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Commentary

The Pouchy Dewlap-Ameloblastoma

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Ameloblastoma is a commonly discerned, benign neoplasm arising from odontogenic epithelium. Characteristically, an expansible, gradually progressive, locally aggressive tumefaction with possible reoccurrence is delineated.

Tumefaction exhibits diverse microscopic variants as follicular, plexiform, basal, acanthomatous, granular or desmoplastic. Metastasizing ameloblastoma is an exceptional neoplasm demonstrating metastasis to lymph nodes, lung or diverse viscera with possible occurrence of organ-associated symptoms. Tumefaction arises secondary to multiple surgical procedures, neoplasms of extensive duration or radiation therapy [1,2].

Conventional ameloblastoma is additionally designated as adamantinoma, solid ameloblastoma or multicystic ameloblastoma whereas cystic ameloblastoma is comprised of conventional ameloblastoma with macro-cystic alterations [1,2].

Unicystic ameloblastoma exhibits microscopic variants as luminal, intraluminal or mural. Luminal and intraluminal neoplasms are minimally aggressive subtypes which are amenable to conservative therapy [1,2].

Conventional ameloblastoma is commonly discerned within fourth to fifth decade followed in frequency by unicystic, peripheral ameloblastoma which arises within second to third decade. Extra-osseous, peripheral ameloblastoma generally emerges between fifth to seventh decade [1,2].

Ameloblastoma represents an estimated 1% of jaw cysts and neoplasms. A specific gender predilection is absent [1,2].

Intraosseous gnathic ameloblastoma, conventional or unicystic subtype is predominantly confined to mandible with few lesions occurring within maxilla. Desmoplastic ameloblastoma is predisposed to occur within anterior jaw, especially anterior maxilla [1,2].

Extra-osseous, peripheral ameloblastoma is discerned within soft tissue of posterior gingiva or retro-molar area with a predilection for lingual aspect of mandible [1,2].

Exceptionally, non-gnathic sites such as sino-nasal tract, middle ear, temporal bone or infratemporal fossa, especially in reoccurring lesions, may be implicated [1,2].

Majority (~80%) of ameloblastomas enunciate mitogen activated protein kinase (MAPK) pathway. Few lesions may exemplify Hedgehog signalling pathway [1,2].

Intraosseous gnathic ameloblastoma is engendered from remnants of enamel organ or dental lamina or may occasionally arise from an odontogenic cyst [1,2].

Extra-osseous, peripheral ameloblastoma emerges from gingival remnants of dental lamina or rests of Serres [1,2].

Conventional or unicystic ameloblastoma can be asymptomatic or may be discovered as an incidental lesion upon imaging. Clinical symptoms are generally nonspecific [1,2].

Unicystic ameloblastoma represents as a singular cystic cavity associated with or devoid of intramural tumour progression.

Gradual, painless expansion of incriminated jaw contributes to malocclusion, loose teeth, pain with infection or haemorrhage within circumscribing soft tissues. Enlarged neoplasms engender facial deformity, restricted opening of oral cavity followed by airway obstruction [1,2].

Unicystic ameloblastoma may exceptionally be associated with nevoid basal cell carcinoma or Gorlin syndrome and diverse, inherited conditions [1,2].

Tumefaction appears as a solid, multi-cystic or grossly macro-cystic, expansible, locally aggressive lesion necessitating extermination of uninvolved peripheral tissue [1,2].

Extra-osseous, peripheral ameloblastoma is an asymptomatic, commonly discerned lesion confined to soft tissue of posterior gingiva or retro-molar area. Cogent categorization of extra-osseous, peripheral ameloblastoma mandates exclusion of an intraosseous tumefaction with extra-osseous extension, simulating a gingival neoplasm [1,2].

Upon gross examination, conventional ameloblastoma appears as a solid tumefaction imbued with multiple cystic spaces. Resorption of tooth roots, impacted tooth or tumour expansion beyond incriminated bone is observed. Inferior alveolar nerve may about the neoplasm [1,2].

Cut surface depicts foci of macro-cystic degeneration with enlarged, cystic spaces exuding viscous, mucoid, clear or red-brown fluid. Unicystic ameloblastoma is pervaded with red-brown fluid [1,2].

Upon cytological examination, compact, cohesive cellular clusters with focal, peripheral, miniature branches of tumour cells are observed. Nests of elliptical to spherical basaloid cells with enhanced nuclear/cytoplasmic ratio, finely dispersed nuclear chromatin and nuclei with miniature, peripheral or inconspicuous nucleoli are enunciated. Centric aggregates of squamous cells imbued with abundant, eosinophilic cytoplasm may occur [1,2].

Upon microscopy, islands, cords or cystic odontogenic epithelial cell layer of conventional or unicystic ameloblastoma exhibits columnar cells incorporated with basal, hyperchromatic nuclei with sub-nuclear vacuoles, peripheral palisading and reverse polarization, directed away from basement membrane. Supra-basal cells exemplify a loose, network-like configuration simulating stellate reticulum articulated within normal odontogenesis. Dentin or enamel is absent [1,2].

Conventional ameloblastoma displays macro-cystic alterations within islands of tumour cells. Fragmented tissue sampling of conventional ameloblastoma with macro-cystic degeneration exhibits peripheral palisading of hyperchromatic basal epithelial cell layer, reverse polarization and stellate reticulum-like areas [1,2].

Inferior alveolar nerve is infrequently incriminated [1,2].

Conventional ameloblastoma exhibits diverse histopathological patterns devoid of prognostic significance. A singular pattern may predominate or an admixture of various configurations is observed within a solitary lesion [1,2]:

- Follicular configuration is common and exemplifies islands of odontogenic epithelium embedded within fibrous connective tissue. Lesion may be cystic or demonstrates classic peripheral palisading with stellate reticulum-like zones [1,2].
- Plexiform ameloblastoma exhibits cords and sheets of anastomosing odontogenic epithelial cells wherein classic peripheral palisading or reverse polarization of epithelium may be indiscernible [1,2].
- Acanthomatous variant enunciates focal squamous metaplasia and variable keratinization of stellate reticulum-like cells [1,2].
- Granular cell ameloblastoma displays stellate reticulum-like cells or peripheral cells of tumour aggregates imbued with granular, eosinophilic cytoplasm [1,2].
- Basal cell or basaloid variant is infrequent and comprised of islands of hyperchromatic, basal cells along with an absence of stellate reticulum-like areas [1,2].
- Desmoplastic ameloblastoma is constituted of compressed, angular islands of epithelial tumour cells commingled with dense, moderately cellular fibrous connective tissue or collagenous stroma. Focal metaplastic bone trabeculae may be discerned [1,2].

Unicystic ameloblastoma enunciates variable histological patterns denominated as:

- Singular cystic lesion layered by ameloblastic, columnar epithelium with basal cells imbued with vacuolated cytoplasm, hyperchromatic nuclei, reverse polarization and peripheral palisading [1,2].
- Supra-basal cells are non-cohesive and loosely configured simulating stellate reticulum. Besides, epithelial invagination, epithelial oedema and cellular separation may ensue [1,2].

Unicystic ameloblastoma exemplifies diverse variants as:

- Luminal variant which depicts cystic odontogenic epithelium with characteristic histological features surrounded by a layer of fibrous connective tissue [1,2].
- Intraluminal variant demonstrates typical cystic odontogenic epithelium with layered fibrous connective tissue and tumour extension into cystic luminal spaces. Intraluminal plexiform configuration may concur [1,2].
- Mural variant describes classic cystic odontogenic epithelium with circumscribing layer of fibrous connective tissue and definitive tumour islands embedded within encompassing fibrous connective tissue [1,2].

Upon ultrastructural examination, epithelial differentiation of tumour cells is configured by tonofilaments or complex desmosomes. Lysosomal aggregates within cytoplasm of granular cells and apoptotic cellular fragments may concur with neoplastic cellular clusters [3,4].

Ameloblastoma exhibits repetitive somatic and activating mutations within mitogen activated protein kinase (MAPK) pathway. Genomic mutations are discerned in BRAF V600E, RAS or FGFR2 genes [3,4].

Ameloblastoma is immune reactive to CK14, CK19, CK5, CD56, calretinin, BRAF V600E, p63, p40 or FOXP1 and infrequently reactive to β-catenin [3,4].

Tumefaction is immune non-reactive to SOX10 [3,4].

Ameloblastoma requires segregation from neoplasms such as odontogenic fibroma, ameloblastic fibroma, adenomatoid odontogenic tumour, squamous odontogenic tumour, dentinogenic ghost cell tumour, sclerosing odontogenic carcinoma, clear cell odontogenic carcinoma or ameloblastic carcinoma [3,4].

Unicystic ameloblastoma necessitates demarcation from dentigerous cyst, odontogenic keratocyst or calcifying odontogenic cyst [3,4].

Ameloblastoma can be appropriately discerned with concurrence of pertinent clinical, radiographic and histopathological features [3,4].

Incriminated subjects depict hypercalcemia [3,4].

Upon radiography, conventional ameloblastoma demonstrates an expansible, multi-locular, well defined, radiolucent lesion with corticated margin. A classic, soap-bubble appearance may occur [3,4].

Desmoplastic ameloblastoma configures reactive bone [3,4].

Unicystic ameloblastoma may articulate a unicystic countenance upon radiography. Tumefaction exhibits a unilocular, well defined, radiolucent lesion with corticated perimeter. Cortical perforation may ensue [3,4].

Features such as impacted tooth, resorption or displacement of tooth roots may be discerned [3,4].

An expansive, peripheral, intraosseous ameloblastoma delineates 'cupping of bone' and lesions with subsequent perforation of cortical bone can simulate a peripheral gingival lesion [3,4].

Cross sectional imaging is optimal in detecting miniature intraosseous ameloblastomas with prominent extra-osseous component [3,4].

Conventional ameloblastoma of mandible can be subjected to marginal or segmental surgical resection along with removal of ~ one centimetre bone perimeter and singular, adjacent, uninvolved, anatomic tissue barrier. Maxillary neoplasms are excised en bloc [3,4].

BRAF inhibitors can be unsuccessfully employed for treating instances with non surgical therapy [3,4].

Unicystic ameloblastoma can be subjected to enucleation or surgical resection [3,4].

Extra-osseous, peripheral ameloblastoma is amenable to conservative treatment with surgical eradication. Proportionate tumour reoccurrence is minimal [3,4].

Cogent treatment of mural unicystic ameloblastoma or metastasizing ameloblastoma remains debatable [3,4].

Simple curettage of conventional ameloblastoma is associated with enhanced tumour reoccurrence. Marginal or segmental eradication of mandibular lesions with removal of one centimetre of uninvolved tissue decimates proportionate reoccurrence [3,4].

Maxillary lesions or tumefaction devoid of BRAF V600E genetic mutations display elevated proportionate tumour reoccurrence [3,4].

Reoccurrence of unicystic ameloblastoma is contingent to histological pattern or extent of tumour proliferation and is ~10% [3,4].

Extra-osseous, peripheral ameloblastoma may display reoccurrence with persistence of intraosseous component [3,4]. Extended monitoring is recommended on account of possible delayed representation of disease reoccurrence which appears minimally responsive to therapy [3,4].

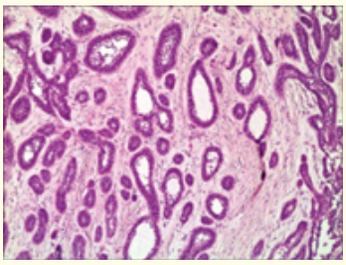

Figure 1: *Ameloblastoma with cysts and glands layered with columnar epithelium with hyperchromatic nuclei and reverse polarization and circumscribing fibrotic stroma [5].*

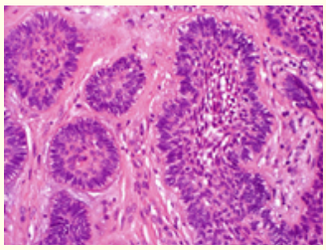

Figure 2: *Peripheral ameloblastoma depicting glandular articulations layered with columnar epithelium with hyperchromatic nuclei and reverse polarization enveloped by fibrotic stroma [6].*

Tumour	Node	Metastasis
TX: Primary tumour cannot be assessed	NX: Lymph nodes cannot be assessed	
Tis: Carcinoma *in situ*		
T0: No evidence of primary tumour	N0: Lymph node deposits absent	M0: Distant metastasis absent
T1: Tumour ≤ 2 cm	N1: Metastasis in single, ipsilateral node ≤ 3 cm	M1: Distant metastasis present
T2: Tumour between 2 cm to 4 cm	•N2a: Metastasis in single, ipsilateral node ≤ 6 cm. •N2b: Metastasis in multiple, ipsilateral nodes ≤ 6 cm •N2c: Metastasis in bilateral or contralateral nodes ≤ 6 cm	
T3: Tumour > 4 cm	N3: Metastasis in a lymph node > 6 cm	
•T4a: Tumour invades cortical bone, extrinsic lingual muscles genioglossus, hyoglossus, palatoglossus, styloglossus, maxillary sinus, facial skin •T4b: Tumour invades masticator space, pterygoid plates, skull base or encases internal carotid artery		

Table: *TNM classification of carcinoma of oral cavity [1,2].*

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5. Image 1 Courtesy: srmjrds.in.

6. Image 2 Courtesy: Springer link.

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