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Pleomorphic Carcinoma of Pulmonary Location: Case Report and Review of the Literature

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Abstract **Case Report**

Pulmonary pleomorphic carcinoma is a rare primary lung tumor, part of the group of non-small cell cancers. We report the case of a 54-year-old patient, a chronic smoker who presented with hemoptysis evolving in a context of deterioration in general condition. Chest imaging revealeda left upper lobar tissue density mass with some hypodensity. The diagnosis of pleomorphic lung carcinoma was made based on histological data from bronchial biopsies, and classified as stage IVb according to the TNM classification (8th edition). Palliative chemotherapy was then offered to the patient. The evolution was marked by the death of the patient 2 months after diagnosis. Through this observation, we wish to shed light on this rare and particular histological type but just as aggressive as other pulmonary neoplasias.

Keywords: lung tumor, cell cancers, smoker.

INTRODUCTION

Pulmonary pleomorphic carcinoma is a rare malignant lung tumor belonging to the group of non-small cell cancers (NSCC), the incidence of which ranges from 0.1 to 0.3% of all lung cancers. It occurs in male patients, aged over 60, who smoke, and without specific clinical signs. Its diagnosis is based on histology which finds two components; one carcinomatous or epithelial and one sarcomatous. The prognosis remains poor due to the rapidly metastatic nature.

OBSERVATION

It was a patient of 54years old, chronic smoker at 36 PA, dyspneic on exertion for 7 years, known diabetic for 5 years on oral antidiabetics and hygiene and diet rules and hypertensive for 2 years. The respiratory symptoms dated back 3 months with the progressive worsening of his dyspnea becoming stage 2 MMRC and the appearance of episodes of hemoptysis of low to medium abundance. All evolving in a context of deterioration in general condition made up of weight loss estimated at 10 kg over a period of 3 months with anorexia and asthenia, without feverish sensations or night sweats. The clinical examination found a patient in fairly good general condition with a Performans status equal to 1, apart from obvious clubbing, the rest of the somatic examination was unremarkable. On chest imaging, standard radiography showed a homogeneous dense opacity with left interhilo-axillary projection, with spiculated boundaries without costal lysis on the side. Chest computed tomography (CT) (Figure 1) identified a left upper lobar tissue density mass with some hypodensity, probably related to areas of necrosis. The analysis of the lymph node territories, although difficult, did not find any lymphadenopathy.

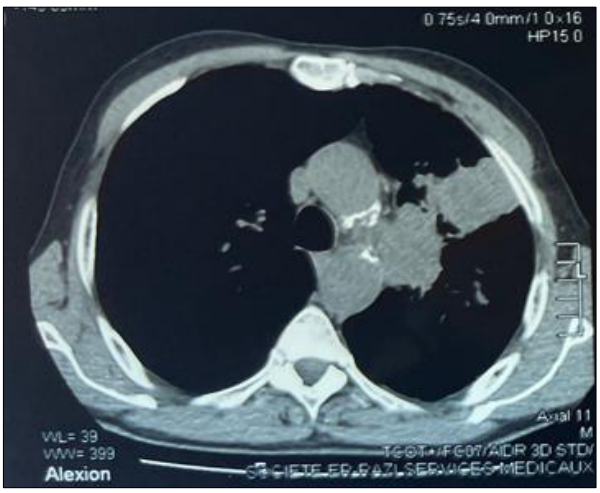

Figure 1: Mediastinal window CT scan of a left upper lobar tissue density mass

Bronchoscopic examination showed a budding tumor with a whitish surface, bleeding at the slightest

contact and completely obstructing the left upper lobar orifice. Bronchial biopsies were taken at his level. On histopathological study, the fragment of the tumor was largely necrotic, with evidence of a diffuse malignant tumor proliferation made of spindle-shaped cells (Figure 2) with elongated nuclei with the presence of several mitotic figures thus pointing towards a sarcomatous component. The immunohistochemical study showed that the tumor cells expressed cytokeratin AE1/AE3 (clone AE1/AE3) and expressed neither TTF1 (clone 8G7G3) nor p40 (clone ZR8, Bio-SB) nor WT1 nor CD20., nor CK5/6 and on the other hand they focally expressed P63 (Figure 3). These data made it possible to retain the diagnosis of pleomorphic lung carcinoma.

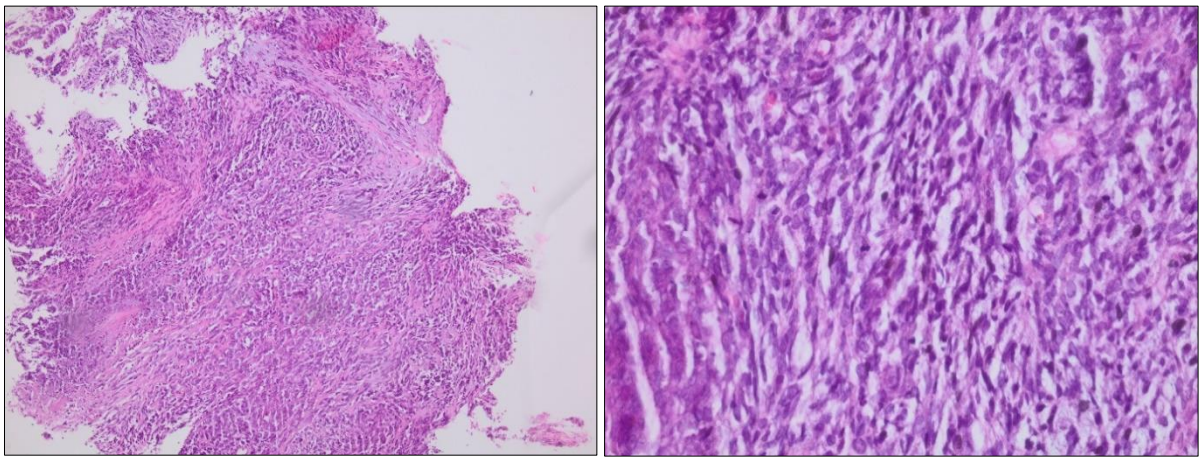

Figure 2: Highlighting spindle cells

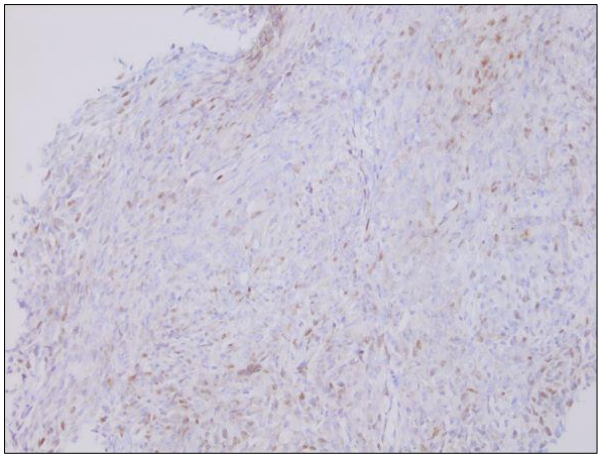

Figure 3: Immunostaining of epithelial cells by P63

The brain CT, done as part of the extension assessment, was normal, however the positron emission tomography (PET scanner) had also found the hypermetabolic left upper lobar pulmonary process (SUV Max: 15.3) locally. advanced, homo and contralateral hypermetabolic nodules, right adrenal involvement and bone lesions. The diagnosis of stage IVb pleomorphic lung carcinoma was then made. The file was discussed in a multidisciplinary consultation meeting (RCP) and the decision was to start palliative chemotherapy based on paclitaxel and carboplatin. Immunotherapy could not be started given the absence of PDL-1 measurement. Death occurred 2 months after diagnosis.

DISCUSSION

According to the definition of the WHO classification of lung tumors, pleomorphic lung carcinoma is a rare lung tumor belonging to non-small cell carcinomas and more precisely to sarcomatoid lung carcinomas [1]. Its incidence varies between 0.1% and 0.3% of all lung tumors [2, 3], and can reach 0.9% according to the study by K.I to *et al.*, [4].

The epidemiological profile is similar to that of other lung cancers. It mainly affects men aged over 60 who smoke; no clinical sign is specific to it, but two forms are classically described. A central endobronchial form responsible for cough, hemoptysis, dyspnea and recurrent pneumonia. A second peripheral form, responsible for pain following invasion of the chest wall [4, 5]. Deterioration of general condition is common due to the rapidly metastatic nature of the tumor [6]. An incidental radiological discovery was reported in 6.7% of cases [4-7].

On chest imaging, these tumors are most often large, peripherally located with a predilection for the upper lobes and invasion of adjacent mediastinal and vascular structures [1-8]. The definitive diagnosis is anatomopathologically based on bronchial or transparietal biopsies. But only a complete examination of the resection specimen allows precise typing due to the histological heterogeneity of sarcomatoid carcinomas [4]. Macroscopically, these tumors are composed of an intimate mixture of carcinomatous and sarcomatous elements [1-9]. The carcinomatous component most often corresponds to an adenocarcinoma (45%), a large cell carcinoma (25%), a squamous cell carcinoma (8%) or a poorly differentiated type of NPCC [4]. The sarcomatous contingent is made up of spindle and/or giant cells representing at least 10% of tumor proliferation [10].

Immunohistochemistry is very useful and has great interest in confirming thediagnosis when the macroscopic study is not sufficient [11]. Indeed, the epithelial component expresses cytokeratins, epithelial membrane antigen, carcino-embryonic antigen, p 21, p 27 and the tumor suppressor gene. In contrast, the sarcomatous component expresses vimentin, fascin and CD 34 [19].

A treatment standard for pulmonary pleomorphic carcinoma (PPC) has not been established due to its rarity [12]. Among NPCCs, PPC is known to be very aggressive in its biological behavior and resistant to conventional treatments, such as chemotherapy and radiotherapy [12, 13].

Its therapeutic management is still based on the same principles as that of CNPC, both surgically and medically; depending on the stadium and the field. The surgical approach, although it is the best therapeutic solution, remains delicate due to the frequent invasion of adjacent elements [14-16]. However, the use of immunotherapy has proven effective [12-19], as well as the various targeted therapies when the search for mutations (EGFR, ALK) comes back positive [18].

The prognosis is often poor [17, 18], given the early occurrence of metastases at the usual sites; brain, liver, bones and adrenals, but also in unusual sites with secondary esophageal, small intestine, peritoneal, gastric, pancreatic, gingival, subcutaneous or even renal lesions.

CONCLUSION

Pleomorphic carcinoma remains a rare malignant tumor with a poor prognosis, the diagnosis of which is essentially based on the histological study which identifies the presence of a double component; carcinomatous and sarcomatous. The treatment of choice is surgical in the absence of metastases, which is rarely the case.

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