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Extraosseous Adenoid Ameloblastoma in Gardner Syndrome: an unusual presentation for a new entity

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CLINICAL HISTORY

- A 74 years old male presented with an expanding mass of the floor of the mouth
- He is affected by Gardner Syndrome, with demonstrated germinal mutation of APC gene
- He had been previously diagnosed by multiple colo-rectal adenomatous polyps and mixed AOT+CEOT of the left hemi-mandible in 2011

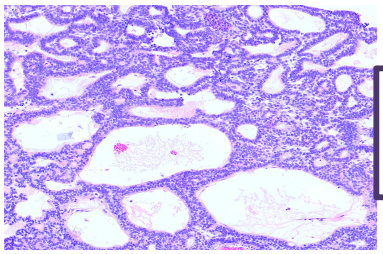
HISTOLOGY

- The intra-oral lesion was poorly circumscribed, with expansile margins and infiltrated the submucosal soft tissues of the floor of the mouth
- The tumor was composed by lobules of tumor cells, presenting a cribriform architecture and duct-like structures
- Ameloblastoma-like components, including nests and strands of tumor cells in a myxoid matrix, were present
- Moreover, squamous morules and dentinoid deposition were detectable

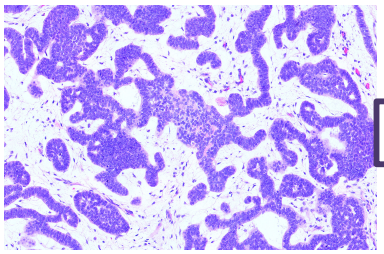
IMMUNOHISTOCHEMISTRY

- **CK34betaE12 +**
- **CK7 +**
- **p40 +**
- **Ki67 >20%**
- **Beta-Catenin + (diffuse nuclear positivity, more evident in squamous morules)**

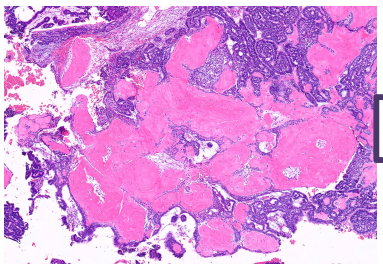
FIGURES



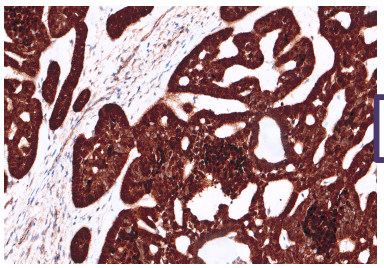
Cribriform architecture and duct-like structures



Ameloblastoma-like components



Dentinoid deposition



Nuclear beta-catenin

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CONCLUSIONS

On the basis of the clinical, histological and immunohistochemical features, a final diagnosis of adenoid ameloblastoma was made. Adenoid ameloblastoma is a rare entity, occasionally described over the last years and recently included as a new entity in the latest WHO classification of Head and Neck Tumors. The case presented herein is the first one in an extraosseous localization, being entirely placed in the soft tissues of the floor of the mouth. Even though acquired somatic mutations involving the beta-catenin pathway genes have been described for this entity, this is the first known case arising in a patient harboring APC germline mutation. Our findings demonstrate that adenoid ameloblastoma might be considered as part of the spectrum of neoplasms possibly occurring in Gardner syndrome, and, furthermore, the extraosseous localization may also be included as a possible primary site for this rare entity.