CASE REPORT

Primary Sinonasal Clear Cell Carcinoma: Case Report

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SUMMARY

Primary sinonasal clear cell carcinoma is a rare neoplasm classified under malignant epithelial tumours of salivary gland – type carcinomas under World Health Organization (WHO) classification. We report a case which occurred on a 69 year old gentleman presented with epistaxis and nasal endoscopy examination showed tumour arising from the right ethmoid cells. Endoscopic excision of the tumour was done and histopathological examination revealed clear cell carcinoma. In addition, other secondary or primary sites of the lesion were excluded by clinical, immunohistochemical and radiological examinations. 10 months into the postoperative period, patient remains well without recurrence of the tumour. In conclusion: We report a rare case of primary sinonasal clear cell carcinoma in addition to the limited literature available and emphasize the differentials with other probable tumour through meticulous microscopic examination and use of special immunostains.

KEY WORDS:
Sinoasal; clear cell carcinoma

INTRODUCTION

Primary sinonasal clear cell carcinoma is a rare neoplasm. It is classified in the World Health Organization (WHO) classification under malignant epithelial tumours of salivary gland –type carcinomas. There are only a limited number of cases reported in the primary literature with the focus on sinonasal clear cell carcinoma. Clear cell carcinoma occurs most common in the kidney which accounts for 60% of renal cell carcinoma. These carcinomas are named clear cell carcinoma due to failure of their cytoplasm to stain with hematoxylin and eosin (H & E) on light microscopy attributable to accumulation of glycogen, lipid and rarity of organelles in the cell.[1] The aim of this article is to report a rare case of primary nasal clear cell carcinoma on a gentleman who presented with epistaxis.

CASE PRESENTATION

A 69 year old Chinese gentleman presented to otorhinolaryngology clinic with two episodes of right sided epistaxis and occasional right nasal blockage for 6 months. On nasal endoscopy examination noted a mass in right nasal cavity displacing the middle turbinate laterally. Biopsy of nasal mass showed tissue covered by respiratory epithelium infiltrated by malignant cells which consist of round nuclei with distributed chromatin and vacuolated cytoplasm containing glycogen and specimen was immunochemistry positive to vimentin, Epithelial Membrane Antigen (EMA), Pan cytokeratin and S100 suggestive of metastatic clear cell carcinoma.

To identify the primary tumour, we proceeded with computer tomography (CT) scans of paranasal sinuses, abdomen and pelvis which revealed a mass arising from the right ethmoid sinus extending into the nasal cavity. Similar CT scan also revealed a small left renal cyst. However there was no evidence of renal carcinoma.

Endoscopic resection of the right nasal mass was performed via endonasal approach. Tumour appeared to arise from the ethmoid cells of the right nasal cavity. Histology of the excised tumour showed ulcerated respiratory type mucosa infiltrated by lobules of tumour cells which are round to polygonal, clear cytoplasm and central to eccentrically placed nuclei with mild pleomorphism. In addition, the cytoplasm contains glycogen as confirmed by Periodic acid Schiff (PAS) stain positivity and diastase soluble. Tumour cells are positive for Pan cytokeratin, EMA, Vimentin, Inhibin alpha, Neuron-Specific Enolase (NSE) and S100 stains though negative to CD 10, Thyroid transcription Factor-1(TTF-1) and Cytokeratin 7.

Thus a primary sinonasal clear cell carcinoma diagnosis was made. Post-surgery there was no sign of residual; neither by endoscopy, magnetic resonance imaging (MRI) nor by FDG-PET (18 fluorodeoxy glucose positron emission tomography) scan. He remains asymptomatic 10 months after surgery and has been placed on a routine monthly endoscopic surveillance.

DISCUSSION

Primary sinonasal clear cell carcinoma is an extremely rare neoplasm with epistaxis as one of the common early presentation and the diagnosis requires an exclusionary approach.[1] Clear cell carcinoma may originate in different primary tissues like kidneys, lungs, thyroid, and female genital urinary tract with each one having their unique clinical and pathologic features.[1]

Most of the clear cell carcinoma tumours microscopically have dominant cell cytoplasm not staining with H & E. Cells are round to polygonal, rich in glycogen, nuclei are round with little pleomorphism and cytoplasmic glycogen stains with PAS and is diastase soluble.[1] Mucicarmine usually stains negative in clear cell carcinoma.

This article was accepted: 15 April 2015
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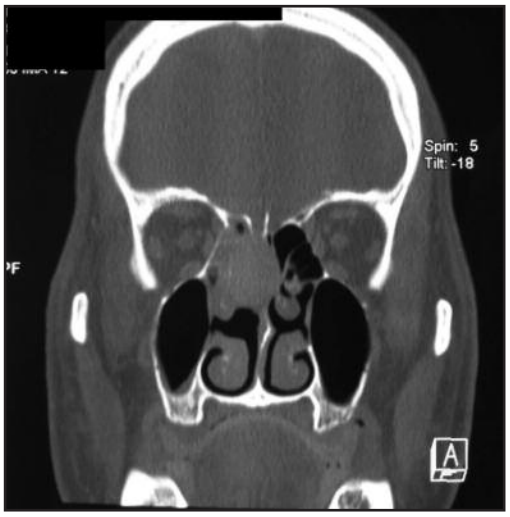

Fig. 1: Coronal CT scan of the paranasal sinus showing lesion arising from right ethmoid sinus extending into nasal cavity.

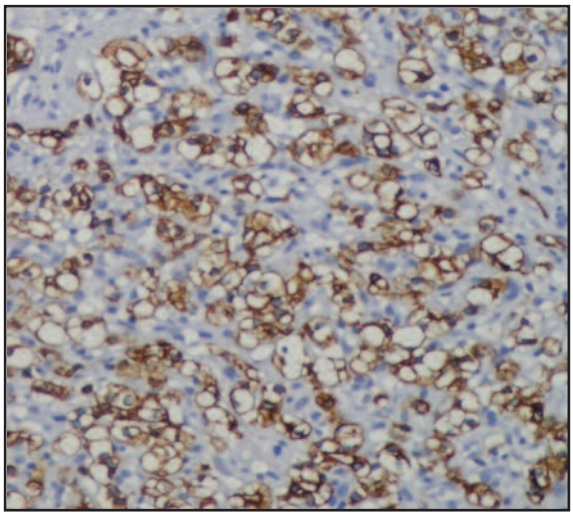

Fig. 2: Pan- CK staining tumour cells with clear cytoplasm (20x).

It is of utmost importance to distinguish this clear cell carcinoma from other tumour as different treatments are required for each tumour. Histological differential diagnoses that require consideration are metastatic renal clear cell carcinoma (RCCC), squamous cell carcinoma (SCC), mucoepidermoid carcinoma, acinic cell carcinoma and metastatic thyroid carcinoma. Nevertheless, tumours which can occur adjacent to this area should be ruminated especially olfactory neuroblastoma, hemangioblastoma as well as chondroma.

First and foremost primary sinonasal clear cell carcinoma must be differentiated from metastatic RCC as it has a remarkable resemblance. Overall, metastatic RCC comprises approximately 6% of all metastatic tumours to head and neck.[2] This can be excluded by clinical and radiological correlation. Microscopically the hallmark of renal cell carcinoma has a high degree of vascularity and significant atypia with lack of prominent hyaline stroma. In addition CD10 immunoreactivity was observed in most RCC which was negative in this case.

SCC is the most frequent type of malignant tumour in the nose and paranasal sinuses (70%–80%).[3] Clear cell change can be prominent, however the evidence of intercellular bridges, infiltrative growth pattern and keratinisation distinguish SCC from clear cell carcinoma.

Even though mucoepidermoid is one of the most common salivary malignancies, it is comparatively rare to be found in sinonasal malignancies. Histological evaluation of mucoepidermoid reveals epidermoid cells and mucin producing cells.[1] Conversely in acinic cell carcinoma the cytoplasm is devoid of glycogen.[1] Clear cell variant of thyroid carcinoma should also be taken into consideration if the immunohistochemistry of thyroglobulin and TTF1 show positive results.

In consideration of the anatomical site of the tumour, olfactory neuroblastoma was not disregarded as one of the differential diagnosis. Nevertheless, neuroblastoma is immunochemistry positive to CD99 and EMA.

There is no specific treatment protocol on primary sinonasal clear cell carcinoma most possibly due to the fact that it is a rare neoplasm. Wide excision is the treatment of choice for most clear cell carcinoma although neck dissection, radiotherapy and chemotherapy have been performed depending on the presence of positive margins, invasion of vascular or neural, grade of histology and positive neck nodes.

CONCLUSION

We have reported a rare primary sinonasal clear cell carcinoma case. It is exceptionally important to distinguish this rare tumour from other clear cell carcinomas and approximately similar cell histologically with the help of special stains, immunohistochemical testing and radiological evaluations considering the fact that different therapeutic decisions are required. The actual prevalence of this tumour is still unknown, however it is quite rare. In addition, long term follow-up and more case studies are necessary to assist in understanding the clinical behaviour and appropriate treatment for this disease.

REFERENCES

1. Moh'd Hadi U, Kahwaji GJ, Mufarrij AA, Tawil A, Noureddine B. Low grade primary clear cell carcinoma of the sinonasal tract. Rhinology. 2002; 40(1): 44-7.
2. Flocks RH, Boatman DL. Incidence of head and neck metastases from genito-urinary neoplasms. Laryngoscope. 1973; 83(9): 1527-39.
3. Mendenhall WM, Werning JW, Pfister DG: Treatment of head and neck cancer. In: DeVita VT Jr, Lawrence TS, Rosenberg SA: Cancer: Principles and Practice of Oncology. 9th ed. Philadelphia, Pa: Lippincott Williams & Wilkins, 2011, pp 729-80.