Primary sinonasal renal cell-like adenocarcinoma: a case report and review of the literature

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Case Report

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Abstract

Background

Primary sinonasal renal cell-like adenocarcinoma (SNRCLA) is an extremely rare neoplasm with unique clinical and pathological features. At present, there is no summary of its clinical and pathological characteristics. We treated one case and reported to here. Review the domestic and foreign literature, summarize its clinical pathological characteristics and diagnosis and treatment.

Case

A 69-year-old female presented with repeated epistaxis, nasal obstruction of 2-years’ duration. Computed tomography (CT) was performed revealing an irregular mass of the right nasal cavity involving choana and nasopharynx. The patient was treated with endonasal endoscopic resection. Histologically, SNRCLA was very similar to clear cell renal cell carcinoma (RCC), the tumor cells were round or polygonal in size with abundant clear cytoplasm and uniform nuclei. The tumor cells positive for CK7, Vimentin, EMA, S-100, Ki-67 (5% +) and negative for CEA, P53, P63 by immunohistochemical staining,. The patient is free from recurrence over 27 months after the surgery.

Conclusion

SNRCLA seems to be a low-grade localized carcinoma associated with favorable prognosis. Surgical resection is recommended as the mainstay of treatment.

Introduction

Primary sinonasal renal cell-like adenocarcinoma (SNRCLA) is quite rare and limited number cases reported. “renal-cell like” was originally described by Zur et al[1] in 2002, who noted its resemblance to the clear-cell type of renal cell carcinoma (RCC) but with no evidence of RCC after a detailed inspection. Herein, we presented a case of primary SNRCLA and described its clinical, histopathologic, immunophenotypic, and prognostic features.

Case Report

A 69-year-old woman presented with a 2-years history of right-sided epistaxis, she complained of progressive right nasal obstruction without any symptoms such as headaches, tinnitus, diplopia, and numbness of face. Computed tomographic (CT) scan showed an irregular mass in the right nasal extending to the right choana and nasopharynx, no invasive bone destruction (Fig. 1). To exclude metastatic RCC, CT scan of abdomen and pelvis were performed, no primary renal tumor was identified. Chest CT and whole-body bone scan using single photon emission computed tomography (SPECT) were normal. The patient treated with endonasal endoscopic resection followed by adjuvant chemotherapy with paclitaxel and cisplatin. Histopathology revealed the tumor consisted of transparent cells arranged in nests and separated by fibrocellular and hyalinized fibrous septa. The nuclei were uniform and small round, with abundant clear cytoplasm, centrally or eccentrically located with inconspicuous nucleoli. Mitosis and necrosis were absent. Immunohistochemica stain revealed the tumor cells positive for cytokeratin (CK), CK5/6, CK7, CK8/18, Vimentin, EMA, S-100, Ki-67 (5% +) and negative for CEA, CgA, Syn, P40, P53, P63, CD34, HMB45, Melan-A, LCA (Fig. 2). The radiological, histological, and immunohistochemical foundings were consistent with primary SNRCLA. There was no recurrence over 27 months after aggressive treatment.

Discussion

SNRCLA is an extremely rare neoplasm, which only 19 cases have been reported in the English literature[1–19]. SNRCLA was first described as low grade adenocarcinoma of the nasal cavity and parenasal sinuses by heffner et al[2]. Later, Moh’d Hadi et al[3] and Zur et al[1] found that the tumor cell was very similar to clear cell RCC and no evidence of RCC was identified. Studies have shown that the primary clear cell carcinoma of head and neck may be originated from the salivary gland or the thyroid gland[3]. They have their unique clinical and pathologic features.

In the group of 20 patients, including our present case, have been identified SNRCLA (Table 1). The clinical presentation is no difference of other types of nasal neoplasm. The ages of the patients ranged from 22–89 years, with a median age of 62.5 years.
There was predominant in women with 13 females and 7 males. The presenting symptom was epistaxis and nasal obstruction in the majority of patients, headache, epiphora, hyposmia and tinnitus were also reported, although not specific for SNRCLA. The primary location were as follows: nasal cavity (11, 55%); Sinonasal (6, 30%), only 3 cases in nasopharynx have been reported.
| Author          | Age | Gender | Location                  | Symptoms                              | Initial diagnosis                          | Metastasis | Treatment                  | Follow up                                         |
|----------------|-----|--------|---------------------------|---------------------------------------|------------------------------------------|------------|----------------------------|---------------------------------------------------|
| Heffner (1982)² | 62  | Female | Left side of nasal cavity | Nasal obstruction and epistaxis       | Low-grade adenocarcinoma                | None       | Resection and radiotherapy | Recurred at ≥ 3 years operation, recurred at ≥ 5 years, alive with disease at 7.5 years |
| Moran (1991)⁵  | 67  | Male   | Sinonasal                 | NA                                    | Clear cell carcinoma                    | NA         | NA                         | NA                                                |
| Newman (1993)⁴ | 77  | Male   | Right side of nasal cavity| Epistaxis and nasal obstruction       | Clear cell carcinoma                    | None       | Resection and radiotherapy | Recurrence at ≥ 7 months operation                |
| Zur (2002)¹    | 50  | Female | Sinonasal                 | Epistaxis, headaches, ocular pressure and epiphora | Sinonasal renal cell-like adenocarcinoma | None       | Resection and radiotherapy | No recurrence after 8 years                      |
| Moh'd Hadi (2002)³ | 22  | Female | Olfactory cleft           | Epistaxis and nasal obstruction       | Clear cell carcinoma                    | None       | Resection                  | No recurrence after 4 years                      |
| Storck (2008)⁶ | 36  | Female | Nasal cavity              | Epistaxis                             | Sinonasal renal cell-like adenocarcinoma | None       | Resection and radiotherapy | No recurrence after 4 years                      |
| Storck (2008)⁶ | 69  | Male   | Nasopharynx               | Epistaxis                             | Sinonasal renal cell-like adenocarcinoma | None       | Radiotherapy and chemotherapy | No recurrence after 2 years                      |
| Cheng (2008)⁷  | 63  | Female | Nasopharynx               | Epistaxis, tinnitus and weight loss   | Clear cell carcinoma                    | None       | Resection and radiotherapy | No recurrence after 12 months                    |
| Negahban (2009)⁸ | 52  | Female | Left side of nasal cavity | Facial mass and nasal obstruction     | Clear cell carcinoma                    | NA         | Resection                  | NA                                                |
| Wang (2010)⁹   | 57  | Male   | Nasopharynx               | Purulence of left ear                 | Clear cell carcinoma                    | None       | Radiotherapy and chemotherapy | No recurrence after 2 months                     |
| Suzuki (2012)¹⁰ | 59  | Female | Sinonasal, skull base and intracranial | Nasal obstruction and double vision | Clear cell carcinoma                    | None       | Radiotherapy               | No recurrence after 24 months                    |
| Brandwein-Gensler (2014)¹¹ | 56  | Female | Nasal cavity, skull base  | NA                                    | Sinonasal renal cell-like adenocarcinoma | NA         | Resection and radiotherapy | No recurrence after 22 months                    |
| Davina (2015)¹² | 69  | Male   | Sinonasal                 | Epistaxis and nasal obstruction       | Clear cell carcinoma                    | None       | Resection                  | No recurrence after 10 months                    |
| Author       | Age | Gender | Location                     | Symptoms                          | Initial diagnosis                                  | Metastasis | Treatment                                   | Follow up                |
|--------------|-----|--------|------------------------------|-----------------------------------|---------------------------------------------------|------------|--------------------------------------------|--------------------------|
| Shen (2015)  | 56  | Female | Nasal cavity, skull base     | NA                                | Sinonasal renal cell-like adenocarcinoma          | None       | Resection and radiotherapy                | No recurrence after 22 months |
| Shen (2015)  | 89  | Female | Sinonasal                     | NA                                | Sinonasal renal cell-like adenocarcinoma          | None       | Resection                                  | No recurrence after 4 months     |
| Shen (2015)  | 73  | Male   | Nasal cavity                  | NA                                | Sinonasal renal cell-like adenocarcinoma          | None       | Resection and radiotherapy                | No recurrence after 20 months   |
| Imai (2016)  | 80  | Female | Right side of nasal cavity    | Nasal obstruction                 | Clear cell carcinoma                              | None       | Resection                                  | No recurrence after 12 months   |
| Kim (2017)   | 63  | Male   | Ethmoid sinus                 | Epistaxis and nasal obstruction   | Sinonasal renal cell-like adenocarcinoma          | None       | Resection and radiotherapy                | No recurrence after 15 months   |
| Wu (2018)    | 26  | Female | Olfactory cleft               | Epistaxis, nasal obstruction and hyposmia | Sinonasal renal cell-like adenocarcinoma          | None       | Resection                                  | No recurrence after 2 years     |
| Present case | 69  | Female | Nasal cavity                  | Epistaxis and nasal obstruction   | Sinonasal renal cell-like adenocarcinoma          | None       | Resection                                  | No recurrence after 27 months   |

NA = not available

Histological differential diagnosis that require consideration are hyalinizing clear cell carcinoma (HCCC), squamous cell carcinoma (SCC), metastatic renal clear cell carcinoma (RCCC), mucoepidermoid carcinoma, myoepithelial carcinoma, and epithelial-myoeipithelial carcinoma (EMC). Tumor cells of SNRCLA were round or polygonal in size, arranged in a solid nest-like manner, with abundant clear cytoplasm and uniform nuclei, centered or biased on one side, mitosis is rare, rich glycogen and lipoid, clear cell membrane, obvious boundary and slight pleomorphism. As summarized in Table 2, most of previously reported SNRCLA case were positive for CK7 (13/14), EMA (8/8), and negative for vimentin (7/13), S100 (9/15). The current case was positive for CK7, vimentin and S100. Most of SNRCLA characteristically expresses CK7, vimentin expression is variable and the Ki-67 proliferation index is usually low. In previous reports, only 9 of the 19 tumors reported to date were initially diagnosed as SNRCLA. About 9 of 19 tumors were initially diagnosed as clear cell carcinoma and 1 of 19 diagnosed a low-grade adenocarcinoma.
The clinical outcomes of SNRCLA carries a favorable prognosis. 18 patients (including ours) had follow-up examinations, the median follow-up duration was 23 months (range, 2–96 months). 15 patients were treated with primarily surgery, of which 9 patients received adjuvant radiotherapy, 2 patients received radiotherapy and chemotherapy, 1 patient received radiotherapy alone, and 1 patient received chemotherapy alone. Only 2 patients have been local recurrence, 1 patient recurrence at 3 and 5 years after surgical excision\(^2\), the other patient developed recurrence at 7 months after surgery and postoperative radiotherapy\(^4\). Neither regional lymph node involvement nor distant metastases has been documented. The most effective treatment is surgical resection, adjuvant radiotherapy may not be necessary in patients with negative margins.

In conclusion, we have reported a rare case of primary SNRCLA which is regarded as a low grade tumor. Surgical resection is recommended as the mainstay of treatment, radiotherapy and chemotherapy are still controversial. Further studies on clinical information, radiology, cytology, and immunohistochemistry is helpful for diagnosing SNRCLA.

**Abbreviations**

SNRCLA
Primary sinonasal renal cell-like adenocarcinoma
CT
Computed tomography
RCC
Renal cell carcinoma
SPECT
Single photon emission computed tomography
HCCC
hyalinizing clear cell carcinoma
SCC
Squamous cell carcinoma
RCCC
Renal clear cell carcinoma
EMC
Epithelial-myoepithelial carcinoma

**Declarations**

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**Authors' contributions**

Xiaoxia Gou and Yanzhe Wang did the literature search, study design, data analysis, interpretation of results and drafted the main manuscript. Fang Chen and Xiaoli Liu and Peigang Ruan and Xin Tian and Jinzhi Wu reviewed patients file charts and did the follow up of cases. Xiaoxia Gou and Hu Ma did critically review and approval of final manuscript. The author(s) read and approved the final manuscript.

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**Availability of data and materials**

All data generated or analyzed during this study are included in this published article.

**Consent for publication**

Informed consent was obtained from the patient.

**Competing interests**

The authors declare that they have no competing interests.

**Consent for publication**

Not applicable.

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**Figures**
Figure 1

Head CT scan. CT scan shows a soft tissue mass filling the right nasal and extends posteriorly to the choana in soft tissue window (A) and bone window (B). CT = computed tomography.
Features of tumor cells. The tumor consisted of transparent cells arranged in nests and separated by fibrocellular and hyalinized fibrous septa. Tumor cells were shown to have abundant clear cytoplasm, small round nuclei, and distinct cytoplasmic borders. (A, B, H&E stain, original magnification ×100 and ×400 respectively). Immunohistochemistry revealed strong positivity for CK7 in tumor cells. (C, D, immunohistochemistry, original magnification ×100, and ×400, respectively). H&E = hematoxylin-and-eosin.