Unicystic Intraluminal Ameloblastoma: An Unusual Case Report

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Abstract

Ameloblastoma is a slow-growing, persistent and locally aggressive neoplasm of epithelial origin with high rate of recurrence. This is the second most common odontogenic tumour (odontoma is the most common). Ameloblastomas typically occur as hard painless lesions near the angle of the mandible in the region of the 3rd molar tooth (48 and 38) although they can occur anywhere along the alveolus of the mandible (80%) and maxilla (20%). According to the WHO, ameloblastomas are classified into the following types: conventional, unicystic, and peripheral. Unicystic ameloblastoma (UA) refers to those cystic lesions that show clinical, radiographic, or gross features of a mandibular cyst, but on histologic examination shows a typical ameloblastomatous epithelium lining part of the cyst cavity, with or without luminal and/or mural tumor growth. Here a case of unicystic ameloblastoma of mandible in young female and its management is reported.

Key words- *Ameloblastoma, unicystic, odontogenic*

Introduction

Ameloblastoma is a true neoplasm of enamel organ type tissue which does not undergo differentiation to the point of enamel formation. It was described by Robinson as benign tumor that is "usually unicentric, non-functional, intermittent in growth.[1] The term "ameloblastoma" as applied to this particular tumor was suggested by Churchill in 1934 to replace the term "adamantinoma", coined by Malassez in 1885, since the latter term implies the formation of hard tissue & no such material is present in this lesion.[1-3] It is generally surrounded by a fibrous capsule, which is occasionally penetrated in some areas by proliferating tumor. It is a distinctive odontogenic tumor, grows slowly and persistently. The tumor spreads into the cancellous marrow spaces, without concomitant resorption of the trabecular bone. It is considered to be locally invasive & recurs frequently after surgical procedures.[1]

Thus the tumor may be derived from [1,2]

1. Cell rests of the enamel organ, either remnants of the dental lamina or remnants of hertwig's sheath, the epithelial rest of malasez.
2. Epithelium of odontogenic cysts, particularly the dentigerous cyst, & odontomas.
3. Disturbances of the developing enamel organ.
4. Basal cells of the surface epithelium of the jaws or
5. Heterotrophic epithelium in other parts of the body, especially the pituitary gland.

Ameloblastoma is the second most common odontogenic neoplasm, and only odontomes outnumbers it in reported frequency of occurrence.[1,3] Excluding odontoma, the incidence of ameloblastoma is at least equal to the incidence of all other odontogenic neoplasm combined. According to the reports in the literature approximately 80% of the tumours are found in the mandible.[2] The molar ramus area is most frequently involved. The frequent occurrence of the tumour at the angle of the mandible is explained by the fact that the

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posterior end of the dental lamina proliferates continuously & the aberrant tooth germs are most often found in this region.[3]

McForland & Patterson & Robinson indicated that irritation may be a causative factor for these tumors.[1] New has also mentioned that the lower 3rd molar have the most difficulty in erupting & that this region of the mouth receives the greatest amount of irritation which may account for the higher incidence at the angle.[11] This assumption may also explain the high incidence of ameloblastoma associated with impacted teeth. Here a case of unicystic ameloblastoma of mandible in young female patient and its management is described.

Case Report

A 29 year old female patient reported to the department of Oral & Maxillofacial Surgery, at Rural Dental College & Hospital with chief complaint of swelling on the left side of mandible since last 2 years. Swelling was gradually increased in size in the past 2 years to attain the present size of concern. Past medical history of patient was not significant. Past dental history of patient revealed history of extraction of left premolars at private dentist 6 months back. Patient's general condition was fair & vital signs were stable. There was no evidence of pallor, icterus & cynosis. On extraoral examination swelling 3 x 6 cm in size was present on the left side of mandible, anteriorly it starts from left corner of mouth extends and posteriorly 1 cm anterior to left angle of mandible. Superiorly it extends from 2 cm below ala of nose extends inferiorly till the lower border of mandible. (Figure 1) The swelling was hard in consistency with smooth surface, diffuse margins and was nontender on palpation. Pain associated with the swelling was insidious in origin and dull, nonradiating, and intermittent in nature. Left side submandibular lymph nodes were palpable, nontender, mobile & firm in consistency. Oral opening & TMJ movements were within normal limit.

Intraoral examination revealed swelling which extends from lower left lateral incisor till lower left second molar. (Figure2) Intraoral pus discharge was evident. Obliteration of buccal vestibule was seen. Overlying mucosa was normal. On palpation, a swelling was palpable from lower left lateral incisor region to distal to the lower left second molar region, with expansion of both buccal & lingual cortices. The buccal and lingual cortices were thinned out. OPG showed a unilocular radiolucency of approximately 3cm x 6cm in size with sclerotic borders extending from 32 regions to 37 regions with distal extension into the angle of mandible (Figure 3). 33, 34, 35 were missing; root resorption of 36 & 37 was seen.

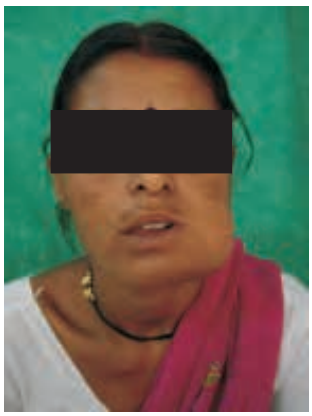

Figure 1 Preoperative Extraoral view showing swelling on the left side of mandible

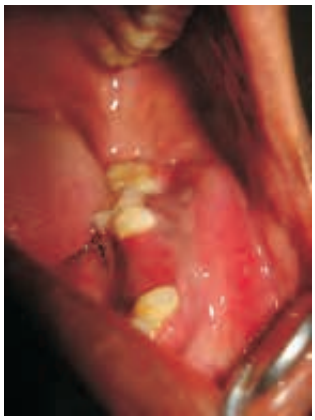

Figure 2 Preoperative Intraoral view showing swelling which caused obliteration of buccal vestibule.

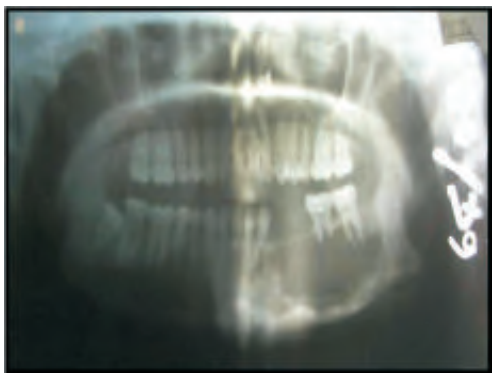

Figure 3 Preoperative OPG showing unilocular radiolucency with sclerotic borders extending from 32 to distal to 37.

Aspiration biopsy was done. Aspirate was found to contain pus & few inflammatory cells. From clinical & radiological examination a provisional diagnosis of cystic lesion of the left side mandible was given. As the lesion was large in size and locally invasive incisional biopsy of swelling was carried out, histopathologic report was suggestive of unicystic ameloblastoma of left side mandible. Thus on clinical & histologic examination the final diagnosis of unicystic ameloblastoma of left side mandible was confirmed. As lesion was locally invasive in nature, resection of left side of mandible with preservation of condylar & coronoid process was carried out. (Figure 4) Defect was reconstructed with 16 hole stainless steel reconstruction plate. (Figure 4 & 5) Number 10 infant feeding tube was sutured extraorally as a drain. Intraorally closure was achieved with 3-0 Vicryl. Layerwise suturing was done extraorally with 3-0 vicryl interrupted sutures for deep layers & suturing of the skin with 3-0 mersilk. The specimen was sent for histopathological examination in 10% formalin solution. Postoperatively recovery was uneventful.

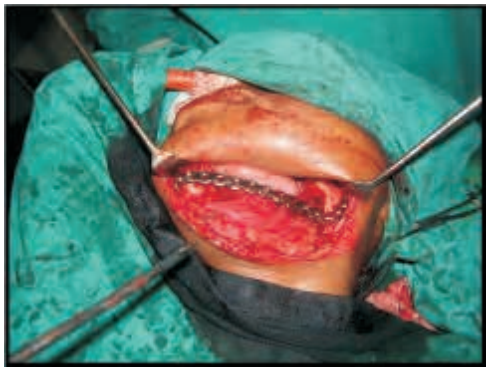

Figure 4 . Resection of the left side of mandible & Reconstruction with reconstruction plate

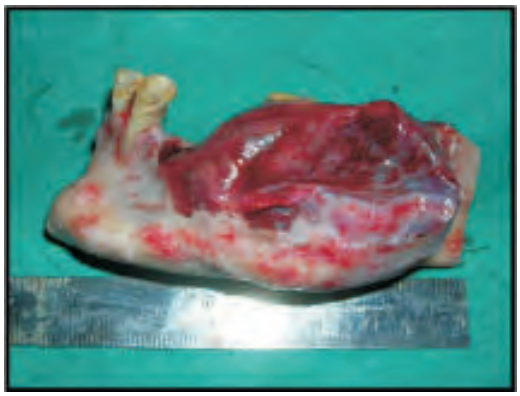

Figure 5. Specimen Buccal View

Discussion

Ameloblastomas (previously known as an adamantinoma of the jaw) are benign, locally aggressive tumors that arise from the mandible, or less commonly from the maxilla.[1,3,5] They are widely described as tumors that present most commonly in the 4th decade of life, however the average age of 38 years decreased to 21 years.[5] The ameloblastoma occur with equal frequencies in two sexes.[1,6] The average age of patient at the time of discovery of the lesion is approximately 33 years. The typical ameloblastoma begins insidiously as a central lesion of bone which is slowly destructive, but tends to expand the bone rather than perforate it. Occasionally a patient allow ameloblastoma to persist for many years without treatment, and in such cases, though the expansion may be extremely disfiguring, ulcerative type of growth characteristic of carcinoma does not occur.[7] Seldom there is breakdown of the oral mucosa.

In this patient the lesion was started as a small swelling which was gradually increased to present size. (Figure1&2) The patient was having lesion of approximately 3x6 cm since last 3 years. Age of the patient was 29 years & it was present in the mandibular molar area extending distally into the left angle of mandible. (Figure 2) and there was expansion of buccal as well as lingual cortices.[6,7] The resorption of teeth roots is an extremely common finding in association with ameloblastoma. Massive tumors readily fenestrate the cortical bone & periosteum but do not penetrate the oral mucosa. Ulceration is probably created by impingement of tooth or by extraction of a tooth in the tumor site.

Ameloblastoma appears macroscopically as solid or cystic lesion. However, a sharp difference does not exist between the two layers, since zones of both solid tumor & cystic spaces are present in almost all growths. It would appear that ameloblastoma begins as a solid tumor & gradually becomes more cystic with age, probably as a result of the degenerative transformation of the tumor. On the other hand, ameloblastoma have developed in the walls of dentigerous cysts.[8,15] Radiographically ameloblastoma has been described classically as a multilocular cystlike lesion of the jaw.[4,6,14-17] This is especially true in advanced cases of ameloblastoma.[11,13&15] The tumor exhibits a compartmented appearance with septa of bond extending into the radiolucent tumor mass. The lesion is a unilocular in many cases & presents no characteristic or pathognomonic features.[1,8,9] In this patient OPG

showed unilocular radiolucency of approximately 3 cm x 6 cm in size with sclerotic borders extending from 32 region to 37 region with distal extension into the angle of mandible. 33, 34, 35 were missing , root resorption of 36 & 37 was seen.(Figure 3) Radiographic findings are important in the prediction of the clinical course of the tumor, because the unicystic type exhibits less aggressive biologic behavior than does the multicystic type.[10,12, 18- 20]

Unicystic Ameloblastoma (UA), first described by Robinson and Martinez in 1977, refers to those cystic lesions that show clinical radiologic or gross features of a mandibular cyst, but on histologic examination show a typical ameloblastomatous epithelium lining part of the cyst cavity, with/without luminal and/or mural tumor growth.[17,18,20] It accounts for 5-10% of all intraosseous ameloblastomas.3–5 UA is believed to be less aggressive and responds more favourably to conservative surgery than the solid or multicystic ameloblastomas.[6] Histologically, the minimum criteria for diagnosing a lesion as UA is the demonstration single cystic sac lined by odontogenic (ameloblastomatous) epithelium often seen only in focal areas.[11,18-20] In a clinicopathologic study of 57 cases of UA, Ackermann 18 classified this entity into three histologic groups:

I- luminal UA (tumor confined to the luminal surface of the cyst);

II- intraluminal/plexiform UA (nodular proliferation into lumen without infiltration of tumor cells into connective tissue wall); and

III- mural UA (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium

Another sub grouping by Philipsen and Reichar 18 has also been described as follows: Subgroup 1: luminal UA;

1.2: luminal and intraluminal;

1.2.3: luminal, intraluminal and intramural; and

1.3: luminal and intramural.

The UA diagnosed as subgroups 1 and 1.2 can be treated conservatively, whereas subgroups 1.2.3 and 1.3 showing intramural growths require radical resection.[3]

In the present case, radiologic finding was suggestive of a cystic lesion of mandible and FNAC report was inconclusive. As the lesion was a large in size extending from 33- 37 region and locally invasive in nature incisonal biopsy of cystic lining was performed before surgery. Histopathologic examination of cystic lining revealed Unicystic Ameloblastoma, intraluminal type. (Figure 8) Hence, finally taking into account of clinical and radiologic features and histopathologic examination resection of left side of mandible with preservation of condylar & coronoid process was carried out.(Figure 4) Reconstruction of defect was done with stainless steel reconstruction plate.(Figure 5) Patient's recovery was unevenful. The

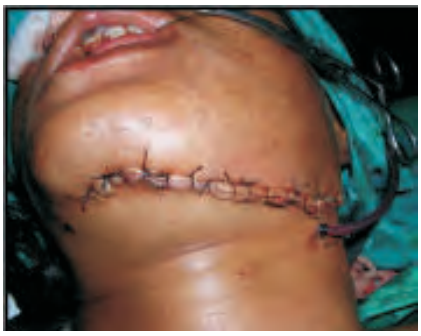

Figure 6. Immediate postoperative

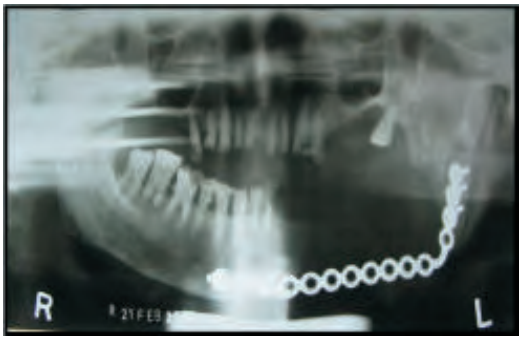

Figure 7. Postoperative OPG

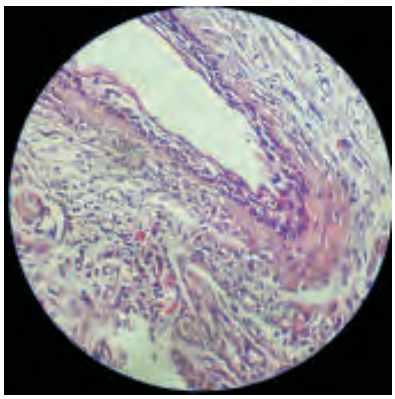

Figure 8. Hematoxylin and Eosin stained sections showing cystic area lined by ameloblastic epithelium with cuboidal to columnar basal cell layer with hyperchromatic nuclei. Thin superficial layer consisting of stellate reticulum like cells is also evident. (H and E, 10×) Plaque like intraluminal proliferation is seen at some area.

patient is being followed up at regular intervals to check for any recurrences.(Figure 6 & 7)

There is no recurrence for 2 years period after that patient lost follow up. (Figure 7) Recurrence of unicystic ameloblastoma (UA) is also related to the histologic subtypes, among which those invading the fibrous wall have a rate of 35.7%, but others have a rate of only 6.7%.[10] Though UA is considered to be less aggressive form of ameloblastoma, in our patient the lesion was aggressive in nature and large in size extending from 32-distal to 37 with extension into left angle of mandible. There was perforation of both buccal as well as lingual cortices hence resection of left side of mandible was performed with preservation of condylar & coronoid process.

Conclusion

Ameloblastoma has a high rate of local recurrence if it is not adequately removed. In our opinion, radical surgical resection of ameloblastoma is the treatment of choice in aggressive and large size tumor to avoid recurrence.

Conflict of interests

The author reports no conflicts of interest related to this study.

References

1. Neville BW, Damn DD, Allen CM, Bouqout JK, editors. 2nd ed. Philadelphia: WB Saunders Co; 2002. Oral and maxillofacial pathology
2. Lagares DT, Cossío PI, José M, Pérez JUL. Mandibular ameloblastoma. Med Oral Patol Oral Cir Bucal (Ed.impr.) Valencia mayo. 2005; 10:231–8
3. Torres-Lagares D, Infante-Cossío P, Hernández-Guisado JM, Gutiérrez-Pérez JL. Mandibular ameloblastoma. A review of the literature and presentation of six cases. Med Oral Patol Oral Cir Bucal.2005;10:231–8.
4. Mehlisch DR, Dahlin DC, Masson JK. Ameloblastoma: A clinicopathologic report. J Oral Surg. 1972;30:9–22.
5. Pinsolle J, Michelet V, Coustal B, Siberchicot F, Michelet FX. Treatment of ameloblastoma of the jaws. Arch Otolaryngol Head Neck Surg. 1995;121:994–6.
6. Kameyama Y, Takehana S, Mizohata M, Nonobe K, Hara M, Kawai T, et al A clinicopathological study of ameloblastomas. Int J Oral Maxillofac Surg. 1987;16:706–12.
7. Robinson L, Martinez MG. Unicystic ameloblastoma: A prognostically distict entity. Cancer.1977;40:2278–82.
8. Williams TP. Management of ameloblastoma: A changing perspective. J Oral Maxillofac Surg.1993;51:1064–70.
9. Stanley HR, Jr, Krogh HW. Peripheral ameloblastoma; Report of a case. Oral Surg Oral Med Oral Pathol.1959;12:760–5.
10. Corio RL, Goldblatt LI, Edwards PA, Hartman KS. Ameloblastom carcinoma: A clinicopathologic study and assessment of eight cases. Oral Surg Oral Med Oral Pathol. 1987;64:570–6.
11. Adekeye EO. Ameloblastoma of the jaws: A survey of 109 Nigerian patients. J Oral Surg. 1980;38:36–41.
12. Cawson RA, Binnie WH, Speight PM, Barrett AW, Wright JM, Thorogood P. 5th ed. London: Churchill Livingstone; 1998. Lucas's pathology of tumors of the oral tissues; pp. 25–44.
13. Shatkin S, Hoffmeister FS. Ameloblastoma: A rational approach to therapy. Oral Surg Oral Med Oral Pathol. 1965;20:421–35.
14. Mehlisch DR, Dahlin DC, Masson JK. Ameloblastoma: A clinicopathologic report. J Oral Surg. 1972;30:9–22.
15. Sehdev MK, Huvos AG, Strong EW, Gerold FP, Willis GW. Ameloblastoma of maxilla and mandible. Cancer. 1974;33:324–33.
16. Tsaknis PJ, Nelson JF. The maxillary ameloblastoma: An analysis of 24 cases. J Oral Surg.1980;38:336–42.
17. Vedtofte P, Hjorting-Hansen E, Jensen BN, Roed-Peterson B. Conservative surgical treatment of mandibular ameloblastomas. Int J Oral Surg. 1978;7:156–61.
18. Robinson L, Martinez MG. Unicystic ameloblastoma: A prognostically distinct entity. Cancer.1977;40:2278–85.
19. Gardner DG, Corio RL. Plexiform unicystic ameloblastoma. A variant of ameloblastoma with a low-recurrence rate after enucleation. Cancer. 1984;53:1730–5.
20. Gardner DG. A pathologist's approach to the treatment of ameloblastoma. J Oral Maxillofac Surg.1984;42:161–6.