Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Abbreviated Key Title: Sch J Med Case Rep **ISSN 2347-9507 (Print)**

A United of Scholars Academic and Scientific Society, India

Acanthomatous Ameloblastoma: A Rare Case Report

Neha Vaid[1], Deepak Bhargava[2], Puja Bansal[3], Rajeshwar Chawla[4], Deepak Goyal[5], Ruby Bansal[1]
[1]Assistant Professor, JCD Dental College, Sirsa, India
[2]Professor & HOD, School of Dental Sciences, Sharda University, Greater Noida, India
[3]Reader, School of Dental Sciences, Sharda University, Greater Noida, India
[4]Professor & HOD, JCD Dental College, Sirsa, India
[5]Reader, JCD Dental College, Sirsa, India

***Corresponding author**
Neha Vaid

Article History
Received: 08.12.2017
Accepted: 17.12.2017
Published:30.12.2017

DOI:
10.36347/sjmcr.2017.v05i12.013

Abstract: Ameloblastoma is the most common clinically significant odontogenic tumor of the gnathic arches. Ameloblastomas show wide morphological spectra and may pose diagnostic difficulties. Ameloblastomas are locally aggressive jaw tumors with a high propensity for recurrence that are believed to arise from remnants of odontogenic epithelium, lining of odontogenic cysts and basal layer of overlying oral mucosa. Of the many types encountered, acanthomatous ameloblastoma is a rare variant that possesses distinctive features. This paper reports a rare case of ameloblastoma in 50 year old male patient and its diagnosis using clinical, radiological and histological findings.
Keywords: Acanthomatous ameloblastoma, squamous metaplasia.

INTRODUCTION

Ameloblastoma accounts for approximately 1% of all oral tumors [1] Ameloblastoma is a benign [2] & most frequent odontogenic tumor arising from dental epithelium, and is characterized by its histological resemblance to the enamel organ of the developing tooth germ, yet enamel formation is not observed [3]. It is often aggressive and destructive, with the capacity to attain great size, erode bone and invade adjacent structures [2]. Guzak first described it in 1826, and in 1879 Falkson performed a histological description [4].

This lesion was first designated as an "Adamantinoma" by Malassez in 1885, while the term ameloblastoma was coined by Ivy and Churchill in 1930 [5]. Acanthomatous ameloblastoma is considered as an aggressive tumor of the canine jaw, characterized by irregular verrucous masses adjacent to the tooth. In 1993 Gardner and Baker described that acanthomatous epulides were a type of ameloblastoma that developed from the gingival epithelium (peripheral) or from alveolar bone (intraosseous) [6]. Acanthomatous ameloblastoma has an aggressive local behaviour and often invades bone in the periodontal apparatus but it does not metastasize to other organs [7].

This paper reports a case of an acanthomatous ameloblastoma presenting as a unilocular radiolucency in the ramus region on the right side of the mandible in a 50-year-old male patient, which showed histopathological features of acanthomatous pattern.

CASE HISTORY

The present case report is about a 50-year-old male patient, reported to the Department of Oral and Maxillofacial Pathology with a chief complaint of swelling in the right lower back teeth region since 1 year. The present history revealed that the swelling was slow growing, gradual in onset over a period of one year and was not associated with pain, tenderness or discharge. There was history of tooth extraction with respect to right mandibular first molar 2 years back. Past medical history was unremarkable and non-significant.

Extra oral examination revealed facial asymmetry on the right side. Swelling measuring 2.3cmx1cm in dimensions as shown in Figure-1. Anterioposteriorly swelling extending from mentalis to ramus of the mandible. Superiorly 2cm away from the angle of mouth to inferiorly lower border of the mandible. On palpation swelling was hard, non tender, non fluctuant.

Intraorally no swelling was present. Obliteration of vestibule- from 46 to right posterior region of mandible, Caries- 46 (proximal caries), Missing-38,45,48 (shown in figure 2). No lymphadenopathy or fistulae were present.

On radiographic investigations OPG revealed unilocular radiolucency present irt 44,46,47 & surrounding the crown of 48 measuring around 2cm x 1cm in dimension. Resorption & displacement of roots

of involved teeth were also present as shown in Figure 3.

A provisional diagnosis of Dentigerous Cyst, Unicystic ameloblastoma, Odontogenic keratocyst, Residual cyst, Ossifying fibroma, intraosseous squamous cell carcinoma was made.

An incisional biopsy was carried out which showed numerous epithelial cell nests present in connective tissue stroma. At the periphery, follicle was lined by single layer of tall columnar ameloblast like cells with reverse polarity. In center island showed loosely arranged polygonal or angular cells resembling stellate reticulum, some follicle showed cystic degeneration of central stellate reticulum like cells (Figure 4). Many follicles showed squamous metaplasia. Thus, on clinical, radiographical and histopathological findings, final diagnosis was given of acanthomatous ameloblastoma.

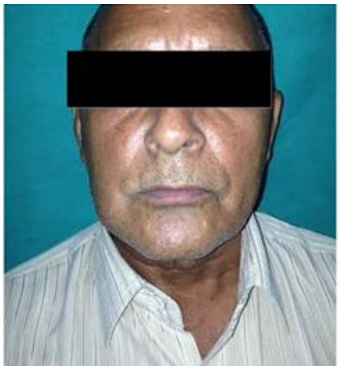

Fig-1: Clinical presentation of the lesion showing swelling on the right lower facial region.

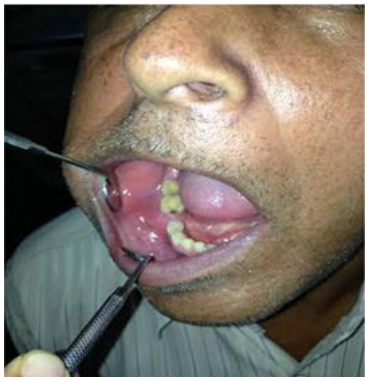

Fig-2: Intraorally obliteration of vestibule- from 46 to right posterior region of mandible.

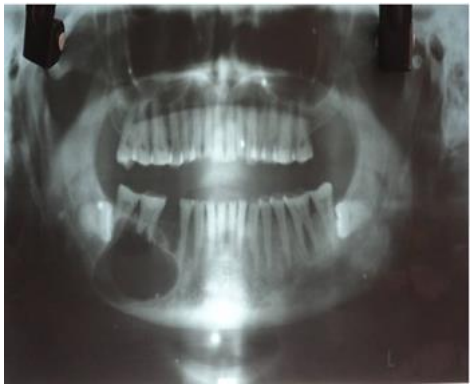

Fig-3: Orthopantomograph showing a unilocular radiolucency irt 44,46,47 & surrounding the crown of impacted 48

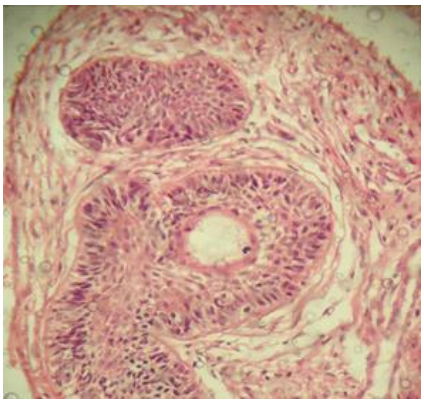

Fig-4: Photomicrograph showing solid epithelial cell nests with peripheral palisading columnar cells and central stellate reticulum like cells, in some follicle cystic degeneration also present in the stellate reticulum like cells

DISCUSSIONS

Ameloblastoma is derived from the English word “amel” which means enamel and the Greek word “blastos” which means the germ. It arises from the epithelium of the dental lamina, and it is characterized by its local aggressive behavior and a high recurrence rate [9]. It was described by Robinson in 1937, as a benign tumor that is “usually unicentric, nonfunctional, intermittent in growth, anatomically benign and clinically persistent [10].” The World Health Organization (WHO) (1991) defined ameloblastoma as a benign but locally aggressive tumor with a high tendency to recur, consisting of proliferating odontogenic epithelium lying in a fibrous stroma [9].

Histopathologically, ameloblastoma resembles normal odontogenic/enamel epithelium and ectomesenchyme. Odontogenesis consists of chronographic and reciprocal interactions between the ectomesenchymal cells, which are derived from the neural crest, and the oral cavity lining epithelium [11]. It is suggested that the tumor may be originating from a) cell rests of the enamel organ, either remnants of the dental lamina or hertwig’s root sheath, the epithelial rests of malassez; b) epithelium of odontogenic cysts; c)

disturbances of the developing enamel organ; or d) basal cells of the surface epithelium [4] heterotopic epithelial from other parts of the body, perhaps [11].

Molecular study of ameloblastoma by Kahn stated for human papilloma virus (HPV). Further studies have found various subtypes of HPV associated with a minority of ameloblastomas, the most common being HPV 6 [11].

Table-1: showing Tumor marker & their expression in ameloblastoma [12]

ABCG2, CD-133, Bmi-1	Increased	Oncogenesis Cell differentiation Malignant potential
Syndecan-1	Decreased	Aggressiveness Invasiveness
α5β1 integrin	Increased	Invasiveness
Podoplanin	Increased	Invasiveness through Collective cell migration
MT	Increased	Invasiveness High recurrence
Ki-67	Increased in peripheral cells	Aggressiveness
PCNA	Unicystic ameloblastoma Follicular ameloblastoma Plexiform ameloblastoma Ameloblastic Carcinoma	Recurrence Aggressiveness
Cyclin D1	Increased	Invasiveness Aggressiveness Poor prognosis Lymph node metastasis
HGF & c-Met	Increased	Malignant transformation
Anti-apoptotic (bcl-2, bcl-x)	Peripheral cells	Cytodifferentiation alignant transformation
Fas, FasL	Central cells in Acanthomatous and Granular cell ameloblastoma	Cytodifferentiation
p14ARF, MDM2, p53	Increased	Neoplastic transformation
Rb	Increased	Cell proliferation Differentiation
PTEN	Increased	Aggressiveness
MMP-1, -2, -9	Increased	Aggressiveness Invasiveness
PTHrP	Increased	Infiltrative growth & destructive behaviour
RANKL, TNF-α	Increased	Osteoclastogenesis tumor expansion
PTCH1	Increased	Proliferation of odontogenic epithelium
BMP-2, -4, -7	Increased	Cytodifferentiation apoptotic cell death
β-catenin	Increased	Cell migration

ABCG2: ATP-binding cassette subfamily G member 2, PCNA: Proliferating nuclear cell antigen, HGF: Hepatocyte growth factor, Rb: Retinoblastoma, PTEN: Phosphatase and tensin, PTHrP: Parathyroid hormone-related protein, RANKL: Receptor activator of nuclear factor–κB ligand, TNF-α: Tumor necrosis factor-alpha, BMP: Bone morphogenic protein, MT: Metallothenin.

Classification of ameloblastoma

Based on the clinical, radiographic, histopathologic and behavioral aspects [13] and the International Agency for Research on Cancer, 2003 [9] it is further classified into:

- Solid/multicystic
- Extraosseous/peripheral
- Desmoplastic ameloblastoma
- Unicystic

The solid/multicystic is the most common type, comprising 91 % of the ameloblastomas in the largest series. This is followed by the unicystic type 6 %, the extra osseous ameloblastoma 2 %, and the desmoplastic type 1 %. The most aggressive clinical/pathologic association is seen in the

solid/multicystic type, which is associated with the highest recurrence rate of up to 90 % with conservative operations such as enucleation and curettage. The unicystic type is the most benign. Unlike solid, unicystic and desmoplastic ameloblastomas which are centered within the marrow space, encapsulated by bone, and thus are designated ‘‘central ameloblastoma’’, the peripheral ameloblastomas are extra-osseous and do not involve the underlying bone [11].

Six histopathologic subtypes of solid ameloblastoma include follicular, plexiform, acanthomatous, basal cell, granular and desmoplastic ameloblastoma [9].

Waldron CA *et.al*. 1987 identified as predominantly common patterns in ameloblastomas the follicular (64.9%), the plexiform (16.9%) and the follicular/plexiform association (12.9%). Adebiyi *et al*. reported that most of the solid lesions belonged to the follicular type (70.4%), plexiform (14.1%) and acanthomatous (4.2%). Reichart *et al*. also reported the following solid lesions as being the most common patterns found: follicular (35.4%), plexiform (31.5%) and acanthomatous (11.8%) [4].

Acanthomatous type is a benign tumor, but is locally aggressive and frequently invades the alveolar bone or recurs after marginal surgical excision [6]. Acanthomatous ameloblastoma is a rare variant with distinctive features. Acanthomatous ameloblastomas has been found to occur more commonly in elder patients rather than younger ones[9]. 80% of ameloblastomas occur in the mandible, usually in the posterior (molar-ascending ramus) region, and 10-15% may be associated with a non-erupted tooth [5]. The present case was seen in a 50-year-old male, in the angle and ramus region of the mandible, and was associated with an impacted mandibular third molar.

Radiographically ameloblastomas varies from unilocular (cyst-like) to multilocular (presence of bony septae which create internal compartments), with well-defined sclerotic margins which may appear scalloped or may expand the cortical plate [5]. Our case presented as a large unilocular radiolucency in association with 44,46,47 & surrounding the crown of impacted third molar measuring around 2cm x 1cm in dimension.

Histopathological features of both follicular and plexiform variants, possessing islands and anastomosing strands of odontogenic epithelium enmeshed in a fibrous stroma. These islands and strands contain basal cells that are columnar or cuboidal, hyperchromatic and lined up in a palisaded fashion at the periphery. The cells have vacuolated cytoplasm and the nuclei exhibit reverse polarization away from the connective tissue. The cells in the central portions of the epithelial islands are more loosely arranged, mimicking stellate reticulum [3]. In granular cell ameloblastoma, cytoplasm of stellate reticulum-like cells appear coarse granular and eosinophilic. Basal cell type, the epithelial tumor cells are less columnar and arranged in sheets. Desmoplastic variant is composed of the dense collagen stroma, which appears hypocellular and hyalinised [11], while the acanthomatous type shows extensive squamous metaplasia and keratin formation in the central portion of the islands [5]. Squamous metaplasia such as that seen in acanthomatous ameloblastoma may be attributed to chronic irritation. Calculus and oral sepsis (which could be a source of chronic irritation) have been suggested to play a role in aetiology of ameloblastoma [3]. The present case showed numerous follicles of tumor cells in connective tissue stroma. At the periphery, follicle was lined by single layer of tall columnar ameloblast like cells with reverse polarity. In center island showed loosely arranged polygonal or angular cells resembling stellate reticulum. Many follicles showed squamous metaplasia. Some follicle showed cystic degeneration of central stellate reticulum like cells. Multinucleated reactive giant cells were also found.

The treatment of ameloblastomas depends on the clinico-radiologic variant, anatomic location and clinical behavior of the tumor. The age and the general state of health of the patient are also important factors [5]. The diagnosis has to be confirmed by a biopsy, but occasionally in cystic variants it may be made only after excision. Ameloblastomas, although benign are relentlessly infiltrative. Conventional ameloblastoma infiltrates into surrounding bone and extends beyond the apparent radiographic boundaries seen in plain radiographs. The rate of recurrence reported varies from 20-90% [14]. Surgical resection is the fastest and most curative treatment for AA [7]. If possible, conservative surgery can be used if an assured complete removal can be performed [6]. Surgical treatment of ameloblastoma may be either conservative or radical. The conservative approach is usually carried out for unicystic/small unilocular lesions and includes enucleation, curettage or surgical excision with peripheral osteotomy. Radical treatment measures are advocated for large lesions, which include marginal or segmental resection of the diseased section of the jaw and inclusion of about 1 or 2 cm of apparently uninvolved bone. Supra-periosteal resection of the bone is necessary when extensive thinning or perforation of the cortical plates is noted. Therapeutic irradiation should not be used in the treatment of ameloblastomas as it can lead to osteonecrosis and has a potential for inducing post-radiation malignancies [5]. Intralesional (IL) chemotherapy is another option for treating AA. In 1998, Yoshida *et al*. studied the effectiveness of IL bleomycin on four dogs with AA; all the dogs studied had a positive response to the treatment. In humans, bleomycin is typically administered subcutaneously, intramuscularly or intravenously weekly at 10–20 U m.

Side effects are generally mild and consist of gastrointestinal effects, myelosuppression, allergic reactions, renal toxicity, hepatotoxicity and pulmonary toxicity [7].

CONCLUSION

Ameloblastomas are uncommon benign odontogenic neoplasms that rarely become malignant. Careful clinical examination combined with thorough and histopathological confirmation of the diagnosis will allow for the selection of the best individual therapeutic approaches, increasing the treatment efficacy in patients diagnosed with this tumor. The present case has some interesting findings distinguished it from other reported cases of ameloblastoma. First, this is a case of acanthomatous ameloblastoma which is an extremely rare variant. Second, radiographically unilocular radiolucency was seen. Finally histologically numerous follicles of tumor cells in connective tissue stroma. At the periphery, follicle was lined by single layer of tall columnar ameloblast like cells with reverse polarity. In center island showed loosely arranged polygonal or angular cells resembling stellate reticulum. Many follicles showed squamous metaplasia. Some follicle showed cystic degeneration of central stellate reticulum like cells. Multinucleated reactive giant cells were also found. The case highlights the diagnostic challenges and the importance of clinicopathological correlation in the diagnosis.

REFERENCES

1. Sandra F, Mitsuyasu T, Nakamura N, Shiratsuchi Y, Ohishi M. Immunohistochemical evaluation of PCNA and Ki-67 in ameloblastoma. Oral oncology. 2001 Feb 28;37(2):193-8.
2. Bansal N, Sheikh S, Bansal R, Sabharwal R, Gupta A, Goyal A, Kainth N. Acanthomatous ameloblastoma of mandible crossing the midline: A rare case report. Annals of African medicine. 2015 Jan 1;14(1):65.
3. Bhargava A, Saigal S, Chalishazar M. Acanthomatous ameloblastoma of mandible. Journal of dental sciences & Research. 2011;2(2):1-5.
4. Dahiya P, Kamal R. Acanthomatous ameloblastoma of mandible–a rare case report. Annals of Dental Research. 2014 Jan 23;3(2):42-6.
5. Figueiredo NR, Meena M, Dinkar AD, Khorate M. Ameloblastoma of the acanthomatous and plexiform type in the mandible presenting as a unilocular radiolucency. Indian Journal of Oral Sciences. 2015 Jan 1;6(1):34.
6. Singh G, Agarwal R, Kumar V, Passi D. Acanthomatous ameloblastoma-a case report. Journal of International Oral Health: JIOH. 2013 Apr;5(2):54.
7. Kelly JM, Belding BA, Schaefer AK. Acanthomatous ameloblastoma in dogs treated with intralesional bleomycin. Veterinary and comparative oncology. 2010 Jun 1;8(2):81-6.
8. Mayer MN, Anthony JM. Radiation therapy for oral tumors: canine acanthomatous ameloblastoma. The Canadian Veterinary Journal. 2007 Jan;48(1):99.
9. Masthan KM, Anitha N, Krupaa J, Manikkam S. Ameloblastoma. Journal of pharmacy & bioallied sciences. 2015 Apr;7(Suppl 1):S167.
10. Rajendran R. Shafer's textbook of oral pathology. Elsevier India; 2009.
11. McClary AC, West RB, McClary AC, Pollack JR, Fischbein NJ, Holsinger CF, Sunwoo J, Colevas AD, Sirjani D. Ameloblastoma: a clinical review and trends in management. European Archives of Oto-Rhino-Laryngology. 2016 Jul 1;273(7):1649-61.
12. Spandana P, Shylaja S, Sekhar MS, Krishna A, Bhavani SN, Raj Y. Molecular etiopathogenesis of ameloblastoma-Current concepts revisited. J Med Radiol Pathol Surg. 2015;1(2):3-7.
13. Barnes L, Eveson JW, Reichart P, Sidransky D. World Health Organization classification of tumours: pathology and genetics of head and neck tumours. Word Health Organization Classification of Tumours: Pathology and genetics of head and neck tumors. 2005.
14. Angadi PV. Head and neck: odontogenic tumor: ameloblastoma.Atlas Genet Cytogenet Oncol Haematol 2011;15(2):223-29.