A Rare Case Presentation of a Midface Pleomorphic Adenoma with Transformation to Myoepithelial Carcinoma and Possible Glomus Tumor as a Second Primary

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More commonly, glomus tumors are solitary benign neoplasms arising from the glomus body, an arteriovenous anastomosis that serves in thermal regulation in the dermis, found under the nail on the digit tip or in the nail bed. Glomus tumors of the head and neck, specifically of the oral region, uncommon, asymptomatic and painless. Fu and Perzin reviewed 256 cases of non-glomus tumors of the nasal cavity, paranasal sinuses, and nasopharynx, and found only 1 glomus tumor, indicating an incidence of 0.4%. The sites most commonly affected are the head and palate and the upper lip. Based on our literature review, there have been only 22 cases of oral glomus tumor reported in the literature, none presenting as a second primary neoplastic lesion. It is believed that glomus tumors develop from proliferation of glomus perivascular cells or from smooth muscle progenitor cells located around the blood vessels. Microscopically, glomus tumors possess a characteristic appearance of glomus cells (small, round placed around vessels with a cytoplasm that stains eosinophilic) and a nesting pattern called zellballen. Rallios et al demonstrated that glomus tumors comprise of positive immunoreactivity for muscle specific actin, smooth muscle actin, and vimentin. Additionally, an IHC profile negative for CD31 and CD34 rule out vascular origin and negative for epithelial membrane antigen, cytokeratin, S-100 and chromogranin rules out a neuroendocrine origin. However, various authors including Kusama et al, Kesseli et al, and Boros et al have found variations in S-100 reactivity providing evidence of cellular instability of the tumor. Thus S100 positivity alone should not be considered an exclusion factor for a glomus tumor. Our patient’s neoplastic lesion excised from the nasal cavity, superior maxillary wall and posterior palate demonstrated positive vimentin, smooth muscle actin, and S-100 while negative for chromogranin, CD31 and CD34.

The partial differential diagnosis for glomus tumors includes myoepithelioma and pleomorphic adenoma. While reviewing the pathology slides with our chair of pathology we came to the conclusion that it is unlikely the patient had a second primary of glomus tumor and more likely that the patient was misdiagnosed with a glomus tumor. Myoepithelial tumors, both benign and malignant, can show a phenotype with clear cells features. When a clear cell myoepithelial tumor has an organized pattern with islands and cords, as in this case, it may be easily confused with a glomus/neuroendocrine tumor. Immunohistochemically myoepithelial neoplasms and glomus/neuroendocrine tumors share reactivity for S100. Smooth muscle actin, Vimentin and NSE. When dealing with clear cells tumors of salivary glands it is wise to perform a complete panel to differentiate myoepithelial cancers (and likely clinically myoepithelial tumors that will not be reactive in a glomus/neuroendocrine cell but will be positive in myoepithelial cells). This small panel (IHC panel of S100, smooth muscle actin, and cytokeratin) was positive in the present case and supported the diagnosis of a myoepithelial neoplasm, which is consistent with the possibility that specimen was already a low grade myoepithelial carcinoma. This brings us to a light important point, it is critical to perform the extended panel because it could alter the therapy and intervention that a patient may receive.

CONCLUSIONS

In conclusion, our case presents a rare presentation of a pleomorphic adenoma of the sinus and palate regions with a subsequent transformation to a myoepithelial neoplasm. Our case also presents a rare presentation of a glomus tumor despite surgical resection due to the patients refused to follow up. Additionally, the patient was likely misdiagnosed with a second primary glomus tumor in the proximal region. Our experience demonstrates the importance of the utilization of immunohistochemistry analysis in the diagnosing and treatment of rare neoplastic lesions of the head and neck.

BIBLIOGRAPHY