Sinonasal renal cell-like adenocarcinoma

Sinonasal renal cell-like adenocarcinoma (SRCLA) is a rare tumor first described by Zur et al. [1] and Hadi et al. [2].

Case report

A 35-year-old African-American male with von Hippel Lindau syndrome developed Sinonasal renal cell-like adenocarcinoma (SNRCLA). He underwent surgical resection followed by adjuvant radiation and has no recurrence one year from diagnosis. A review of the literature yielded two similar cases in the setting of VHL. In this case with associated VHL syndrome, next-generation sequencing detected MST1R mutation, a possible driver. SNRCLA is an emerging tumor associated with VHL syndrome and it is hoped that future studies shed light on the underlying biology of this unique tumor [3].

A 63-year-old man presented with repeated epistaxis, nasal obstruction and hyposmia of 2-month duration. Radiological studies revealed a mass of the left ethmoid sinus involving anterior skull base.

The patient was treated with craniofacial resection, bifrontal craniotomy combined with an endonasal endoscopic approach. Intraoperatively, a hypervascular paranasal mass invading the dura mater was removed en block. Histologically, the tumor resembled a clear cell renal cell carcinoma, with cuboidal shaped cells having clear cytoplasm. The tumor cells were positive for CK7, S100, vimentin and PAX-8 and negative for CD10 and PAX-2 by immunohistochemistry. No evidence of renal malignancy was found by radiological and clinical examinations.

Following local radiation therapy, the patient was in good health without recurrence for 15 months after the operation.

To the best of Kim et al. knowledge, this is the first reported case of SNRCLA in Korea. Because of its histological feature of clear cytoplasm, SNRCLA needs to be differentiated from clear cell renal cell carcinoma and other salivary clear cell carcinomas. The prognosis of SNRCLA is generally favorable as shown in the previously reported cases. Considering the limited number and follow-up periods of the cases, however, delayed recurrence should be kept in mind for clinicians. [4].

1)
Zur KB, Brandwein M, Wang B, et al. Primary description of a new entity, renal cell-like carcinoma of the nasal cavity: van Meegeren in the house of Vermeer. Arch Otolaryngol Head Neck Surg. 2002;128:441–7.
2)
Moh'd Hadi U, Kahwaji GJ, Mufarrij AA, et al. Low grade primary clear cell carcinoma of the sinonasal tract. Rhinology. 2002;40:44–7.
3)
Maharaj S, Seegobin K, Wakeman K, Chang S, Potts K, Williams B, Redman R. Sinonasal renal cell-like adenocarcinoma arising in von Hippel Lindau (VHL) syndrome. Oral Oncol. 2022 Jan 5;125:105705. doi: 10.1016/j.oraloncology.2021.105705. Epub ahead of print. PMID: 34998175.
4)
Kim NI, Yang JI, Kim SS, Lee JS, Lim SC, Jung S, Lee JH, Moon KS, Lee KH. Sinonasal renal cell-like adenocarcinoma, a unique variant of primary clear cell carcinoma of the head and neck: The first reported case in Korea. Medicine (Baltimore). 2017 Aug;96(31):e7711. doi:

10.1097/MD.0000000000007711. PubMed PMID: 28767609.

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