Unicystic ameloblastoma of mandible: imaging features and literature review

Abstract

Ameloblastoma are benign tumors whose importance lies in its potential to grow into enormous size with resulting bone deformity. They are typically classified as Unicystic, multicystic, peripheral and malignant subtypes. Unicystic ameloblastoma (UCA) refers to those cystic lesions that show clinical, radiographic, or gross features of an odontogenic cyst, but on histological examination show a typical ameloblastomatous epithelium lining, with or without luminal and/or mural tumor growth. We present a very rare case of Unicystic ameloblastoma in a girl child with an age of 10 years, clinical and radiographic features of UCA, its differential diagnosis, histopathology and current concepts of management has also been discussed in the presenting paper.

Keywords: ameloblastoma, computed tomography, imaging of ameloblastoma, Unicystic ameloblastoma

Introduction

Ameloblastoma is the most common benign odontogenic tumor accounting for approximately 1% of tumors and cysts of the jaw and 10% of all the odontogenic tumors. It is a slow-growing, persistent and locally aggressive neoplasm that may originate from the epithelium involved with the formation of teeth such as enamel organ, odontogenic rests of Malassez, reduced enamel epithelium and odontogenic cyst lining. Ameloblastoma may occur centrally within the bone or peripherally, without an intraosseous component in the soft tissues overlaying the alveolar ridge. Intraosseous lesions are of two types solid/conventional/multicystic and unicystic. Unicystic Ameloblastoma (UA), a variant of ameloblastoma first described by Robinson & Martinez in 1977, refers to those cystic lesions that show clinical and radiologic characteristics of an odontogenic cyst, but in histologic examination, show a typical ameloblastomatous epithelium lining part of the cyst cavity with or without luminal and/or mural tumor proliferation. Prior to the report by Robinson and Martinez, this variant had been referred to as a mural or intraluminal ameloblastoma. Recognition of this growth pattern is very important because of its tumor proliferation. Prior to the report by Robinson and Martinez, this variant had been referred to as a mural or intraluminal ameloblastoma.

Case report

A ten year old female child came with a complaint of swelling on right lower side of her jaw since one month which was insidious in onset and got gradually progressed to present size. It was associated with severe, intermittent and dragging type of pain which radiates to right ear. Pain relieved with medication. It was also associated with extra oral swelling since 15 days. On Extraoral examination facial asymmetry was seen in the right lower third of the face. On Intra oral examination, a solitary diffuse swelling was seen in the buccal vestibular region of 85 and 46, extending anteroposteriorly from middle third of 85 to distal surface of 46. Superoinferiorly 0.5cms below the marginal gingiva of 85 and 46 extending into the buccal vestibule with vestibular obliteration. Sinus opening with pus discharge was seen on the attached gingiva of 85 (buccal or lingual). On palpation, swelling was tender, hard in consistency, non-compressible, non-reducible. Expansion of buccal cortical plate was felt in 85 and 46. Based on the history and clinical examination a provisional diagnosis of ameloblastoma was made. Under differential diagnosis ameloblastic fibroma, odontogenic keratocyst was considered. IOPA, Occlusal view, Orthopantanogram and CT scan was taken. IOPA taken in 85 and 46 (Figure 1) showed diffuse radiolucency distal to 46 with absence of 47 tooth bud. OPG (Figure 2) view showed a solitary, well defined radiolucency of size 3x2cms extending anteroposteriorly from mesial root of 46 to 1cm from the posterior border of ramus of the mandible, superoinferiorly 1cm below the sigmoid notch to inferior border of mandible thinning the inferior cortical margin. Internal structure was radiolucent with the displaced tooth bud of 47 in the ramus region. Panoramic CT section (Figure 3) and Sagittal CT section (Figure 4) is showing hypodense area of size 2.5x3cms surrounding the developing tooth bud of 47. Axial CT section (Figure 5A) showing tooth within the hypodense area and axial CT section (Figure 5B) showing buccal cortical plate expansion with breakdown of lingual cortical plate. An incisional biopsy was done and it showed epithelial lining with ameloblast-like cells and adjacent connective tissue stroma. There was no luminal proliferation of epithelium, suggestive of intra luminal ameloblastoma (Figure 6). Following the diagnosis, the parents were informed about the condition and proposed treatment. Surgical enucleation along with chemical cauterization with Carnoy’s solution (Figure 7) was done under general anesthesia along with extraction of 47 (Figure 8) considering age of the patient. Patient is under follow up, with no functional or aesthetic complaints. 6 months post treatment OPG shows signs of new bone formation (Figure 9).

Discussion

UA accounts for 6% to 15% of all intraosseous ameloblastomas. It is less aggressive, usually occurs in an earlier age group than the solid or multicystic with about 50% of the cases occurring in the second decade of life. As in the present case more than 90% are located in the mandible. In most cases UA are associated with impacted tooth, mandibular third molar being the most common. The term unicystic...
is derived from the macroscopic and microscopic appearance, the lesion being essentially a well-defined, often large monocystic cavity with a lining, focally but rarely entirely composed of odontogenic (ameloblastomatous) epithelium. The pathogenesis of cystic ameloblastomas remains obscure. Some investigators believe that UCA arises from pre-existing odontogenic cysts, in particular a dentigerous cyst, while others maintain that it arises de novo. The reason why some ameloblastomas become completely cystic may be related to epithelial dysadhesion (e.g., defective demosomes) or, more likely, to the intrinsic production of proteinases (e.g., metalloproteinases, and serine proteinases); enzymes that normally degrade the central zone of the enamel organ after tooth development. Radio-graphically, the unilocular: multilocular ratio is 13:3 when the lesion is associated with an impacted tooth. For the 'non-dentigerous' variant this ratio changes to 8:7. Further, the 'dentigerous' type occurs on average 8 years earlier than the 'non-dentigerous' variant. Lastly, the mean age for unilocular, impaction-associated UAs is 22 years, whereas the mean age for the multilocular lesion unrelated to an impacted tooth is 33 years. Ackerman et al., classified unicystic ameloblastoma into three types with prognostic and therapeutic implications.

Figure 1 Intraoral periapical radiograph showing diffuse radiolucency distal to 46 with the absence of 47 tooth bud.

Figure 2 Pre-op OPG showing solitary, well defined radiolucency of size 3x2cms surrounding the crown of 47 tooth bud which got displaced into the ramus region with thinning of inferior border of mandible.

Figure 3 Panoramic CT section showing complete extension of the lesion.

Figure 4 Sagittal CT section showing hypodense area surrounding the developing tooth bud of 47.

Figure 5 (A) Axial CT section showing tooth within the hypodense area.

Figure 5 (B) Axial CT section is showing buccal cortical plate expansion with breakdown of lingual cortical plate.

Figure 6 Low power histopathological picture of unicystic ameloblastoma showing intraluminal proliferation.

Group I: Luminal UA (tumor confined to the luminal surface of the cyst).

Group II: Intra luminal/plexiform UA (nodular proliferation into the lumen without infiltration.

Citation: Avinash TML, Paramkusam G, Bhayya H, et al. Unicystic ameloblastoma of mandible: imaging features and literature review. J Dent Health Oral Disord Ther. 2018;9(4):298–302. DOI: 10.15406/jdhodt.2018.09.00396
of tumor cells into the connective tissue wall).

Group III: Mural (invasive islands of ameloblastomatous epithelium in the connective tissue wall not involving the entire epithelium).

The microscopic pattern that exhibits mural invasion in UA suggests a more aggressive potential.\(^\text{11}\)

**Figure 7** Surgical site of enucleation.

**Figure 8** Surgical specimens with Extracted 47.

Another histologic subgrouping by Philipsen & Reichart\(^\text{12}\) has also been described as follows:

1. Subgroup 1: Luminal
2. Subgroup 1.2: Luminal and intraluminal
3. Subgroup 1.2.3: Luminal, intraluminal and intramural
4. Subgroup 1.3: Luminal and intramural.

The Unicystic Ameloblastomas diagnosed as subgroups 1 and 1.2 can be treated conservatively (enucleation), whereas subgroups 1.2.3 and 1.3 showing intramural growths require radical resection, as for a solid or multicystic ameloblastoma. Following enucleation, vigorous curettage of the bone should be avoided as it may implant foci of ameloblastoma deeper into bone. Chemical cauterization with carnoy’s solution\(^\text{13}\) is also advocated for subgroups 1 and 1.2. Subgroups 1.2.3 and 1.3 have a high risk for recurrence, requiring more aggressive surgical procedures.\(^\text{14}\) Recurrence rates for Unicystic Ameloblastoma after conservative surgical treatment (curettage or enucleation) are generally reported to be less than 25%. For intraluminal and plexiform type of Unicystic Ameloblastoma recurrence rate was found to be as low as 10.7%.\(^\text{15}\) Recurrence rates for Solid Multicystic Ameloblastoma was found to be about 50 to 90%. The present analysis included only publications in English. All well-documented publications during the last 20 years were collected, and several clinicopathological features of each case were studied. The following data were recorded: age (less than or equal to 10 years), sex, location, clinical features/symptoms, histological type, radiographic appearance, treatment. Included only reports of Unicystic ameloblastoma in children less than 10 years confirmed by histological analysis with all the data required for tabulation were included and the Articles not having enough information were excluded (Table 1).

| S. no | Year | Author | Sex | Age | Location | Clinical features | Histological features | Radiological features | Treatment |
|-------|------|--------|-----|-----|----------|-------------------|----------------------|----------------------|-----------|
| 1     | 1998 | Li et al.\(^\text{16}\) | F   | 10  | Mandible | Mild fullness over the cheek | Unicystic ameloblastoma | UL      | Enucleation |
| 2     | 2000 | Li et al.\(^\text{17}\) | M   | 5   | Maxilla (premolar to second molar) | Cystic lesion | Mural type | INA     | Enucleation |
| 3     | 2003 | Al-Khateeb & Ababneh\(^\text{18}\) | F   | 9   | Mandible | Painless swelling | Unicystic ameloblastoma | UL      | Enucleation plus peripheral ostectomy |
| 4     | 2007 | Huang et al.\(^\text{19}\) | M   | 9   | Body–angle of the mandible | INA | INA | UL | Enucleation and peripheral ostectomy |
| 5     | 2008 | Qureshi et al.\(^\text{20}\) | F   | 10  | Mandible | Mild fullness over the cheek | Unicystic ameloblastoma | UL | Enucleation, curettage |

Citation: Avinash TML, Paramkusam G, Bhayya H, et al. Unicystic ameloblastoma of mandible: imaging features and literature review. J Dent Health Oral Disord Ther. 2018;9(4):298–302. DOI: 10.15406/jdhodt.2018.09.00396
Table Contined

| S. no | Year | Author | Sex | Age | Location | Clinical features | Histological features | Radiological features | Treatment |
|-------|------|--------|-----|-----|----------|-------------------|----------------------|--------------------|-----------|
| 6     | 2008 | Gulen U et al. | M   | 8   | Right Mandible | painless hard swelling | unicystic ameloblastoma | UL | Enucleation and extraction of related teeth |
| 7     | 2011 | Chacko & Kuriakose | M   | 9   | Mandible | Pain and swelling in relation to the right side of the lower jaw | Plexiform unicystic ameloblastoma | UL | Enucleation, curettage |
| 8     | 2011 | Kalaskar et al. | M   | 9   | Right maxilla | Painless swelling | Unicystic Ameloblastoma with intra-luminal proliferations | UL | Enucleation+ Carnoy’s solution and extraction of related teeth |
| 9     | 2011 | Ponniah et al. | F   | 8   | Left ramus of the mandible | Painless swelling on the left side of the mandible | Unicystic ameloblastoma | UL | Enucleation then segmental resection |
| 10    | 2011 | Sudhakar K Reddy et al. | M   | 6   | Anterior mandible | Slow growing painless swelling | Unicystic ameloblastoma | UL | Enucleation and extraction of related teeth followed by application of Carnoy’s solution |
| 11    | 2012 | Scariot et al. | F   | 9   | Right mandibular body | Painless swelling | Plexiform unicystic ameloblastoma | UL | Curettage with extraction of two adjacent teeth |
| 12    | 2013 | Bhutia O et al. | M   | 5   | Right Mandible | painless hard swelling | type I unicystic ameloblastoma | UL | Enucleation of the cyst with extraction of the involved teeth followed by application of Carnoy’s solution |
| 13    | 2013 | Arora S et al. | F   | 3   | Left Maxilla | Bony hard swelling | Unicystic ameloblastoma (Type 1.2) | UL | Enucleation of the cyst with extraction of the involved teeth |
| 14    | 2014 | Present case | F   | 8   | Right Mandible | Swelling with mild pain | Unicystic ameloblastoma | UL | Enucleation with chemical cautery |

Conclusion

Ameloblastomas in children differ from adults with a higher percentage of unicystic tumors. Unicystic ameloblastoma is a tumor with a strong propensity for recurrence, especially when the ameloblastic focus penetrates the adjacent tissue from the wall of the cyst. Although enucleation has been claimed to give acceptable recurrence rates in unicystic ameloblastoma, there are no large series with long follow-up in children. The histologic pattern that exhibits mural invasion in unicystic ameloblastoma suggests that more aggressive surgery is necessary. Present case was treated with Carnoy’s solution along with the enucleation, which suggests a possible benefit against recurrence.

Acknowledgements

None.

Conflict of interest

The author declares that there is no conflict of interest.

References

1. Olaitan AA, Adekeye EO. Clinical features and management of ameloblastoma of the mandible in children and adolescents. Br J Oral Maxillofac Surg. 1996;34(3):248–51.
2. Gerzenshtein J, Zhang F, Caplan J, et al. Immediate mandibular reconstruction with microsurgical fibula flap transfer following wide resection for ameloblastoma. J Craniofac Surg. 2006;17(1):178–82.
3. Reichart PA, Philipson HP, Sonner S. Ameloblastoma: biological profile of 3677 cases. Eur J Cancer B Oral Oncol. 1995;31B(2):86–99.
4. Robinson L, Martinez MG. Unicystic ameloblastoma: A prognostically distinct entity. Cancer. 1977;40(5):2278–2285.
5. Gardner DG, Corio RL. The relationship of plexiform unicystic ameloblastoma to conventional ameloblastoma. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 1983;56:54–60.
6. Pizer ME, Page DG, Svirsty JA. Thirteen-year follow-up of large recurrent unicystic ameloblastoma of the mandible in a 15-year-old boy. J Oral Maxillofac Surg. 2002;60(2):211–215.
7. Philipson HP, Reichart PA. Unicystic ameloblastoma. A review of 193 cases from the literature. Oral Oncol. 1998;34(5):317–325.
8. Kessler HP. Intraosseous Ameloblastoma. Oral Maxillofac Surg Clin North Am. 2004;16(3):309–322.
9. Rosenstein T, Pogrel AM, Smith RA, et al. Cystic ameloblastoma: Behavior and treatment of 21 cases. J Oral Maxillofac Surg. 2001;59(1):1311–1316.
10. Ackermann GL, Altini M, Shear M. The unicystic ameloblastoma: A clinicopathological study of 57 cases. J Oral Pathol. 1988;17(9-10):541–546.

Citation: Avinash TML, Paramuskam G, Bhavya H, et al. Unicystic ameloblastoma of mandible: imaging features and literature review. J Dent Health Oral Disord Ther. 2018;9(4):298–302. DOI: 10.15406/jdhodt.2018.09.00396
11. Ord RA, Blancaert RH, Nikitakis NG, et al. Ameloblastoma in children. J Oral Maxillofac Surg. 2002;60:762–770.
12. Philpens HP, Reichert PA. Classification of odontogenic tumors and allied lesions. In: Odontogenic tumors and Allied lesions. Quintessence Pub. Co Ltd: 2004:309–322.
13. Lee PK, Samman N, Ng IO. Unicystic ameloblastoma—use of Camoy’s solution after Enucleation. Int J of Oral Maxillofac Surg. 2004;33(3):263–267.
14. Rakesh RS, Suraj M, Tanveer HS. Unicystic ameloblastoma of the mandible An unusual case report and review of literature. Head and Neck Oncol. 2010;2:1–5.
15. Gardner DG, Corio RL. Plexiform unicystic ameloblastoma. A variant of ameloblastoma with a low-recurrence rate after enucleation. Cancer. 1984;53(8):1730–1735.
16. Li TJ, Kitano M, Arimura K, et al. Recurrence of unicystic ameloblastoma: a case report and review of the literature. Arch Pathol Lab Med. 1998;122(4):371–374.
17. Li TJ, Wu YT, Yu SF, et al. Unicystic ameloblastoma: a clinicopathologic study of 33 Chinese patients. Am J Surg Pathol. 2000;24(10):1385–1392.
18. Al-Khateeb T, Ababneh KT. Ameloblastoma in young Jordanians: a review of the clinicopathologic features and treatment of 10 cases. J Oral Maxillofac Surg. 2003;61(1):13–18.
19. Huang YJ, Lai ST, Chen CH, et al. Surgical management of ameloblastoma in children. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2007;104(4):478–85.
20. Qureshi SS, Qureshi SS, Medhi SS, Kane SV. Unicystic ameloblastoma of the mandible masquerading as carcinoma of the oral cavity in a 10 year old girl. Am J Surg. 2008;196(3):e7–e9.
21. Gulten U, Vedat T, Hilal A. Unicystic ameloblastoma in 8 years old child: a case report review of unicystic ameloblastoma. Int Dent J. 2008;58(1):29–33.
22. Chacko V, Kuriakose S. Conservative management of a case of plexiform ameloblastoma. Dent Update. 2011;38(5):336–338.
23. Kalaskar R, Unawane AS, Kalaskar AR. Conservative management of unicystic ameloblastoma in a young child: report of two cases. Contemp Clin Dent. 2011;2:359–63.
24. Ponniah I. Recurrent unicystic ameloblastoma in a child. J Oral Maxillofac Pathol. 2011;15(2):236–238.
25. Sudhakara Reddy K, Girish Rao S. Unicystic Ameloblastoma in 6-Year-Old Child and Its Significance. World J Dentistry. 2011;2(4):363–366.
26. Scariot R, da Silva RV, da Silva Felix W. Conservative treatment of ameloblastoma in child: a case report. Stomatologia. 2012;14:33–36.
27. Bhutia O, Roychoudhury A, Arora A, et al. Management of unicystic ameloblastoma of the mandible in a 5-year old child. Natl J Maxillofac Surg. 2013;4(2):232–234.
28. Arora S, Kumar P, Urs AB, et al. Unicystic ameloblastoma in 3 year old paediatric patient – A rare entity. J Clin Exp Dent. 2013;5(1):e54–e57.