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Lung Synovialosarcoma: A Case Report and Review of the Literature

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Abstract **Case Report**

Synovialosarcoma is a rare malignant tumor, accounting for 10% of soft tissue sarcomas. It usually develops in the limbs and its pulmonary location is exceptional. We report a clinical case of synovialosarcoma of the lung in a 65-year-old man discovered at a localized stage. This extremely rare tumor has a particular immunohistochemical phenotype, which greatly contributes to the diagnosis. The cytogenetic study confirms the diagnosis by showing the presence of the specific translocation t (X; 18), which characterizes the synovialosarcoma whatever its anatomical location. Through this observation, we insist on the radiology-clinical, therapeutic and prognostic characteristics of this rare tumor often unrecognized by clinicians.

Keywords: Synovialosarcoma, malignant tumor soft tissue.

INTRODUCTION

Soft tissue sarcomas are rare tumors characterized by great anatomical, histological and prognostic heterogeneity. The general incidence of soft tissue sarcomas is estimated between 3 and 4 per 100,000 inhabitants and 50 to 60% of these cancers develop in the limbs [1]. Primary thoracic sarcomas are rare and represent less than 1% of all primary thoracic tumors [2]. Synovialosarcoma is the 4 e soft tissue sarcoma (8-10%) in terms of frequency; it affects the young adult (15-40 years) with a slight male predominance [3]. About thirty cases of primary pulmonary synovialosarcoma have been described in the literature. We report a new observation by presenting the peculiarities of this tumor rarely encountered in clinical practice.

CASE REPORT

Mr. HM is a 65-year-old patient, chronically smoking 25 pack-years and weaned, consulted in decembre 2020 for chest pain and dyspnea associated with a cough producing haemoptoic sputum in a context of deterioration of the general condition. The physical examination was unremarkable. In addition, a notion of tuberculosis contagion was not retained. Frontal chest x-ray showing multiple tone opacities right water.

A thoraco-abdominopelvic scan was requested and showed an ovoid mass of lung (12x 11 x 10 cm) lower right lobe associated with an ipsilateral nodule, pleural and fissural effusion and seat calcifications. This tumor mass presents an hourglass endocanal extension through the intercostal space bone lysis next to it (figure 1, 2).

The abdominal stage did not present any abnormality. The patient underwent a lung biopsy. The anatomopathological examination of the lung biopsy : The tumor is made up of a fairly polymorphic cell population, sometimes arranged in more or less intersecting swirling bundles, sometimes with a pseudo-hemangiopericyte appearance; Moreover, these cells line cavities of variable size taking on an endotheliform appearance; they show moderate to frank cytonuclear atypia, the mitotic index is high, evaluated at 53 mitoses over 10 fields, on The immunohistochemistry complement showed positivity for the anti-EMA , anti-Ki67, anti-cytokeratin and anti- vimentin marker and negativity for the CD34, PS100 type AE1 / AE3 markers.

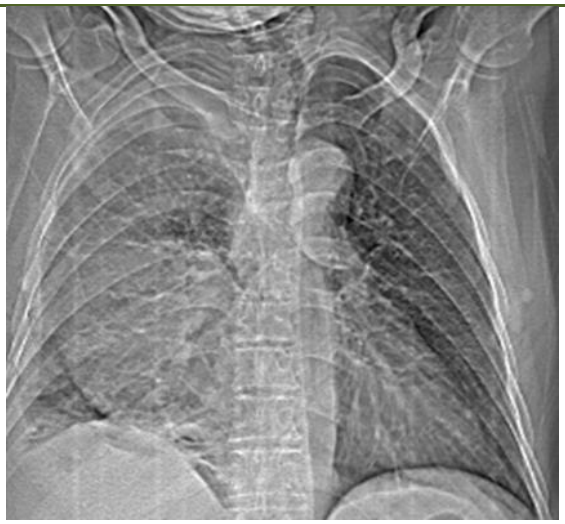

Fig-1: Frontal chest x-ray showing multiple tone opacities right water

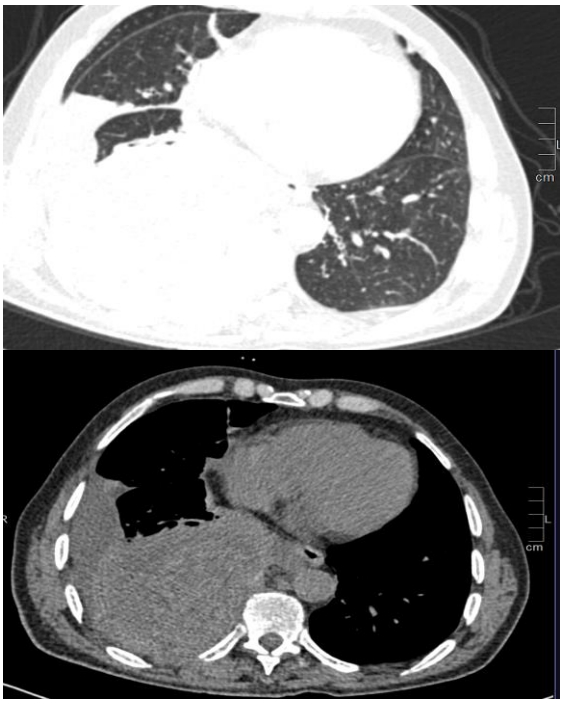

Fig-2: Thoracic mediastinal and lung window computed tomography objectifying a tissue process endothoracic development with encysted pleurisy

DISCUSSION

Soft tissue sarcomas are rare tumors characterized by great anatomical, radiology, histological and prognostic heterogeneity. The incidence of soft tissue sarcomas is estimated between 3 and 4 per 100,000 inhabitants and 50 to 60% of these cancers develop in the limbs [1].

Primary thoracic sarcomas are rare and represent less than 1% of all primary thoracic tumors [2]. Synovialosarcoma is the 4e soft tissue sarcoma (8-10%) in terms of frequency; it affects the young adult (15-40 years) with a slight male predominance [3]. The age of our patient is 65 years, therefore far superior to the data in the literature, but joins the series of Mastroianni *et al*., where the average age of the patients was between 50 and 70 years [4]. Most sarcomas have not been associated with risk factors, but certain genetic and environmental predispositions have been suggested in a minority of patients [5].

Clinically, patient s with synovialosarcoma of the lung usually consults for chest pain, cough progressive hemoptysis and dyspnea over several months [6].

Our patient's picture is typical of this description. Radiologically, there is a mass containing calcifications in 25% of cases. The tomodensitometry makes it possible to better appreciate the presence of micro-calcifications as well as the endothoracic and parietal extension. On magnetic resonance imaging, approximately 90% of synovialosarcomas are well limited with a capsule appearance; the presence of lobulations or septa is frequent. Tumors are heterogeneous in T2 with liquid, solid or fibrous tone signals. The usefulness of positron emission tomography (PET-CT) has been little studied and bone scintigraphy is recommended in cases of bone appeal [6].

Histologically, faced with a malignant tumor proliferation of sarcomatous appearance, a fortiori with spindle-shaped cells, it is always advisable to look for a carcinomatous component in order to rule out the diagnosis of carcinosarcoma. Once the purity of the sarcomatous proliferation has been established, the possibility of sarcomatoid carcinoma must first be ruled out. Unlike synovialosarcoma, this is always rich in cyto-nuclear atypia. In addition, the proliferating cells are there intensely and diffusely positive for epithelial markers. After ruling out these 2 more frequent possibilities, the diagnosis of sarcoma can be made. The question then arises is it a primary or secondary lung tumor? The 2econtingency is by far the most common. The parent tumor is usually located in soft tissue. Only the absence of extra-pulmonary tumor localization in the past, at the time of diagnosis and after 2 years of follow-up will attest to the primary nature of the pulmonary tumor. Concomitant pulmonary metastases can also be indicative of extra-thoracic sarcoma [7]. At the time of death, our patient did not present an extra-thoracic tumor location, in about 2 years of follow-up.

We distinct three subtypes of synovialosaromas : the monophasic form (31%) which a pure fibrosarcomatous form, the biphasic form which combines epithelial cells and spindle cells (36%) and the poorly differentiated form (36%) which contains small oval-shaped cells characterized by a scant cytoplasm and a dense nucleus. On immunohistochemistry, synovialosarcomas express in 90% of cases the Epithelial Membrane Antigen (EMA)

and cytokeratins, in 60% of cases CD99 and in 30% of cases the S100 protein [8]. In most cases, there is a characteristic t (X; 18) translocation that involves the SSX1 or SSX2 genes of the X chromosome (Xp11).

In the absence of metastases, surgery remains the treatment of choice and a large resection is imperative to reduce the risks of locoregional recurrences and at a distance the advantage of adjuvant radiotherapy is to allow better local control of the tumor [11]. It is indicated when the tumor has a diameter greater than or equal to 5cm and incomplete margins. No study has been able to assess the benefit of adjuvant chemotherapy in this situation [11]. Treatment with Doxorubicin and or Ifosfamide constitutes the first-line treatment in inoperable forms and in metastatic forms with a response rate of around 50% [11]. The average rate of locoregional or metastatic recurrence at two years is 50% [12]. The most frequent metastatic sites are lymph nodes, bone and hepatic. A tumor diameter less than 5 cm, a low mitotic index (Ki 67 <10%), the absence of tumor necrosis, the absence of residual tumor after surgical resection are considered to be factors of a good prognosis.

The 5-year survival varies between 35% and 76% depending on the absence or presence of factors with a good prognosis, respectively [12].

CONCLUSION

Synovialosarcoma of the lung is a rare malignant tumor. Its diagnosis is difficult and requires immunohistochemical and cytogenetic analysis to distinguish it from other mesenchymal tumors. The tomodensitometry makes it possible to better appreciate the presence of micro-calcifications as well as the endothoracic and parietal extension. It is important to observe two years of evolution without extra-thoracic involvement to confirm the primary pulmonary nature of the synovialosarcoma. Surgery remains the standard treatment followed more or less by radiotherapy.

Recurrences are frequent and the prognosis is reserved for the advanced stage with modest benefit from chemotherapy. Our case shows the interest of multidisciplinary collaboration in the management of this tumor.

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