Clear Cell Carcinoma of the Nasopharynx

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Summary

Nearly all cases of clear cell carcinoma in the head and neck are a clear cell variant of mucoepidermoid carcinoma which affects the salivary glands. More rarely are cases that involve an unknown primary renal clear cell carcinoma that had metastasized to the upper airway tract. Very few cases reported in literature are a true primary clear cell carcinoma affecting the nasopharynx or oral cavity.[1]

Clear cell carcinoma affects the minor salivary glands which are sparsely located in the nasopharynx. The optimal treatment for this malignancy has not been established given its rare nature. Here we describe a case of primary nasopharyngeal clear cell carcinoma staged pT2N0MO and its management.

Case

44-year-old male with past medical history of right sided eustachian tube dysfunction status post right ear tube placement in 2020, presenting to the emergency department four times between 7/6/2022 and 9/23/2022 for recurrent epistaxis. Each presentation to the emergency department required increasing levels of intervention. On 9/23/2022 patient experienced significant epistaxis persistent despite nasal packing. He was brought to the operating room for nasal endoscopy and cauterization. Upon evaluation a large right-sided nasopharyngeal mass was visualized and biopsied followed by cauterization as need to achieve hemostasis.

Following cauterization in the operating room, a CT scan was performed which demonstrated a right sided nasopharyngeal neoplasm measuring 5.2 cm in diameter with subtle erosion of skull base. Biopsy results indicated hyalinizing clear cell carcinoma. Patient was treated with extensive resection including nasopharyngectomy, pterygopalatine fossa exenteration, and craniotomy. Reconstruction was performed utilizing a lateral forearm free flap. Tumor was classified as a pT2N0M0. Resection was followed by adjuvant chemoradiation therapy including Docetaxel, Cisplatin, and Fluorouracil.

Figures

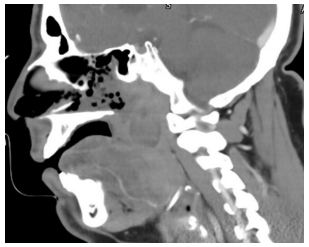

Figure 1 CT demonstrating right sided 5.2cm nasopharyngeal tumor with erosion of skull base along clivus.

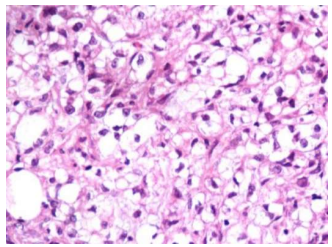

Figure 2 Microscopic findings include polygonal to round-shaped cells with clear cytoplasm and uniform nuclei with little pleomorphism.

Discussion

Minor salivary gland tumors have a much higher rate of malignancy compared to the major salivary glands. However, the concentration of minor salivary glands in the nasopharynx are extremely sparse compared to the amount found within the oral cavity and oropharynx. The Fossa of Rosenmuller contains the highest concentration of minor salivary glands in the nasopharynx.[2] The relatively small amount of minor salivary glands in nasopharyngeal region, along with the rare nature of clear cell carcinoma (CCC) involvement of minor salivary glands, accounts for the reasoning why nasopharyngeal clear cell carcinoma is rarely encountered.

There have been just over 100 case reports of clear cell carcinoma of the head and neck, with only 10 of these reported to occur within the nasopharynx. Occurrences have been most frequent in patients in their sixth or seventh decade.[3] No definitive risk factors have been identified. While renal clear cell carcinoma has a propensity for distant metastases, it has not been reported in any of the 10 case reports of nasopharyngeal CCC.[4] Presenting symptoms of nasopharyngeal CCC include eustachian tube dysfunction, epistaxis, nasal congestion, and tinnitus.[5]

Typically, CCC presents as a low-grade tumor that is easily resectable. If negative margins cannot be obtained, radiation +/- chemotherapy can be used as adjuvant therapy. When these tumors are discovered in the T1 stage, which includes 8 of the 10, prognosis is excellent with all reporting disease free state 1 year following intervention. Tumors in the T4 stage have only been reported three times prior. One patient had recurrences 4 times in the following 10 years, one patient was lost to follow up, and the third had no recurrences after 1 year.[1] No reports of a T2 tumor such as the present patient have been reported.

Histopathology of CCC Tumor cells are arranged into sheets, nests, or cords. Cytoplasmic glycogen assessed by periodic acid Schiff stain varies from marked to not evident. Tumor cells usually stain negative for mucin. Mitotic figures are rare. Tumors cells stain positive for cytokeratin and variable results are reported regarding S100 protein, glial fibrillary acidic protein, actin, and vimentin staining.[6] EWSR1 (Ewing sarcoma breakpoint region 1) rearrangement has been described as a hallmark of clear cell carcinoma of the salivary glands and may be critical in differentiating from other variants.[6]

In summary, we present this extraordinarily rare nasopharyngeal clear cell carcinoma. To date, standard treatment modality has not been established given the rarity of the malignancy. The hope is that with each case of nasopharyngeal CCC, our understanding of this tumor and its characteristics continue to progress.

References

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