

Metastasis in neck lymph nodes as unusual first symptom in a patient with clear cell pulmonary carcinoma

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Citation

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Abstract

Introduction. The purpose of this article is to report a case of clear cell pulmonary carcinoma, an extremely rare type of lung cancer, with unusual neck lymph nodal metastasis. **Case Report.** We present the case report of a 49-year-old man who presented a 1 month history of dysphonic process with left-side neck mass. Cervical nodal biopsy was informed as metastasis of clear cell carcinoma and patient died 3 months after being attended. Clinical presentation, radiological imaging characteristics and histopathological features are presented and revised in this paper. The difficulties in establishing diagnosis are discussed. **Conclusions.** Clear cell pulmonary carcinoma is distinctly rare but aggressive lesion with unusual neck metastasis. Otolaryngologists should be aware of this possibility.

INTRODUCTION

Metastasis of visceral tumours to the extracranial region of the head and neck are uncommon and the usual primary sources of these lesions are breast, colon, and renal carcinoma¹. Lung cancer is the leading cause of cancer death in the world², with the majority of patients having advanced inoperable disease at the time of presentation. Nevertheless, clear cell carcinoma of the lung is an extremely rare type of lung cancer that comprises a diverse group of lesions. The prototypical lesion is the benign clear cell tumour or “sugar tumour,” a tumour of enigmatic histogenesis, whose name derives from the high glycogen content of the cells³. The tumour is composed predominantly of clear cells with some areas showing cells typical for small cell carcinoma. Ultrastructurally, the larger clear cells have cytoplasm containing many rough endoplasmic reticuli and free ribosomes, while the smaller dark cell showed a serrated nucleus and scanty cytoplasm having many free ribosomes without neurosecretory granules. On immunohistochemical study the tumor cells express positive with epithelial marker including cytokeratin (CK) and epithelial membrane antigen (EMA)⁴ but negative with vimentin⁵. The diagnosis should be done carefully, because benign clear cell tumour⁶ and carcinoma show similarities in histological appearance due to its morphologic

epithelioid features with clear cytoplasm, but not in their management and prognosis. Usually clinical investigations do not lead to the final diagnosis so that only subsequent histological examination and immunophenotyping can establish the correct tumour classification⁷.

The purpose of this article is to report one case of clear cell pulmonary carcinoma of a 49-year-old man with neck lymph nodal metastasis. Patient presented a 1 month history of dysphonic process with left-side neck mass and died 3 months after being attended. Clinical, radiographic and histopathological features are presented and revised in this paper.

CASE REPORT

We present a case of a 49-year-old white man that presented to the otolaryngology service with dysphonic, headache and posterior neck and thoracic pain that had been worsening over 2 months. His medical or surgical history was unremarkable and he denied previous pathology. On palpation the patient had a well-circumscribed, nonpainful, nontender, nonfluctuant, nonpulsatile, nonmobile mass at cervical nodal levels IV and V in the left-side area of the neck. The patient had no other significant otolaryngologic or physical findings although fibrolaryngoscopic examination showed a compression of left supraglottis with normal vocal

cord mobility.

The blood, hematometry and biochemical analyses and the chest X-rays were normal. Axial, contrast-enhanced Computed Tomographic (CT) of the neck revealed hypertrophy of the lymph nodes at the cervical nodal levels IV and V (Figure 1). Fine-needle aspiration did not obtain enough material for diagnosis. Given the persistence of the dysphonia, laryngeal microsurgery and a cervical nodal biopsy were performed during the same surgical session. The laryngeal microsurgery shows a slight hypertrophy of the left ventricular band and the laryngeal biopsy was normal.

The lymphatic node biopsy showed the existence of an epithelial tumor consisting of groups of cells separated by connective-vascular septa. Cytologically, they were composed of clear cells with a large clear cytoplasm, with well-defined cell borders. The size of the nuclei varied, with a moderately dense chromatin, sometimes with nucleoli and mitosis figures (Figure 2). The proliferating cells showed intense and diffuse immunoreactivity to AE1-AE3, CK7, CK19 keratins and EMA, whereas they showed no reaction to CK8, 18, 20 and 34, and to BetaE12 (Figure 3). The markers for nervous and myoepithelial differentiation, such as Vimentin, Actin and S100 were negative. The studies for HMB45, thyroglobulin and TTF-1 were also negative. The biopsy was informed as clear cell carcinoma metastasis. The tumor's immunophenotype was compatible with a possible renal or pulmonary origin.

The abdominal CT did not reveal focalized renal, adrenal and hepatic lesions or abdominal lymphatic metastasis. Contrast-enhanced CT scan of the thorax showed the existence of a mass in the left lung vertex involving the region of the aortic-pulmonary window with a site of osseous metastasis involving the upper dorsal vertebral bodies. In addition, bilateral lung metastases were found (Figure 4).

The patient received 20 sessions of radiotherapy but, despite them, patient gradually deteriorated and showed Horner syndrome and progressive dyspnea and died 2 months later before the planned chemotherapy began.

Figure 1

Figure 1: Computed Tomographic (CT) of the neck. Hypertrophy of the cervical lymph nodes is observed.

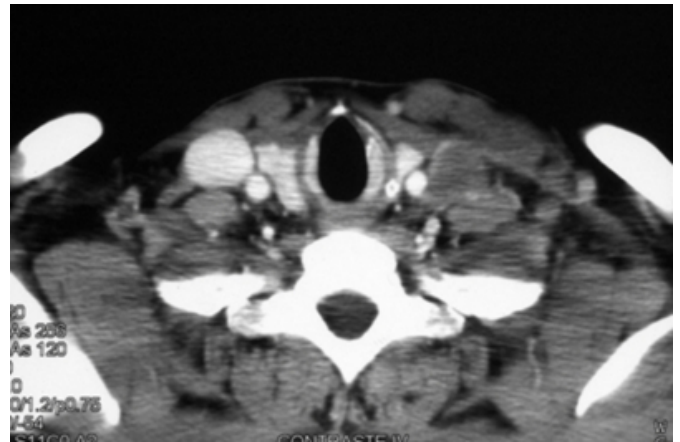


Figure 2

Figure 2: Hematoxylin and eosin; original magnification $\times 40$. Epithelial tumour consisting of groups of cells separated by connective-vascular septa is observed. Clear cells with a large clear cytoplasm with well-defined cell borders are showed.

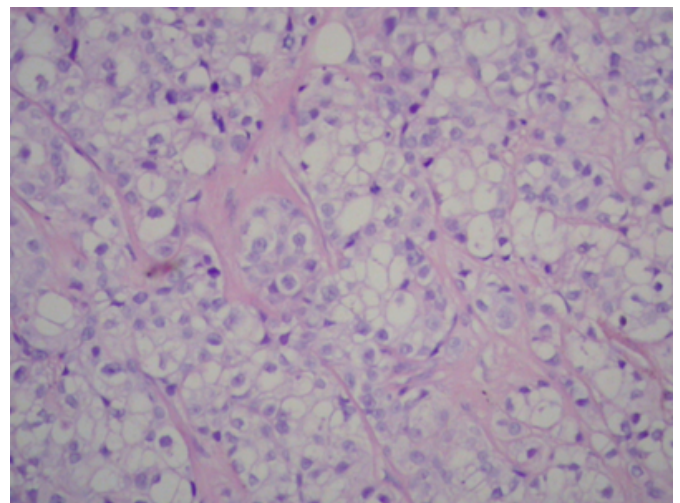


Figure 3

Figure 3: Intense and diffuse immunoreactivity to CK7 keratins; original magnification $\times 40$.

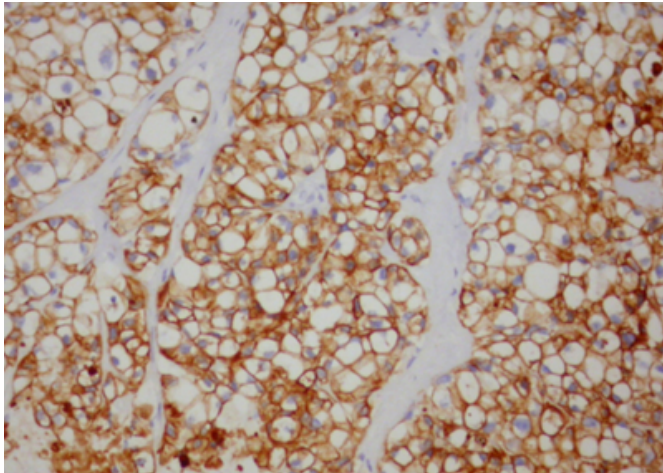
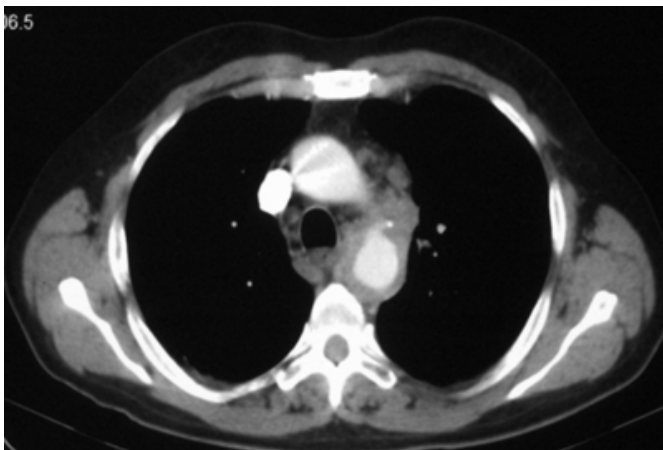


Figure 4

Figure 4: Contrast-enhanced CT scan of the thorax. Bilateral lung metastases and a mass in the left lung vertex involving the region of the aortic-pulmonary window are showed.



DISCUSSION

Clear cell pulmonary carcinoma is a type of lung cancer of low incidence. At the same time, in patients with lung cancer, metastasis is usually located in lymph nodes within the mediastinum but not in neck nodes. An exception is the more superficially located supraclavicular lymph nodes, which are of substantial importance because are associated with incurable disease⁸. Therefore, we have presented a case report of a patient with clear cell pulmonary carcinoma whose first medical symptom was the existence of a neck mass of unknown origin. The diagnosis was made with cervical and thoracic CTs and a biopsy of the cervical tumor formation during the same surgery session.

About radiographic, CT is of choice in assessing the extent of metastatic carcinoma in the head and neck. The appearance may suggest malignancy but is nondiagnostic, although the enhancement with contrast, destruction, and lack of tumour calcification should suggest metastatic clear cell carcinoma as a part of the differential diagnosis. Magnetic resonance imaging is helpful in demonstrating intracranial involvement and residual disease after radiotherapy treatment⁹. At the same time, ultrasonography (US), Fine-needle aspiration cytologic (FNAC), US-guided FNAC analysis and computed tomography (CT) are safe and cost-effective methods for diagnosing supraclavicular lung cancer metastasis and to assess the effect of proved metastasis on TNM stage and diagnostic work-up^{10,11,12,13}. However, in our case report FNAC was not enough for diagnosis, and given the persistence of the dysphonia, laryngeal microsurgery and a cervical nodal biopsy were performed during the same surgical session. So, although all imaging modalities are helpful in the workup, biopsy is the only way to confirm the diagnosis

The evolution of the case report we are presenting was rapidly lethal but some studies have reported the effectiveness of radiotherapy in the treatment of clear cell carcinoma metastatic disease. Response to radiation therapy is site specific, with bone and soft tissue metastasis having the best response rates. Chemotherapy may be used as an adjunct to therapy for postsurgical recurrences. The mainstay of chemotherapy treatment for metastatic clear cell carcinoma uses interleukin-2 or interferon-[alpha] with partial response rates ranged from 5% to 20%⁹. About surgical treatment, head and neck metastasis should be viewed differently because of the lesions can lead to severe disfigurement, airway compromise, recurrent bleeding or other associated morbidity issues. So, depending on the site of presentation, local resection may improve quality of life, provide a chance for cure in the head and neck or allow for appropriate short-term palliation of symptoms, despite the poor long-term prognosis of the disease.

CONCLUSION

Clear cell pulmonary carcinoma is distinctly rare but aggressive lesion that can be fatal and metastasis to the head and neck are rare. Otolaryngologists should be aware of this possibility. The difficulties in establishing clinical, pathological and radiological imaging basis diagnosis are discussed.

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