Case Report

# Primary pulmonary myoepithelial carcinoma in a child: An ambiguous entity

*Wala Ben Kridis, Nabile Toumi, Afef Khanfir, Mongia Hachicha[1], Tahia Boudawara[2], Mounir Frikha*

*Department of Oncology, Habib Bourguiba Hospital, [1]Department of Pediatrics, HediChaker Hospital, [2]Department of Histopathology, Habib Bourguiba Hospital, Sfax, Tunisia*

## ABSTRACT

Primary myoepithelial carcinoma (MC) of the lung is exceedingly rare. We report here, to the best of our knowledge, the first pediatric case having primitive pulmonary MC. The originality of our case was the disappearance of the pulmonary opacity spontaneously, without any treatment. The difficulties in our case were the diagnosis of this rare entity and its subsequent treatment. In fact, given the rarity of these tumors, recommendations regarding chemotherapy or radiation, were difficult to formulate.

**KEY WORDS:** Myoepithelial carcinoma, particularity, treatment

**Address for correspondence:** Dr. Wala Ben Kridis, Department of Oncology, Habib Bourguiba Hospital, Sfax-3029, Tunisia. E-mail: walabenkridis@yahoo.fr

## INTRODUCTION

Myoepithelial carcinoma (MC) is a rare tumor with an incidence of 0.2% of all salivary gland tumors. Most of the reported cases of MC arise in the parotid gland (48–75%), followed by minor salivary glands, and the submandibular gland.[1] Primary MC of the lung is exceedingly rare. The first case was described by Higashiyama *et al.*,[2] in 1998. Since then, only seven cases have been reported in literature. We describe here the first pediatric case, with a review of the literature.

## CASE REPORT

A mentally retarded 13-year-old girl, with a history of congenital hypothyroidism and cystic lymphangioma in the left dorsal region, operated in 2004, had consulted for cough and dyspnea in September 2010. Physical examination showed a decrease in vesicular breath sounds at the basal areas of chest bilaterally, without fever. Chest radiography showed a bilateral basal opacities [Figure 1].

Access this article online

Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/0970-2113.164168

Pulmonary computed tomography (CT) showed bilateral pleural masses measuring 6.3 cm and 7.4 cm at the right and left bases, respectively, with lymph node metastases [Figure 2].

A histopathological study of the left pleural biopsy revealed a monomorphic proliferation of round cells with clear cytoplasm and a weak, mitotic hyperchromatic oval nucleus [Figure 3]. There were some cohesive layers, separated by bands of sclerosis; the stroma was sparse with foci of necrosis. The immunohistochemistry (IHC) study was focally positive for vimentin, CD99, and PS100. The abdominal ultrasound, bone scan, and metaiodobenzylguanidine (MIBG) scintigraphy were normal. The histopathological and the IHC review at the "Institut Bergonié" in France concluded the diagnosis of myoepithelial carcinoma of the soft tissues with intermediate malignancy. IHC was positive for pancytokeratin (AE1/AE3) and PS100, and negative for EMA, CD34, desmin, and CD99. The tumor was inoperable. Monitoring was advocated. The patient was lost to follow up for one year three months. The progression was marked by the disappearance of the pulmonary opacities [Figure 1].

## DISCUSSION

Primary salivary gland-type tumors of the lung are rare. They include mucoepidermoid carcinoma, adenoid cystic carcinoma, acinic cell carcinoma, oncocytoma, epithelial–myoepithelial carcinoma, benign myoepithelioma, and mixed tumors.[3] Primary pulmonary myoepithelial carcinoma (MC)

is exceedingly rare. It arises from the submucosal bronchial glands of the lower respiratory tract. In the World Health Organization Classification, published in 2004,[4] MC was cited as being synonymous with epithelial–myoepithelial carcinoma. As MC and epithelial–myoepithelial carcinoma of the salivary gland are distinguished by the presence or absence of ductal cells, their pulmonary counterparts must also be differentiated. Furthermore, the patient's prognosis is different. In fact, pulmonary epithelial–myoepithelial tumors are low-grade lesions without recurrences or metastasis described after resection, whereas, the rate of metastasis in MC is high,[4] as seen in our case.

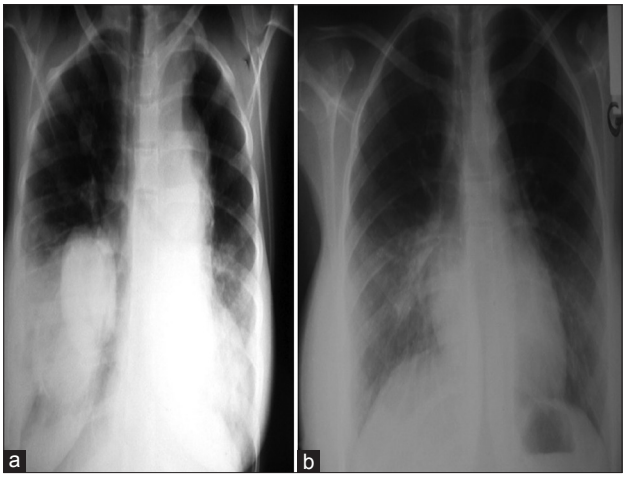


**Figure 1:** Chest x ray. (a) Bilateral pulmonary opacities (b) Disappearance of pulmonary opacities

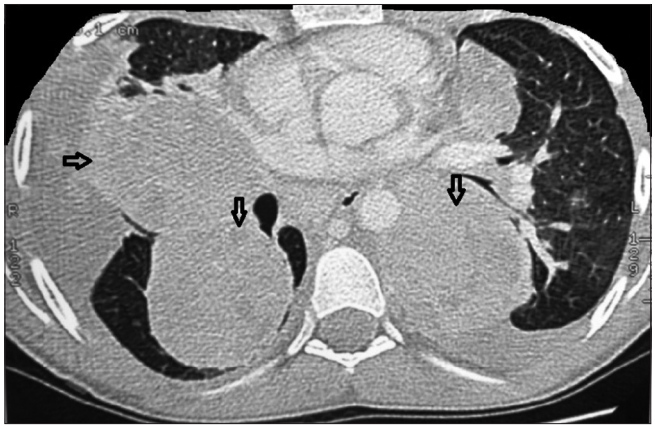

**Figure 2:** Pulmonary computed tomography (CT): Bilateral pulmonary masses

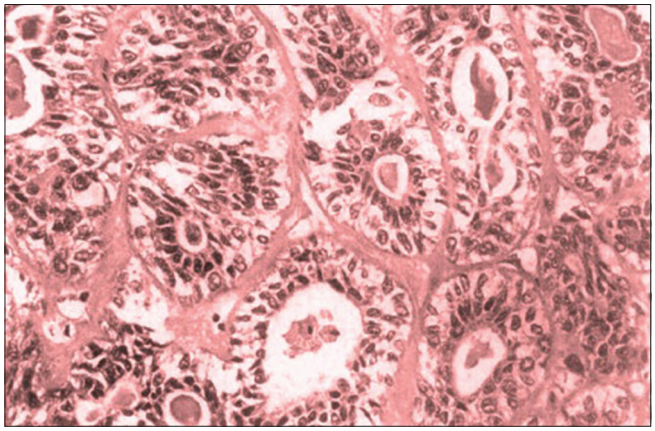

**Figure 3:** Myoepithelial cells with clear cytoplasm. (H and E, stain ×300)

Most of the patients described in the literature are Japanese men. There are only two affected women, aged 60 and 63 years. The mean age is 58.4 years (46 to 76 years). Our patient represents the first pediatric case, described in the literature, having primitive pulmonary MC. The tumor was peripheral and bilateral, measuring 6 cm and 7 cm, which was in agreement with the literature. In fact, the size of the MC ranged from 15 mm to 130 mm (mean 50.7 mm) [Table 1].[4,5]

In our case, the histopathological study oriented to the diagnosis of a primitive neuroectodermal tumor or neuroblastoma. However, an abdominal ultrasound, bone scan, and MIBG scintigraphy were negative. The pathological review, with further IHC analysis in France, had concluded the diagnosis of MC. The following markers were found to be useful in myoepithelial carcinoma: Cytokeratins (AE1/AE3) and vimentin (reported to be positive in neoplastic myoepithelial cells and negative in normal myoepithelial cells). Other variable markers, such as, PS100, Calponin, smooth muscle actin (SMA), muscle-specific actin (MSA), smooth muscle myosin, and P63 protein, could be positive.[5] However, neoplastic transformation of myoepithelial cells could result in a loss or a modification of their smooth muscle phenotype.

In our case, the tumor was inoperable with bilateral lung metastases, which caused a therapeutic challenge. In fact, surgery represented the gold standard in pulmonary MC. It was the only approved treatment in case of operable MC.[4,5] Although the eight patients reported in literature were treated with optimal surgery, metastases were reported in seven of them (87.5%) (contralateral lung, forearm, liver, and brain). It was significant that a patient who had not developed metastasis, had the lowest tumor mitotic rate of

**Table 1: Characteristics of pulmonary MC**

| Case report | Age | Sex | Location | Size | Initial Dg* |
|---|---|---|---|---|---|
| Higashiyama *et al.* | 58 | M | Proximal | 38 mm | NA |
| Higashiyama *et al.* | 58 | M | Proximal | 60 mm | SCC** |
| Sekine *et al.* | NA | NA | NA | NA | NA |
| Miura *et al.* | 46 | M | Proximal | 65 mm | Not atypical cell |
| Masuya *et al.* | 48 | M | Distal | 15 mm | Sarcoma<br>Carcinoma<br>Sarcocarcinoma |
| Tanahashi *et al.* | 76 | M | Distal | 22 mm | NA |
| Sarkaria *et al.* | 63 | F | Distal | 130 mm | Low grade<br>Spindle cell<br>Neoplasm |
| Haysi *et al.* | 60 | F | Distal | 20 mm | Schwannoma |
| Our case | 16 | F | Distal | 74 mm | PNET*** |

*Dignosis, **Squamous cell carcinoma, ***Primitive neuroectodermal tumor. M=Male, F=Female, NA=Not Available, MC=Myoepithelial carcinoma

5/10 HPFs. This fact reasoned that the mitotic rate could be an important prognostic factor of the clinical outcome and survival in primary myoepithelial carcinoma of the lung.

Given the rarity of these tumors, recommendations regarding chemotherapy or radiation, either pre- or postoperatively, are difficult to formulate. The originality of our case is the disappearance of the pulmonary opacity spontaneously, without any treatment.

## CONCLUSION

Primary MC of the lung is exceedingly rare. Our case represents, to the best of our knowledge, the first pediatric case having primitive pulmonary MC. The histopathological study familiarizes the diagnosis, but a further IHC study is needed to confirm the diagnosis and to eliminate other etiologies. Surgery represents the main treatment for the operable forms. To the best of our knowledge, we have reported the first case, with spontaneous regression of this tumor, without any treatment.

## REFERENCES

1. Savera AT, Sloman A, Huvos AG, Klimstra DS. Myoepithelial carcinoma of the salivary glands: Aclinicopathologic study of 25 patients. Am J SurgPathol2000;24:761-74.
2. Higashiyama M, Kodama K, Yokouchi H, Takami K, Kabuto T, Tsuji N, *et al*. Myoepithelioma of the lung: Report of two cases and review of the literature. Lung Cancer 1998; 20:47-56.
3. Doganay L, Bilgi S, Ozdil A, Yoruk Y, Altaner S, Kutlu K. Epithelial-myoepithelial carcinoma of the lung. A case report and review of the literature. Arch Pathol Lab Med 2003; 127:e177-80.
4. Yousem SA, Nicholson AG. Epithelial-myoepithelial carcinoma. In: Travis WD, Brambilla E, Muller-Hermelink HK, Harris CC, editors. World Health Organization Classification of Tumours: Pathology and Genetics of Tumours of the Lung, Pleura, Thymus and Heart. Lyon: IARC Press; 2004. p. 67.
5. Nguyen CV, Suster S, Moran CA. Pulmonary epithelial-myoepithelial carcinoma: A clinicopathologic and immunohistochemical study of 5 cases. Hum Pathol 2009; 40:366-73.

**How to cite this article:** Kridis WB, Toumi N, Khanfir A, Hachicha M, Boudawara T, Frikha M. Primary pulmonary myoepithelial carcinoma in a child: An ambiguous entity. Lung India 2015;32:497-9.

**Source of Support:** Nil, **Conflict of Interest:** None declared.