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Case Report

Obstructing Trachea Tumor: Myoepithelial Carcinoma about a Case

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ABSTRACT

Myoepithelial carcinoma, also called malignant myoepithelioma, is an extremely rare tumor of the salivary gland type. It is defined as a malignant neoplasm in which the tumor cells exhibit exclusively myoepithelial differentiation. Our case is a 48-year-old patient with no specific history who was presented to the hospital emergency room for respiratory difficulty and dysphagia progressively evolving for 08 weeks. A cervical CT was done which revealed a hypodense tumor process in T1 of 4 cm long axis obstructing the trachea well limited and richly vascularized. The diagnosis of tracheal myoepithelial carcinoma was made after histological and immunohistochemical analysis on a biopsy pending analysis of the operative specimen. Treatment remains initially surgical with appropriate postoperative radiotherapy and chemotherapy.

INTRODUCTION

Myoepithelial carcinoma is a rare tumor that mainly affects the salivary glands, breast, and rarely soft tissue. Tracheal myoepithelial carcinoma does not fall under the most recent classification of the World Health Organization (WHO) which defines a set of tumors called the salivary gland type of the lung of which it is not part [4]. It is an extremely rare tumor; only 16 cases reported in the literature [5,6]. Here, we report a case of tracheal myoepithelial carcinoma diagnosed as a result of asphyxiation in a 45-year-old emergency patient, whose endoscopic examination found a polypoid tumor obstructing the trachea.

CASE PRESENTATION

Our case is a 48-year-old patient with no specific history who was presented to the hospital emergency room for respiratory difficulty and dysphagia that had progressed gradually 08 weeks ago. A cervical CT was done which revealed a hypodense tumor process in T1 4 cm long axis obstructing the trachea well limited and richly vascularized [Figure 1]. An emergency bronchial endoscopy showed a polypoid tumor process occupying the entire tracheal lumen bleeding on contact [Figure 2]. A biopsy curettage was done to free the upper airway. Our laboratory received 12 biopsy fragments measuring between 0.5 and 1.5 cm, yellowish-white in color, completely included in three blocks of paraffin.

The histological study shows a bronchial mucosa massively infiltrated by a carcinomatous process with a partly cylindromatous and partly diffuse architecture made up mainly of cells with a clarified cytoplasm and an elongated and filiform nucleus with spindle-shaped cells and epithelioids [Figure 3]. Cytonuclear atypias are moderate and the mitotic index is estimated at 7 mitoses per 10 Gx40 fields and the tumor stroma is poorly represented and inflammatory with a few lymphocytes. No image of vascular embolus or peri-nervous sheath was observed. The immunohistochemical study performed on the tumor shows the positivity of all tumor cells for the myoepithelial markers: AML, P63 and cytokeratin AE1-AE3 while they are negative for CD117 [Figure 4].

A genetic study is underway to identify the abnormalities by FISH technique.

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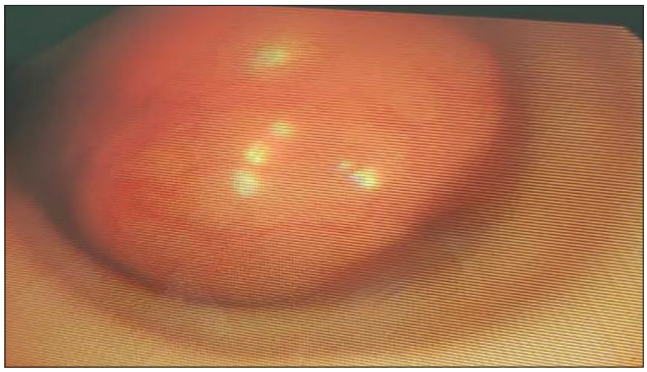

Figure 1. Endoscopic image showing the vascularized polypoid tumor obstructing the trachea

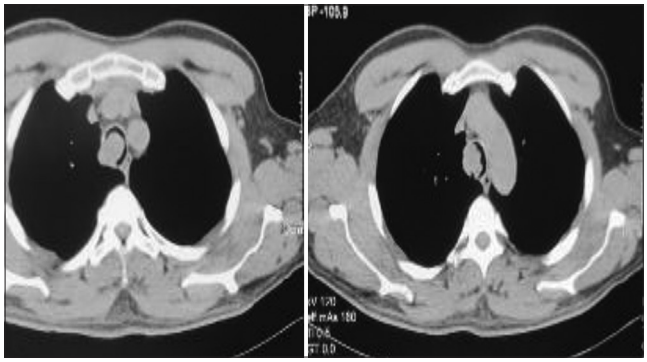

Figure 2. X-ray image of cervical CT showing the hypodense tracheal tumor process in T1

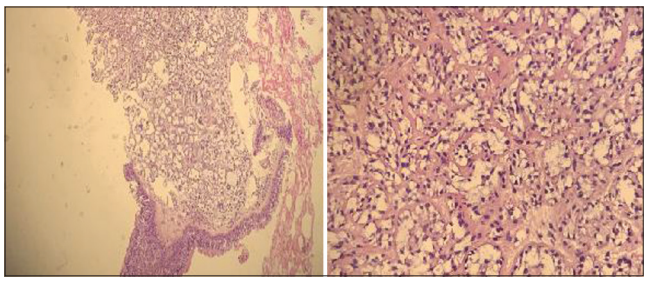

Figure 3. Histological images of a carcinomatous proliferation of diffuse and cylindromatous architecture infiltrating the trachea

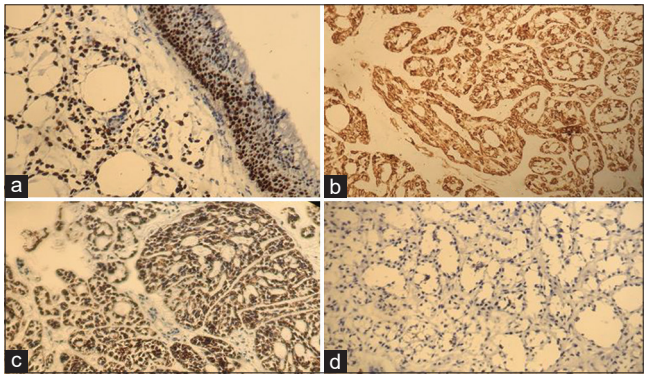

Figure 4. Immunohistochemistry: a- nuclear positivity of P63, b- membrane positivity of AML, c- membrane and cytoplasmic positivity of cytokeratin AE1-AE3, d- negativity of CD117

The diagnosis of tracheal myoepithelial carcinoma was made pending analysis of the operative specimen.

The patient will be a candidate in the coming days for surgery to remove the tumor with a healthy excision margin followed by local radiotherapy and chemotherapy sessions.

DISCUSSION

Myoepithelial carcinoma, also called malignant myoepithelioma, is an extremely rare tumor of the salivary gland type. It is defined as a malignant neoplasm in which the tumor cells exhibit exclusively myoepithelial differentiation.

Myoepithelial carcinoma is mainly localized in the salivary glands and the breast, and rarely occurs in the accessory salivary glands of the airways, such as the nasopharynx, larynx, trachea or lung. There has been one documented case in the nasopharynx [1], four cases in the larynx [2-3] three cases in the trachea, [7,8] and five cases in the lung. [9,10]

Data from the literature confirm that myoepithelial carcinoma is much more common in men. The age of the patients ranged from 23 to 76 years (mean age: 60.5 years) [11].

A wide variety of tumor cells have been described in myoepithelial carcinomas. Therefore, it is important to identify the pathological features in order to avoid a false diagnosis and obviously inadequate treatment. Myoepithelial carcinoma simply shows myoepithelial differentiation translated histologically into five subtypes: epithelioid cells, plasmacytoid cells, stellate cells, spindle cells and clear cells [11]. The co-presence of these subtypes can be described in the same tumor. In our case clear cells are predominant.

Cytonuclear features include cytologic atypia, tumor infiltration, and high mitotic index which are considered useful in distinguishing malignant myoepithelioma. The presence of tumor infiltration is the minimum criterion for making the diagnosis of malignancy [11]. Recently, it has been suggested that assessment of cell proliferative activity may be useful in differentiating between benign and malignant myoepithelioma, and that more than seven mitoses per 10 high magnification fields or a higher Ki-67 proliferation index. 10% are indicative of myoepithelial carcinoma [11]. In our case tumor infiltration is evident and the mitotic index is moderately high.

Immunohistochemically, tumor myoepithelial cells are positive for cytokeratin AE1-AE3, vimentin, and markers of myoepithelial differentiation (SMA, MSA, PS-100, Calponin, P63, CD10 or GFAP). However, neoplastic myoepithelial cells can show various reactions to these markers [11]. Our case is positive for markers of myoepithelial differentiation.

On the genetic level myoepithelial carcinoma is characterized by various abnormalities related to the predominant cell subtype with an EWSR-PBX1 fusion known for the clear cell variant and the EWSR-ZNF444 translocation known for the epithelioid cell variant [12]. In the rhabdoid variant of myoepithelial carcinoma of the soft tissue of the neck for example, FISH analysis revealed an EWSR Translocation without any other genetic fusions characteristic of EWSR, including EWSR1-NR4A3, TAF15- NR4A3, EWSR1-FLI1, EWSR1- ERG, EWSR1-WT1, EWSR1-ATF1, EWSR1-CREB1 or EWSR1-POU5F1 [13]. A single case of clear

cell myoepithelial carcinoma described in the literature in the salivary gland showing a rearrangement of the EWSR1 gene detected by FISH analysis [14]. In this case, the genetic study is ongoing.

The main differential diagnoses remain the other salivary gland type tumors which are described in the trachea and the lung as adenoid cystic carcinoma and epithelial and myoepithelial carcinoma, hence the importance of the immunohistochemical study.

CONCLUSION

In conclusion, we report a rare unusual case of tracheal myoepithelial carcinoma discovered in an emergency situation. The histological description remains important but the immunohistochemical study remains essential to diagnose this type of cancer. The treatment remains initially surgical with appropriate postoperative radiotherapy and chemotherapy.

Abreviations

WHO: World Health Organization
AML: Smooth muscle actin
CT: Computed tomography
FISH: Fluorescence in situ hybridization
GFAP: Glial fibrillar acidic protein

Ethics Approval and Consent to Participate

This work has respected all the rools of medical ethics and has been elaborated by all the authors.

Availability of Material and Data

All data is available in the military hospital Mohammed V, Rabat, Morocco.

Consent to Publish

As the main author and the names of all authors I allow you to publish this article in your review

Author's Contributions

All the authors contributed to the writing of this work.

ENDONOTES

Dear sir,

I have the honor to submitt my work intitulated OBSTRUCTING TRACHEA TUMOR: MYOEPITHELIAL CARCINOMA that remains a rare entity that the recurrent caracter requires a rigorous monitoring and acomplete surgical exeresis.

My work aims to focus on this type of tumor and the role of the pathologist in the diagnostic approach.

ACKNOWLEDGEMENTS

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