery*. 1989; **47:** 11-15.