



ODONTOGENIC TUMORS -A REVIEW.

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**ABSTRACT**

Odontogenic tumors are the heterogenous group of lesions differs from benign lesions to malignancy due to its diversity in clinical behaviour and histological types. It arises from the tooth forming tissues which are unique to jaws and to the dentistry. Odontogenic tumors are capable of inductive interactions between odontogenic ectomesenchyme and epithelium, and the classification of odontogenic tumors is based on this interaction. Odontogenic tumours presents with a large number of histologic patterns and are derived from the primordial tooth forming tissue They occur most commonly in mandible and maxilla. According to the classification of 2017, benign odontogenic tumors are classified into three and many tissues variety into two based upon its emergence into the oral cavity. The tooth-forming tissues can give rise to a wide variety of tumours, both benign and malignant. Benign growths are usually noncancerous, but they can be aggressive and expand, displace or destroy the surrounding bone, tissue and teeth. Malignant odontogenic tumours arise either de novo from the tooth forming tissues or developmental residues or from existing odontogenic epithelial or mesenchymal neoplasms in the jaws. Their management includes extensive surgery due to their infiltrative nature and risk of metastasis. The aim of this review article is to acquire knowledge on the types and it's clinical diversity thereby it will be easy to understand it's course and to make an efficient treatment plan.

**KEYWORDS :** Ameloblastoma, Ameloblastic fibroma, Ameloblastic fibrosarcoma, Benign, Calcifying epithelial odontogenic tumor, Complex and Compound odontoma, Cemento myxoma, Malignant, Odontogenic carcinoma, Odontogenic myxoma.

**INTRODUCTION**

Odontogenic tumors comprises of disorders of growth from malignant and benign neoplasms to malformations of dental tissues of self limited growth<sup>1</sup>. Odontogenic tumors derived from ectomesenchymal and epithelial odontogenic tissues and manifest following normal tooth development<sup>2</sup>. Odontogenic tumors represent interactions between odontogenic ectomesenchyme and epithelium like normal odontogenesis<sup>4</sup>.

There are two primary classification for odontogenic tumors which includes benign odontogenic tumors that arise denovo and malignant odontogenic tumors that arise from benign precursor<sup>2</sup>. Odontogenic tumors are principally jaw lesions but some present as localized gingival swellings which is known as peripheral odontogenic tumors<sup>1</sup>.

The etiology of odontogenic tumors is not exactly determined but the result of next generation sequencing demonstrated specific mutation improved the biological process in tumorigenesis of odontogenic tumors. They involve cell proliferation, cell differentiation, regulation of tooth development, apoptotic factors and extra cellular matrix remodelling.

**Classification**

Table 1 shows list of odontogenic tumors based on the WHO classification.

<b>BENIGN</b>	<b>Odontogenic epithelial tumors</b> Ameloblastoma Squamous odontogenic tumor Calcifying epithelial odontogenic tumor Adenomatoid odontogenic tumor
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	Keratocystic odontogenic tumor (odontogenic keratocyst) Odontogenic epithelial/ Ectomesenchymal Ameloblastic fibroma Ameloblastic fibrodentinoma Ameloblastic fibro-odontoma Complex and compound odontomas Odontoameloblastoma Calcifying cystic odontogenic tumor (calcifying odontogenic cyst) Dentinogenic ghost cell tumor Odontogenic ectomesenchymal Odontogenic fibroma Odontogenic myxoma Cementoblastoma
<b>MALIGNANT</b>	<b>Odontogenic carcinomas</b> Metastasizing (malignant) ameloblastoma Ameloblastic carcinoma Primary intraosseous squamous cell carcinoma Clear cell odontogenic carcinoma Ghost cell odontogenic carcinoma Odontogenic sarcomas Ameloblastic fibrosarcoma

**Ameloblastoma**

Ameloblastoma is a benign tumor of odontogenic epithelium that majorly affects jaws. It is mainly of enamel organ-type tissue which does not undergone any differentiation to the point of hard tissue formation. It arises from the dental lamina epithelium. Ameloblastoma is characterized by local aggressive behavior and it has high recurrence rate. It

involves 1% of all oral tumors and 9-11% of all odontogenic tumors. It is a slow-growing, locally invasive tumor. Its peak incidence occurs in the third to fourth decades of life<sup>5</sup>. The female : male ratio is 1:1. Ameloblastoma accounts for about 60.3% of all odontogenic tumors in Indian population, with average age of 30.2 years.

Ameloblastoma is classified into solid/multicystic, extrasosseous/ peripheral, desmoplastic ameloblastoma, unicystic. The etiological factors are trauma, inflammation, nutritional deficiencies, non-specific irritation from extractions, and dental caries. Ameloblastoma occurs also due to the enamel organ, remnants of odontogenic epithelium, and lining of odontogenic cyst<sup>6</sup>. Most of cases of ameloblastoma (80%) occur in the mandible, mostly in the posterior mandibular region. Maxillary ameloblastoma mostly occurs in the posterior molar region. Maxillary ameloblastoma shows SMO mutations and RAS mutations<sup>9</sup>. Occurrence of ameloblastoma can also be associated with unerupted third molar tooth<sup>7</sup>. The growth occurs in buccolingual direction which results in significant expansion. The average size of ameloblastomas is approximately 4 cm<sup>8</sup>. Pain will occur when hemorrhage present inside or adjacent to the tumor. The other signs and symptoms are soft tissue invasion, loosening of teeth, facial deformity, malocclusion<sup>7</sup>.

Ameloblastoma can be diagnosed by imaging (usually a CT scan) as well as a biopsy. The CT scan shows a well-defined, uni- or multilocular radiolucent expansile lesion. It is good for the evaluation of cortical destruction or soft tissue extension margin.

Differential diagnosis of ameloblastomas includes calcifying epithelial odontogenic (CEOT), central giant cell granuloma, or ameloblastic fibroma, odontogenic myxoma<sup>10</sup>. Histologically, ameloblastoma contains two types of cells: peripherally situated known as 'basal cells' that resemble ameloblasts and centrally situated suprabasal cells known as 'epithelial cells' which resemble stellate reticulum. Presence of hyperchromatic basal cells which is columnar in shape with a palisaded arrangement, vacuolated cytoplasm and reversal of polarity will be present (nuclei displaced away from the basement membrane)<sup>1</sup>. In classical ameloblastoma these basal and epithelial cells are arranged in two characteristic patterns: follicular and plexiform patterns. In the follicular pattern, arrangement of epithelial cells will be in islands or follicles surrounded by connective tissue. In the plexiform pattern, arrangement of epithelial cells will be in an interlacing plexiform network surrounding the connective tissue. Various other histological variants of multicystic ameloblastomas are desmoplastic, granular cell, acanthomatous, basal cell, and keratopapillary ameloblastomas<sup>1</sup>.

The treatment modality for ameloblastoma is surgical treatment. Other treatment modalities like chemotherapy or radiotherapy have role only in select situations. The goal of surgical treatment of ameloblastomas is to restore good function and to minimize recurrence rate and aesthetics with minimum morbidity in the donor area<sup>7</sup>. The currently suggested surgery for classical ameloblastoma is partial or total maxillectomy for the maxilla and complete en bloc resection (radical surgery) which is classified as marginal or segmental osteotomy for the mandible.

#### Calcifying epithelial odontogenic tumor (pindborg tumor):

The calcifying epithelial odontogenic tumor (CEOT) or Pindborg tumor is a rare benign epithelial odontogenic neoplasm<sup>11</sup>. It occurs most commonly in fourth - sixth decade of life and bears no gender predilection. 94% of the lesions are central and intraosseous and 6% are extrasosseous. Mandible is affected commonly than maxilla

with the ratio (2:1). It mostly arises in the premolar/molar region. 52% cases have been associated with unerupted or embedded tooth<sup>12</sup>. In general it occurs as a slow-growing painless, bony hard swelling causing expansion of the cortical plates along with subsequent soft tissue infiltration. It may cause rotation, migration, tipping, and mobility of the adjacent tooth and may also be associated with impacted/unerupted teeth. Root resorption is found in 4% of cases.

The differential diagnosis for CEOT includes adenomatoid odontogenic tumor, odontoma, calcifying odontogenic cyst, ameloblastic fibro-odontoma<sup>12</sup>.

Radiologically, CEOT exhibits as a unilocular or multilocular radiolucent lesion presented with scattered flecks of calcification throughout the radiolucency which gives "driven snow" appearance.

The histopathological features will reveal layers of odontogenic epithelial cells that will form prominent intercellular bridges. Areas of eosinophilic, extracellular and amyloid-like material with formation of concentric calcifications (Liesegang Rings) will be present<sup>11</sup>. Amyloid like substances stained positively with Congo red and produce typical apple green birefringence under polarized light in confocal microscope<sup>13</sup>.

Small intrabony lesions with well defined borders are treated by simple enucleation or curettage followed by removal of a thin layer of bone adjacent to the tumor. Large tumors are treated by segmental resection, hemimaxillectomy and hemimandibulectomy, which causes bone discontinuity requiring reconstruction procedures such as grafting. Recurrence rate following conservative treatment is 10-20%<sup>12</sup>. Malignant transformation and metastasis is rare.

#### Adenomatoid Odontogenic Tumor

Adenomatoid odontogenic tumor (AOT) is an uncommon benign odontogenic lesion which arises from odontogenic epithelium commonly associated with an impacted tooth, usually canine. It accounts 3-7% of all odontogenic tumors<sup>15</sup>. It is a benign neoplasm which occurs commonly in the second decade of life. Females are commonly affected than males. It most often occurs in the anterior maxilla<sup>16</sup>.

AOT is a painless, benign, noninvasive, and slow-growing tumor which does not infiltrate the bone. The tumor appears as an intraoral-extraoral swelling in the maxilla. It is sometimes referred as 'two-third tumor' because it occurs in the maxilla in about two-third cases, about two-third cases occurs in young females, two-third cases are associated with unerupted tooth, and two-third cases affected teeth are canines<sup>17</sup>.

Radiographically, AOT resembles dentigerous cyst. The lesion will be usually unilocular and radiolucent. However, it contains fine calcifications (snowflake) which is a key feature in differentiating an AOT from dentigerous cyst<sup>15</sup>. Presence of unilocular radiolucency which is well demarcated with smooth cortical border. Most lesions are juxta coronal, pericoronar, displacement of teeth and divergence of roots often occurs without root resorption. The differential diagnosis are dentigerous cyst, calcifying odontogenic cyst, calcifying odontogenic tumor, unicystic ameloblastoma, odontogenic keratocyst<sup>17</sup>.

Histopathological features reveals that tumor is usually surrounded by a well-developed connective tissue capsule. A solid mass with a single large cystic space, or as numerous small cystic spaces will be present. The tumor is composed of spindle-shaped or polygonal cells forming sheets and whorled masses in scant connective tissue stroma<sup>17</sup>.

Amorphous eosinophilic material will be present between the epithelial cells, as well as in the center of the rosette-like structure. Dystrophic calcification in varying amounts will be encountered. AOT can be treated by enucleation along with the associated impacted tooth and simple curettage.

Since tumor is not locally aggressive, conservative treatment is adequate. Recurrence rate is rare.

### Keratocystic Odontogenic Tumor

The World Health Organization (WHO) considered odontogenic keratocyst (OKC) to be a tumor and recommended the term keratocystic odontogenic tumor in 2005<sup>18</sup>. The metamorphosis of odontogenic keratocyst into a recognized cystic neoplasm, keratocystic odontogenic tumor (KCOT), occurred after observation of its biological behavior and association of chromosomal and genetic abnormalities consistent with neoplastic progression<sup>20</sup>.

Keratocystic odontogenic tumor involves approximately 11 % of all cysts in the maxillofacial region and is located most commonly in the ramus and angle region of the mandible<sup>19</sup>.

The etiology of KCOT is the epithelial islands derived from the remnants of dental lamina, mostly found in the gingiva and periodontal ligament. Various theories for the rise of KCOT include intraluminal hyperosmolality, active epithelial proliferation and collagenolytic activity of the cyst wall and synthesis of interleukin 1 and 6 by keratinocytes<sup>19</sup>.

It occurs as solitary lesions or as multiple cysts or a component of the basal cell nevus syndrome. Mandible is affected commonly when compared to maxilla. In mandible, ramus-third molar area, followed by first and second molar and then anterior region are affected commonly. In maxilla, third molar area followed by cuspid area is commonly affected. The clinical features are pain, soft tissue swelling and expansion of bone, drainage and various neurologic manifestations, such as paresthesia of the lip or teeth<sup>20</sup>.

Radiographically KCOT may be present as radiolucent entity with well defined borders extending along the cancellous bone. It can either be a unilocular or multilocular radiolucency<sup>19</sup>. Smaller KCOTs appear as asymptomatic unilocular entity with corticated borders. Histopathologically, KCOT consists of epithelial lining composed of a parakeratinized surface which is typically corrugated, rippled or wrinkled. A prominent palisaded, polarized basal cell layer of cells will be present which is often described as having 'picket fence' or 'tomb-stone' appearance<sup>20</sup>.

KCOT can be managed by conservative treatment and radical management. Goal of conservative treatment is preserving the bony architecture as much as possible while removing the pathology as in marsupialisation or decompression. Aggressive forms of management are enucleation with/without curettage along with uses of chemical or cryo cauterisation or resection<sup>19</sup>.

### Ameloblastic Fibroma

Ameloblastic fibroma occurs rarely. It comprises of neoplastic epithelial and mesenchymal tissues. This lesion was previously considered to be a benign lesion with very limited recurrence rate which undergo malignant transformation<sup>14</sup>. Ameloblastic fibroma (AF) is an extremely rare mixed benign tumor which occurs either in the mandible or maxilla<sup>14</sup>. It is commonly found in the posterior region of the mandible, often associated with an unerupted tooth<sup>14</sup>. The tumors are considered as a tumor of childhood and adolescence and occur almost exclusively in the first and second decades of life with a slight female predilection, causing delay in tooth eruption or altering the eruption sequence<sup>14</sup>.

Patients often present with painless swelling of the jaw and the lesion may affect the normal eruption of teeth in that area. Buchner et al. proposed that there are two different types of ameloblastic fibromas - one of neoplastic nature and the other representing a hamartomatous lesion<sup>21</sup>. Small tumors are asymptomatic, and larger ones produce significant swelling of the jaws<sup>14</sup>. On radiographs, smaller lesions are well circumscribed and unilocular with a sclerotic border,<sup>14</sup> occasionally multilocular when larger, with smooth well-demarcated borders. Cortical expansion may or may not be identified on plane film. Grossly, ameloblastic fibroma appears as firm, lobular soft tissue mass with a smooth surface. Histopathologically, ameloblastic fibroma consists of odontogenic epithelium in the dental papilla without dental hard tissue formation<sup>14</sup>.

Microscopically, hematoxylin and eosin sections show islands and strands of epithelial cells in a loose connective tissue stroma which resembles primitive dental papilla. The peripheral epithelial cells lining the islands and strands with low columnar cells, similar to the cells found in ameloblastoma. The connective tissue resembles cellular fibroblastic tissue similar to the dental papilla in the developing tooth. Hyaline-like tissues are also seen adjacent to the epithelial strands and islands<sup>14</sup>. The treatment of choice for ameloblastic fibroma is curettage and enucleation.

### Complex And Compound Odontomas

Odontomas are benign tumors composed of dental tissue. They are the most common odontogenic tumors of the jaws, characterized by their slow growth and nonaggressive behavior. The term odontoma was coined by Broca in 1866<sup>22</sup>. They usually develop from epithelial and mesenchymal components of the dental apparatus, producing enamel and dentin<sup>23</sup>. The etiology may be local trauma or infections<sup>22</sup>. They develop from epithelial and mesenchymal components of the dental apparatus, producing enamel and dentin. They occur at any age, but are most common in the first two decades of life, with an average age of 14–18 years. They occur most commonly in females. They are more common in the maxilla, especially the anterior maxilla, than in the mandible<sup>22</sup>. The odontomas which occur in the anterior region of the maxilla are compound, while the great majority of odontomas which occur in the posterior areas, are of the complex types<sup>23</sup>.

The etiology of odontomas have been associated with trauma during primary dentition as well as with inflammatory and infectious processes, hereditary anomalies (Gardner syndrome, Hermann's syndrome), odontoblastic hyperactivity and alterations in the genetic components<sup>22</sup>. Odontomas are generally small, they may occasionally grow large, resulting in bone expansion. The World Health Organization (WHO) 2005 has classified odontomas into two groups based upon their gross and radiographic features into compound (small tooth-like structures), complex (conglomeration of dentin, enamel and cementum) and cystic<sup>22</sup>. Clinically, odontomas are classified as intraosseous (central), peripheral (soft tissue or extraosseous), and erupted odontomas<sup>23</sup>.

Complex odontomas are made of a mass consisting of an assembly of mineralized tissue (enamel, dentin, and cementum) and dental pulp. The compound odontomas are consisting of a set of small rudimentary teeth, assembling in clusters<sup>24</sup>. Compound odontomas most commonly found in the anterior maxilla, and appear as a collection of small teeth. Histologically, compound odontoma have the tooth-like structures which are arranged in a uniform manner similar to the normal tooth structure<sup>22</sup>. The compound composite odontome most commonly occur in the incisor cuspid region of the upper jaw<sup>23</sup>.

The structures in complex odontomas are found to be mixed

and disorganized<sup>22</sup>. The complex odontomas are commonly found in molar and premolar region of the mandible. Complex odontomas appear as a radiodense mass of hard tissues<sup>22</sup>. Histologically, the odontoma is not diagnostic. It is composed of dentin, cementum, pulpal tissue and enamel, mature enamel is lost during the decalcification process and will not be seen on conventional haematoxylin and eosin stained slides. The compound odontoma appear as recapitulates of the organization of a normal tooth, while the complex odontoma tend to appear as a disorganized mass of hard odontogenic tissue.

Loose, myxoid connective tissue with odontogenic epithelial cell rests may also be seen in close association with the lesion, and most often represents a normal dental follicular tissue. Fibrous connective tissue with a cystic lining representing a dentigerous cyst may also be seen<sup>22</sup>. Radiographic examination seems to be the most effective clinical method of differentiating between the two types. Compound odontoma, shows comparatively a well-organized malformed teeth or tooth like structures, which are radiolucent cyst like lesion where as complex odontoma shows an irregularly shaped oval radiopacity usually surrounded by a well-defined thin radiolucent rim<sup>23</sup>. The treatment of choice for both complex and compound odontoma include surgical removal.

#### Odontoameloblastoma

Odontoameloblastoma (OA) is an extremely rare neoplasm, which is defined as follows: "A neoplasm which includes odontogenic ectomesenchyme in addition to odontogenic epithelium that resembles an ameloblastoma (SMA) in both structure and behavior.

Odontoameloblastoma is a mixed odontogenic tumor with both epithelial and mesenchymal components<sup>25</sup>. Odontogenic Ameloblastoma develops from proliferating odontogenic epithelium and mesenchymal tissue<sup>25</sup>. It is characterized by simultaneous occurrence of ameloblastoma and composite odontoma<sup>26</sup>. It contains an ameloblastomatous component and odontoma-like elements, usually seen to occur in the mandible of younger patients<sup>27</sup>.

Odontogenic Ameloblastoma is a locally invasive, aggressive odontogenic tumor, which spreads by infiltration between the bony trabeculae.

It might cause divergence and resorption of the roots, there might be dull or intermittent pain<sup>27</sup>. Several other names exist for this tumor which include odontoblastoma, adamantodontoma, calcified mixed odontogenic tumor, soft and calcified odontoma, and ameloblastic odontoma<sup>25</sup>.

It usually occurs between 6 months to 40 years, involving mandible with molar-premolar region more common<sup>26</sup>.

There is a male predilection. This tumor usually occurs in the posterior segment of both the jaw<sup>25</sup>. It is an expansile centrally destructive lesion exhibiting slow growing characteristics like ameloblastoma and if left untreated. Symptoms include a slowly progressive swelling of the alveolar plates, dull pain, an altered occlusion, delayed eruption or impacted teeth<sup>26</sup>. Radiological examination usually reveals a unilocular or multilocular radiolucency with radiopaque areas resembling mature dental tissue. It commonly exhibits a well-defined margin, the surrounding erupted teeth rather than producing root resorption<sup>25</sup>.

Histopathology: odontogenic epithelial cells arranged in the form of follicles and stellate reticulum like cells in the center which are surrounded by ectomesenchymal cells, tall columnar ameloblast like cells on the periphery with stellate reticulum like cells at the center and surrounded by condensed mesenchymal cells. Neoplastic odontogenic

epithelium is in the form of large follicles. Tall columnar ameloblasts like cells which shows nuclear palisading, reversal of polarity and stellate reticulum like cells; dense ectomesenchymal cells present in the connective tissue stroma; large masses of dysplastic dentin arranged in a haphazard pattern; and irregular masses of dysplastic dentin, areas of calcification and stromal connective tissue<sup>25</sup>. The treatment methods are en-bloc resection or complete resection of affected part of bone irrespective of size of the lesion. Early and periodic follow-up is also advised to detect any possible relapse<sup>26</sup>.

#### Odontogenic Myxoma

Odontogenic myxoma is considered as a rare, benign, locally aggressive and nonmetastasizing neoplasm. It commonly arises from the odontogenic ectomesenchyme<sup>28</sup>. Odontogenic myxoma, also termed as odontogenic fibromyxoma or myxofibroma, is a subtype of myxoma occurring mainly in the hard, bony tissues of the face<sup>30</sup>. Odontogenic myxoma is a rare benign tumour of the jaw and characteristically appears as a slow, painless, expansion with facial deformity. It is most commonly associated with an unerupted tooth and arises from the mesenchymal portion of the tooth germ<sup>28</sup>.

It usually occurs in the second to the third decade of life, although some authors have reported the age of occurrence as 15–65 years yet a rare case have been reported in pediatric age group in a 17-month old baby. There is no sex predilection, but there is slight female predilection and male to female ratio is 1:1.5. The tumour may occur in any area of the jaws. The mandible is more commonly affected than maxilla (3:1)<sup>28</sup>.

The odontogenic myxoma mostly contains hypocellular matrix and mucoid ground substance of glycosaminoglycan's and chondroitin sulphate, these components accounts for its rapid growth and recurrence if inadequately removed. The cause of recurrence is due to the local invasion into cancellous bone beyond radiographically visible margins<sup>28</sup>.

Histopathologically, the lesion tend to consists of loosely arranged spindle, stellate-shaped or round cells, in an abundant myxoid stroma<sup>29</sup>. On H and E stained section reveals loosely arranged stellate-shaped cells with intermingled fibrillar processes in a homogenous mucoid ground substance with a few collagen fibrils and capillaries. A very few scattered odontogenic epithelial rests, areas of hemorrhage, and foci of bony trabeculae were visible<sup>29</sup>.

Radiographically, it may produce several patterns unicystic, multilocular, pericoronal (less often) and also as radiolucent-radiopaque (rare). In case of occurrence in peri-coronally impacted tooth, it is most likely to have a unilocular outline. Sometimes they appear in patterns such as a soap bubble, honey comb, or tennis-racket pattern<sup>28</sup>. Radiographically, the multilocular appear radiolucent, with well-developed locules, consisting of fine trabeculae, arranged at right angles. It is known as the 'Tennis-racquet' or 'Step-ladder' pattern. A 'sun-ray' or 'sun-burst' appearance has also been reported. On gross examination, the surgical specimen is characteristically loose, slippery or gelatinous in nature<sup>29</sup>.

Small myxomas are generally treated by curettage, but careful follow up is necessary for at least 5 years and larger lesions need extensive resection.

Typical treatment of choice for myxoma consists of surgical resection<sup>28</sup>. Liquid nitrogen cryosurgery tend to limit recurrence because of its ability to devitalize the organic content while leaving the inorganic framework intact<sup>28</sup>.

There is no proper treatment protocol to be followed. In the present case of a growing patient conservative surgery with strict clinical follow-up was recommended because the lesion



has a significant chance of recurrence. Wider surgical excision should be employed in the case of recurrence<sup>30</sup>.

### Cementoblastoma

The benign cementoblastoma is a rare odontogenic neoplasm of the jaw is known as the only true neoplasm of cementum origin<sup>31</sup>. It was first described by Dewey in 1927. Cementoblastoma was first recognized by Norberg in 1930.

Histologically, it is defined as "a neoplasm characterized by formation of sheets of cementum like tissue containing a large number of reversal lines with lack of mineralization at the periphery of the mass or in the active growth area"<sup>32</sup>.

Recently the cementoblastoma is included into the classification under odontogenic ectomesenchyme<sup>31</sup>. The lesion derives from mesenchymal tissue, its aetiology remains unknown<sup>31</sup>. The lesion is slow growing tumor and presents as asymptomatic<sup>31</sup>. This tumor affects adults with a mean age of 20.7 years. The gender male found to be higher for males with a ratio of 2.1:1<sup>32</sup>. The common site of this lesion was found to be the mandible compared to maxilla with high incidence associated with roots of mandibular molar followed by mandibular premolar<sup>32</sup>. This tumor is primarily associated with multiple teeth, impacted molars and deciduous teeth where, The associated tooth is vital unless involved<sup>32</sup>. Histologically, the tumor presents cementum-like tissue with numerous reversal lines<sup>32</sup>. The prominent basophilic reversal lines of the lesion gives a pagetoid appearance<sup>32</sup>. The periphery may show a band of connective tissue which resembles the capsule<sup>32</sup>.

The differential diagnosis of cementoblastoma may be osteoblastoma and osteosarcoma<sup>33</sup>. Osteoblastoma and cementoblastoma are essentially identical histologically and the only distinguishing feature of fusion with the involved tooth in the latter<sup>33</sup>.

Differentiation, of the lesions, from cementoblastoma requires correlation with the clinical and radiographic findings<sup>33</sup>. The treatment choice for cementoblastoma is the complete surgical excision of the mass along with the affected tooth<sup>33</sup>. Recurrence is common and recurrence risk appears to be highest for those treated with curettage alone<sup>33</sup>.

## MALIGNANT ODONTOGENIC TUMORS

### A. Odontogenic Carcinoma

#### a. Metastasizing Ameloblastoma

Metastasizing ameloblastoma (MA) is defined as a pathological entity, despite of its histological appearance<sup>34</sup>. Ameloblastoma is defined as ameloblastic carcinoma with cytological atypia and absence of metastasis<sup>34</sup>. Therefore, MA is defined as a retrospective diagnosis that can only be made when metastasis occurs<sup>34</sup>. The primary and secondary lesions shows histological features of benign ameloblastoma, to rule out a diagnosis of METAM<sup>35</sup>. Notably, the diagnosis can only be made in retrospect and there are no specific histological features that can predict the metastasis<sup>35</sup>. Therefore, the incidence of combining metastasizing ameloblastoma and ameloblastic carcinoma under the umbrella called 'malignant ameloblastoma'<sup>35</sup>. The mean age of the lesion was found to be 42.72 ± 15.68 years (range 8–74 years), depicting the change in the trend over the past decade<sup>35</sup>. Concordant with literature, a slight male predominance was noted in the present review. Mandibular ameloblastoma predominantly shows more tendency to metastasize by the present review<sup>35</sup>. The most common proposed theories for metastasis are lung aspiration theory, surgical implantation theory and the phenomenon of heterotopias<sup>35</sup>. The lymphatogenous spread of tumors cells is accepted but hematogenous route is more favored<sup>35</sup>. The histological features of pulmonary MA have been insufficiently discussed in the literature. Henderson et al.

noted that a metastasizing tumor in the lung was more cellular than the primary tumor<sup>36</sup>. Their histological pictures showed many glandular structures. The histological features of pulmonary MA have been insufficiently discussed in the literature.

Henderson et al. noted that a metastasizing tumor in the lung was more cellular than the primary tumor<sup>36</sup>. Their histological pictures showed many glandular structures. The recurrent mandibular tumor will be a solid/multicystic ameloblastoma with a follicular growth pattern. The nests consisted of stellate reticulum along with peripheral palisading basal cells and local squamous metaplasia separated by fibrous stroma. The pulmonary metastatic tumor of patient 2 was more cellular, with many glandular/papillary structures that did not appear in the primary tumor. Among these structures, nests of spindle/ovoid cells with peripheral palisading basal cells and local squamous metaplasia that resembled acanthomatous ameloblastoma were observed<sup>35</sup>.

Previous studies have reported a 5 years survival of 44%<sup>35</sup>. Five year survival of 70% has been stated elsewhere in the literature<sup>35</sup>. Surgery alone appears to be the optimal treatment of both primary and secondary sites. A long term follow-up is mandatory<sup>35</sup>.

### B. Odontogenic Sarcoma:

#### b. Ameloblastic Fibrosarcoma:

Ameloblastic fibrosarcoma (AFS) is a rare odontogenic neoplasm which exhibits benign ameloblastomatous epithelial component admixed with sarcomatous mesenchyme. It is considered the malignant counterpart of ameloblastic fibroma (AF) and can arise de novo or from transformed AF. AFS is a locally aggressive tumor that seldom metastasizes. According to the World Health Organization classification, AFS is considered an odontogenic sarcoma<sup>37</sup>. AFS a rare malignant neoplasm, consist of : i) A reductive benign odontogenic epithelium arranged in an island and net pattern, dispersing in the mesenchymal tissue; ii) malignant hyper cellular mesenchymal tissue comprised of spindle and stellate cells, exhibiting nuclear pleomorphism with hyperchromatism<sup>38</sup>. Panoramic radiography exhibits a macroscopical which is effective and low-cost image for the lesions involving the bones. Furthermore, CT scans provides more detailed information about the lesion and their association with the adjacent organs<sup>38</sup>. AFS shows recurrence potential even after the conservative treatment. Compared with the initial lesions, the recurrent lesions shows decreased epithelial components with increased pleomorphic and mitotic mesenchymal components (38). AFS is usually local invasive rather than regional or distant metastasis<sup>38</sup>.

Procedures like curettage, nucleation and local excision have typically been utilized in the surgical procedure<sup>38</sup>. Since the proliferative potential and malignant degree is elevated in the recurrent neoplasm, it eventually results in sarcomatous transformation<sup>38</sup>. The inadequate surgical boundary may induce the recurrence of the tumor. Therefore, local control will have its importance in the extent of the initial resection. Conservative surgery is replaced by wide surgical resection surrounding the soft tissue, particularly the cortical plates in cases of perforation. Adjuvant postoperative radiotherapy and chemotherapy may be effective. A number of cases involving cyclophosphamide and fluorouracil, showed positive result<sup>38</sup>. The administration of radiotherapy (40–60 Gy) following surgery can be employed for effective results<sup>38</sup>. Therefore, AFS is difficult to discriminate from AF when relying only on the histopathological features. Resection with a wide margin is the optimal treatment strategy, and adjuvant chemotherapy and radiotherapy may additionally be used to reduce the recurrence rate and enhance the survival rate<sup>38</sup>.

## DISCUSSION

Odontogenic tumors (OT) are generally a heterogeneous group of lesions that differentiates itself in clinical behaviour and histopathologic types, differs from hamartomatous lesions to malignancy<sup>39</sup>. OTs may derive from ectomesenchyme, epithelial tissues or even both around the tooth-forming apparatus<sup>39</sup>. Though odontogenic tumors are not frequent in jaw bones, it should be included in the differential diagnosis of bone lesions so as to overcome the suspicions related odontogenic tumors in first line<sup>40</sup>. Many researchers published several works pertaining to the age, gender predilection, etiology of the tumors of odontogenic origin based on classification of odontogenic classification proposed by WHO in 1992<sup>40</sup>.

The WHO published its several editions of Histological types of odontogenic tumors where its first edition was published in 1971<sup>41</sup>. But the researchers are facing several controversies related to the classification, etiology, terminologies and several other diagnosis related to odontogenic tumor all because of its diversities and mutations<sup>41</sup>. Few studies are reported among Asians, especially from the Indian subcontinent illustrating its relative frequencies of different parts of the world covering different ethnic groups in and around Asian and European countries<sup>41</sup>. The ameloblastoma being the second most common pathology is the most common histological type in Africa, followed by the odontogenic myxoma<sup>40</sup>.

The countries like Chile and Canada, have reported 20 and 18% of such tumors; in turn, the most common tumor in these countries is the odontoma (Chile – 45 and Canada – 46%)<sup>41</sup>. The tumors are comparatively rare comprising of 1% of all oral and maxillofacial pathologies which ranges from hamartomatous or non neoplastic tissue proliferations to malignant neoplasms with metastatic capacity<sup>41</sup>. Therefore, knowledge of various types of OTs along with their clinical characteristics can be supportive for both pathologists and general practitioners when developing a differential diagnosis and may lead to the arrival of conclusion concerning these lesions<sup>41</sup>.

## CONCLUSION

Odontogenic tumors are rare group of tumors which comprises about 1% of all head and neck pathologies. It shows slight predominance in women and commonly occur during the first decades of life. It is evident that most of the tumors occur in mandible and most of them are asymptomatic. By knowing these tumors in detail, we will be able to rule out the differential diagnosis, thereby helpful to conclude its diagnosis and treatment plan.

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