Introduction:

Ameloblastoma is the most frequently encountered neoplasm derived from residual odontogenic epithelial components characterized by a benign but locally invasive behavior with a high tendency to recur. This tumor is most commonly seen in molar ramus area of the mandibular jaw bone of individuals in 3rd to 4th decade of life. The unicystic ameloblastoma, a variant of ameloblastoma, has been reported to occur in decade earlier than its solid counterpart. There are less than 10% cases in the literature where unicystic ameloblastoma has been reported in very young children. Here is a case report of one such rarity where plexiform unicystic ameloblastoma showing mural proliferation, in an otherwise healthy four year old girl involving anterior region of the mandible.

Case report:

A four year old girl child presented with a complaint of a slow growing swelling of the lower jaw, front teeth region since the past eight months. Extra oral examination revealed mild, diffuse swelling on the right lower anterior portion of the mandible (Fig.1), which, on palpation was firm and non tender. Intra oral examination revealed a swelling extending from 82 to 74. Expansion of both buccal and lingual cortical plates was evident (Fig.2). All laboratory findings were found to be within normal limits. Orthopantomography of the lesional area revealed a well defined unilocular radiolucency extending from 82 to 75 (Fig.3). The radiograph also showed displacement of the developing permanent teeth up to the inferior border of the mandibular jaw bone. Computed tomography showed an expansile osteolytic lesion (Fig.4). Incisional biopsy of the lesion was performed and microscopic examination
revealed odontogenic epithelial cells proliferating in broad anastomosing cords showing presence of peripheral tall columnar ameloblast like cells with reverse nuclear polarity and central stellate reticulum like cells. There was also a focal area of ameloblastomatous cystic lining. Mature fibrous connective tissue was seen between these epithelial cords. A diagnosis of plexiform unicystic ameloblastoma was established. The tumor was surgically resected along with the overlying teeth. Gross examination of the specimen revealed a well circumscribed tumor mass measuring about 4cm X 5cm (Fig.5), firm in consistency the cut surface of which showed a cystic sac, focal thickening of the cyst wall and luminal nodules were also detected. Microscopic examination of excisional specimen revealed a focal area of ameloblastomatous cystic lining and extensively invading strands of odontogenic epithelium forming anastomosing network, suggesting plexiform pattern into the surrounding mature connective tissue (Fig.6 and Fig.7). A diagnosis of plexiform unicystic ameloblastoma grade III was reached. Post treatment response of the patient is good.

**Discussion:**

Ameloblastoma is a benign odontogenic tumor arising either from rests of dental lamina, from developing enamel organ, from the epithelial lining of an odontogenic cyst or from basal cells of the oral ectoderm. Its relative frequency equals the combined frequency of all other odontogenic tumors, excluding odontomas. Ameloblastomas are slow growing and locally invasive tumors, occurring in three different clinico-radiographic situations namely, Conventional solid/multicystic, Unicystic and Peripheral.

Unicystic Ameloblastoma is defined as a single cystic cavity that shows ameloblastomatous differentiation in the lining. Solid masses of tumor cells may also extend into the lumen, or islands of tumor may infiltrate the fibrous wall. However to be categorized as unicystic, the ameloblastoma tissue must be confined to the cyst and not extend into the wall.

Ackerman et al, in their study of unicystic ameloblastomas, defined three subgroups. Group I (42%) consisted of a unilocular cyst with a nondescript but variable epithelial lining. Inactive odontogenic cell rests might be present in the fibrous wall, but there was no infiltration by neoplastic epithelium. Group II lesions (9%) featured intraluminal plexiform proliferation but no infiltration of the cyst wall. In Group III lesions (49%), plexiform or follicular — type ameloblastoma, sometimes in continuity with the cyst lining, infiltrated the wall. Group III lesions need to be treated more aggressively similar to solid/multi cystic Ameloblastoma.

The present case shows presence of plexiform type ameloblastoma in continuity with the cyst lining infiltrating the wall and hence was diagnosed as group III plexiform unicystic ameloblastoma.

Ameloblastomas present over a wide age range but in industrialized countries they are usually diagnosed in the fourth and fifth decades of life. In developing countries, ameloblastomas tend to present about 10-15 years earlier.

Several extensive surveys and reviews of ameloblastoma have been published, the most notable being those of Robinson in 1937 and Small and Waldron in 1955. Analysis of more than 1,000 cases by the latter authors revealed that the ameloblastoma most commonly occurs in the 20 to 49 years age, with average age at first diagnosis being about 39 years.

A review of 1,036 ameloblastomas in which the average age is 38.9 years with only 2.2% under 10 years of age and 8.7% between 10 and 19 years of age. Similar were the results in a study by RA Ord et al 2002 and a study by Harvey P Kassler. An observation of these studies, suggests that only about 10% of cases are reported to arise in children, and less than one third of those occur in children younger than 10 years.

From various studies and published material we understand that Ameloblastoma is uncommon in children and that 80 to 90 % of the times ameloblastoma occurs in 3rd to 5th decade of life and only 10% of the times it is reported in children of
around 10 years or lesser age. Various literature also have shown that about 80% of tumors occur in mandible, of which, 70% arise in the molar region and ascending ramus, 20% in the premolar region, except for desmoplastic ameloblastoma which occurs most commonly in anterior region of maxilla.  

Ackerman et al, in a study of 57 unicystic ameloblastomas, concluded that the gender and site distribution corresponded with that of solid and multicystic ameloblastomas, but the mean age at diagnosis of 23.8 years was significantly younger. Shteyer et al found that unicystic ameloblastoma affected younger age group, the average age being 10.8 years. Gardener and Corio in their analysis of 46 plexiform unicystic ameloblastomas also concluded that they were more frequent in patients less than 30 years old. Oliveiro et al, Wassermann et al, Qureshi et al have reported cases each of unicystic ameloblastoma in patients with ages 5 years and 10 years respectively.

The present case report, of a four year old girl with unicystic ameloblastoma with extensive plexiform mural proliferation, occurring in mandibular anterior region, is rare and unique in its age of occurrence and site. This case adds to the very few cases reported in literature, where plexiform unicystic ameloblastoma with extensive mural proliferation histopathologically and reaching a size of 4cm X5cm as evidenced clinically and radiographically in just a 4 year old child.

Conclusion:

Ameloblastomas remain an enigmatic group of oral tumors. This rare case of plexiform unicystic ameloblastoma in four year old girl in anterior region of mandible adds to the pediatric cases in literature. More such cases should provide us an insight to the biologic behavior and clinical course of this tumor which may help in effective treatment plan.

References:

13. Qureshi SS, Medhi SS, Kane SV. Unicystic Ameloblastoma of the mandible masquerading as carcinoma of the oral cavity in a 10-year-old girl.

Fig.1. Swelling on the lower jaw front region
Plexiform Unicystic Ameloblastoma in a four year Old Child

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Fig. 2: Buccal and lingual cortical expansion seen intra orally

Fig. 3: Orthopantomography showing well defined unilocular radiolucency with displacement of the developing Permanent teeth

Fig. 4: Computed tomography showing an expansile osteolytic lesion

Fig. 5. Swelling on the lower jaw front region

Fig. 6: Low power photomicrograph showing unicystic odontogenic epithelial lining along with an area of transition in to a more proliferative epithelium.

Fig. 7: Photomicrograph showing Mural proliferation with anastomosing network of odontogenic epithelium within connective tissue stroma.