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 main stay 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. Immunohistochemical 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 findings 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 paranasal 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.

Table 1
Reported cases of sinonasal renal cell-like adenocarcinoma

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 $\geq$ 3 years operation, recurred at $\geq$ 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 $\geq$ 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) ¹³	56	Female	Nasal cavity, skull base	NA	Sinonasal renal cell-like adenocarcinoma	None	Resection and radiotherapy	No recurrence after 22 months
Shen (2015) ¹³	89	Female	Sinonasal	NA	Sinonasal renal cell-like adenocarcinoma	None	Resection	No recurrence after 4 months
Shen (2015) ¹³	73	Male	Nasal cavity	NA	Sinonasal renal cell-like adenocarcinoma	None	Resection and radiotherapy	No recurrence after 20 months
Imai (2016) ¹⁴	80	Female	Right side of nasal cavity	Nasal obstruction	Clear cell carcinoma	None	Resection	No recurrence after 12 months
Kim(2017) ¹⁵	63	Male	Ethmoid sinus	Epistaxis and nasal obstruction	Sinonasal renal cell-like adenocarcinoma	None	Resection and radiotherapy	No recurrence after 15 months
Wu (2018) ¹⁶	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-myoepithelial 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 lipid, 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.

Table 2
Reported cases of sinonasal renal cell-like adenocarcinoma

Author	Age	Gender	CK7	CK20	CD10	EMA	CEA	Vimentin	S100	P63
Heffner (1982) ²	62	Female	NA	NA	NA	NA	NA	NA	NA	NA
Moran (1991) ⁵	67	Male	NA	NA	NA	NA	NA	NA	NA	NA
Newman (1993) ⁴	77	Male	NA	NA	NA	NA	NA	NA	NA	NA
Zur (2002) ¹	50	Female	+	-	-	+	+	-	+	NA
Moh'd Hadi (2002) ³	22	Female	+	-	-	+	+	-	+	NA
Storck (2008)[36 F] ⁶	36	Female	+	+	+	+	+	-	-	NA
Storck (2008)[69 f] ⁶	69	Male	+	-	NA	NA	NA	-	-	NA
Cheng (2008) ⁷	63	Female	NA	NA	NA	NA	NA	NA	NA	NA
negahban (2009) ⁸	52	Female	+	-	-	+	-	NA	-	NA
wang(2010) ⁹	57	Male	NA	NA	NA	NA	NA	NA	-	NA
Suzuki (2012) ¹⁰	59	Female	-	-	-	+	NA	+	+	NA
Brandwein-Gensler (2014) ¹¹	56	Female	+	+	NA	NA	NA	+	-	NA
Davina (2015) ¹²	69	Male	+	-	-	+	NA	-	-	+
Shen (2015) ¹³	56	Female	+	+	NA	NA	NA	+	+	NA
Shen (2015) ¹³	89	Female	+	-	NA	NA	NA	-	-	-
Shen (2015) ¹³	73	Male	+	-	-	NA	NA	+	+	-
imai (2016) ¹⁴	80	Female	+	NA	+	+	NA	+	-	NA
Kim(2017) ¹⁵	63	Male	+	-	-	+	+	+	+	NA
Wu (2018) ¹⁶	26	Female	+	NA	-	NA	NA	-	-	NA
Present case	69	Female	+	NA	NA	NA	-	+	+	-
NA = not available										

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.

Figure 2

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 $\times 100$ and $\times 400$ respectively). Immunohistochemistry revealed strong positivity for CK7 in tumor cells. (C, D, immunohistochemistry, original magnification $\times 100$, and $\times 400$, respectively). H&E = hematoxylin-and-eosin.