



Case Report

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# Primary Typical Carcinoid of the Pleura: Case Report

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To Cite This Article: Lamyae Nouiakh, Karima Oualla, Hajar Ouahbi, Soumia Berrad, Hayat Erraichi, Primary Typical Carcinoid of the Pleura: Case Report. 2020 - 10(6). AJBSR.MS.ID.001566. DOI: [10.34297/AJBSR.2020.10.001566](https://doi.org/10.34297/AJBSR.2020.10.001566).

Received: 📅 September 01, 2020; Published: 📅 November 09, 2020

## Abstract

Carcinoid tumors are rare, slow-growing neuroendocrine tumors. The most common location of these tumors is in the gastrointestinal tract and broncho-pulmonary system. Primitive pleural localization is very rare. The majority of cases found in the literature are metastatic forms or relapses. We report the case of a 56-year-old patient with a typical carcinoid tumor of the pleura.

**Keywords:** Pleura; Typical carcinoid tumor; Multidisciplinary approach

## Background

Carcinoid tumors are a very rare variety of neuroendocrine tumors, most commonly found in the digestive tract and in the lungs [1]. Primary pleural localization is rarely found. Typical carcinoid tumors pose a diagnostic problem. Because of the scarcity of these tumors, this diagnosis is sometimes difficult. Histologically, it is a neuroendocrine tumor whose morphology often joins that in other locations.

The prognosis of these tumors are generally pejorative given the late diagnosis, the absence of an effective therapeutic approach, even for localized stages where surgery remains the standard treatment as it is impossible to perform carcinological surgery due to the localization of the tumor. We report the case of a 56-year-old patient with a typical carcinoid tumor of the pleura.

## Case Presentation

A 56-year-old patient, 30 years of chronic smoking, weaned 6 years ago, who consulted for grade 2 dyspnea according to NYHA classification evolving for more than a year, becoming grade 4 one

month before your admission without any other associated signs. The clinical examination found a patient in moderate performance status (PS2) with a left pleural effusion syndrome at the pulmonary examination. The rest of the clinical examination was normal. As a result of symptomatology, the patient benefited from a standard X-ray and then a chest scan that showed a left pleural nodular thickening reaching of 18 mm in maximum thickness associated with a pleural effusion of great abundance responsible for the pulmonary collapse and discharge from the medieval elements to the right (Figure 1). A pleural puncture biopsy was performed for diagnostic and evacuating purposes. Histological analysis showed infiltration of the pleural membrane by malignant tumor proliferation consisting of epitheliomatous cell flows, cord, and trabecular structure, sometimes laying out glanduliform structures or acinous formations which are arranged in an endocrine mode in palisades and rosettes, dissociated by a desmoplastic stroma of variable abundance. The tumor cells were rounded, polyhedral, or fusiform with eosinophilic cytoplasm and ovoid nuclei, sometimes voluminous,

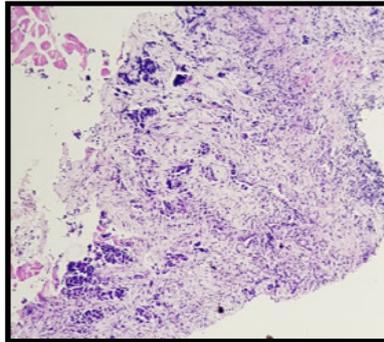


nucleoli with rare mitosis figures: less than 2M/10CFG (Figures 2 & 3). At immunohistochemistry, the tumor cells expressed antibody anti CK7, anti- SYNAPTOPHYSINE, but instead, they were negative for anti-chromogranin, anti-CD56, anti-CK20, and anti-CDX2 antibodies (Figures 4 & 5). The pathological study concluded a typical carcinoid tumor of pleural localization. The patient completed the

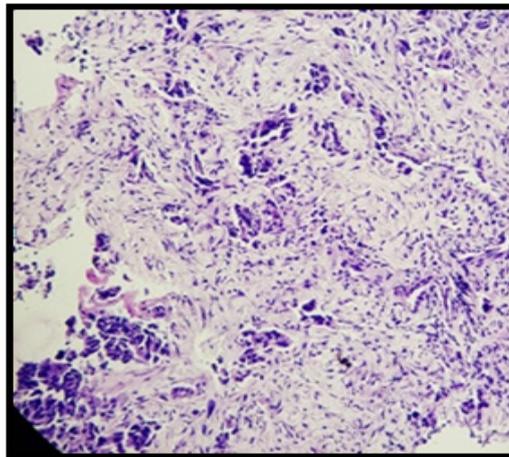
extension assessment with an abdominal-pelvic computed tomography which did not show any secondary localization. The case was discussed in a multidisciplinary consultation meeting, the tumor was found to be unresectable and the therapeutic decision was to start the first-line chemotherapy. Unfortunately, the patient died 15 days later due to respiratory failure.



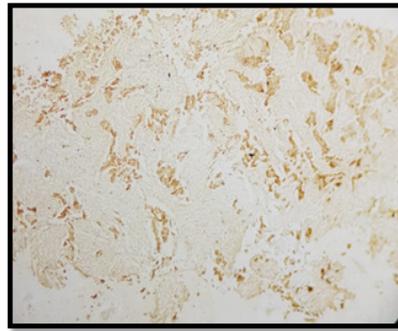
**Figure 1:** Axial scangraphic section showing a left pleural nodular thickening reaching of 18 mm in maximum thickness associated with a pleural effusion of great abundance responsible for the pulmonary collapse.



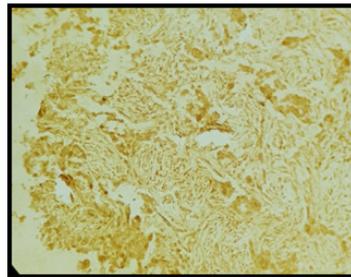
**Figure 2:** Histological study: HESx200: infiltration of the pleural membrane by malignant tumor proliferation consisting of epitheliomatous cell flows, cord and trabecular structure, sometimes laying out glanduliform structures or acinous formations which are arranged in an endocrine mode in palisades and rosettes, dissociated by a desmoplastic stroma of variable abundance.



**Figure 3:** Histological study: HESx400: The tumor cells were rounded, polyhedral or fusiform with eosinophilic cytoplasm and ovoid nuclei, sometimes voluminous, nucleoli with rare mitosis figures: less than 2M/10CFG.



**Figure 4:** Immunohistochemical study: the tumor cells expressed anticorps anti CK7.



**Figure 5:** Immunohistochemical study: the tumor cells expressed anticorps anti-SYNAPTOPHYSINE.

## Discussion

Carcinoid tumors are rare neuroendocrine tumors. The most frequent localization is at the digestive tract in 70% of cases, then at the broncho-pulmonary in 10-30% of cases [1]. In the literature, cases at the gallbladder, breast, thymus, larynx, ovary, prostate, and kidney have been described [2-6]. Primary pleural localization is unusual. Few cases have been reported in the literature [4]. The majority of reported cases were related to either metastatic forms or relapses at the pleural level of another primitive [7-9]. Only one retrospective study conducted by Moss and colleagues including 50 patients with carcinoid syndrome. In this study, pleural thickening was found in nine patients, four of whom were primary [10]. There are two types of carcinoid tumors; typical and atypical. Typical carcinoid tumors are good prognosis compared to atypical tumors and rarely metastasize.

The diagnostic of a carcinoid tumor remains the pathological examination, based on histological examination with immunohistochemical complement. In a histological analysis, the diagnosis of the neuroendocrine nature of these tumors can be suspected to morphology; tumor cells contain secretion granules attached to the membrane and to the cytoplasm level and will be confirmed by immunohistochemical analysis in case of the positivity the tumor cells by one or more endocrine markers (Synaptophysin, chromogranin A, CD56) [7].

The treatment of choice of typical carcinoid tumors is based on surgery because it is the only therapeutic modality that seems

reasonable to improve the prognosis of patients with these tumors. However, the pleural location of these tumors makes this treatment difficult as it is almost impossible to do carcinological surgery. In cases where surgical excision is not feasible and in metastatic forms, there is no consensus on the best treatment to use in the first line due to the scarcity of these tumors. However, chemotherapy seems to be beneficial in these situations to avoid the rapid progression of the disease and relieve symptoms related to the disease.

## Conclusion

Through this work, a very rare case of a typical carcinoid tumor of pleural localization has been reported. Few cases have been found in the literature. These tumors pose a diagnostic problem, hence the interest of a multidisciplinary diagnostic approach to avoid missing out on metastatic disease.

## Acknowledgement

None.

## Competing Interests

The authors declare that they have no competing interests.

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