A Rare Case Of Sarcomatoid Carcinoma Of The Lung

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ABSTRACT

Sarcomatoid carcinoma (SC) is a rare primary malignant tumor in which both carcinomatous and sarcomatous elements occur. Pulmonary location is particularly uncommon. Pulmonary sarcomatoid carcinoma (PSC) is a rare malignant cancer composed of sarcoma and sarcoma-like tumors with spindle or giant cell features. We present a case of a 64 year old man with a three month history of cough, hemoptysis and fatigue. Chest computed tomography (CT) scan revealed a 5 cm tumoral process at the apex of the left lung. Scannaried transparietal biopsy was performed and the histological examination concluded that adenocarcinoma was poorly differentiated and infiltrated the pulmonary parenchyma. The pathological stage was PT3N0MX based on the tumor node metastasis (TNM) staging system. The tumor’s pathology, histology, immunohistochemistry and treatment are discussed.
INTRODUCTION:
Primary pulmonary Sarcomatoid carcinoma (SC) is a particularly uncommon and aggressive biphasic lung tumor, it accounts for only 0.1 – 0.4 % of all lung cancers. The WHO 2015 classification brought together under this term any carcinoma having pseudosarcomatous cells appearance (spindle cells and/or giant cells) or sometimes heterologous sarcomatous component [1-2]. Here, we present a report of the clinical, radiological and anatomo-pathological particularities of a patient with a Primary pulmonary sarcomatoid carcinoma.

CASE REPORT
A 64 year old man, chronic smocker with a history of 60 pack years complaints of coughing, fatigue and hemoptysis evolving for 3 months. The clinical examination was strictly normal. Chest computed tomography (CT) scan revealed a 5 cm tumoral process at the apex of the left lung. Scannaried transparietal biopsy was performed and the histological examination concluded to a poorly differentiated adenocarcinoma infiltrating the pulmonary parenchyma. A left lobectomy was performed. The specimen was immediately transferred to our department as fresh tissue without fixative, it was inflated with fixative (10% neutral buffered formalin) and sectioned. Gross examination showed a well-circumscribed masse of the apex, measuring 5 cm in diameter, hard in consistency. The cut surface was white gray with areas of hemorrhage, necrosis and cavitation (Fig1).

Fig1: Gross examination of the masse of the apex

Fig2: Dual carcinomatous and sarcomatoid tumor proliferation (HE, magnification 200x)
Fig3: Immunohistochemical staining of the resected tumor specimen: Cytoplasmic expression of the glandular contingent for anti-CK7 antibody

Fig4: Immunohistochemical staining of the resected tumor specimen: Cytoplasmic expression of sarcomatoid cells for anti-vimentine antibody

Microscopic examination showed sarcomatoid tumor proliferation. Histologically, the tumor was composed of compact cells arranged in cords. The tumor cells were pleomorphic, irregular, ovoid, or spindle-like in shape, and some of them had an indistinct cell boundary. Importantly, severe necrosis was found inside. Multiples levels for HE staining were performed, and showed afterwards, a minor carcinomatous component composed of glands of varying sizes (Fig2). Immunohistochemistry stained positive for vimentine in the sarcomatoid contingent and for cytokeratin 7 in the glandular contingent. It showed lack of expression of tumor cells of CD45, EMA, Napsin, Napsin, P63 and CK20 antibodies (Fig3). The selected diagnosis was sarcomatoid carcinoma, subtype pleomorphic carcinoma. The pathological stage was PT3N0MX based on the tumor node metastasis (TNM) staging system.

DISCUSSION
Primary pulmonary sarcomatoid carcinoma is a very rare tumor. SC with spindle cells were categorized as a variant of squamous cell carcinoma back in 1981 [3]. Later in 1999, those with spindle and/or giant cells were classified as carcinomas with pleomorphic,
sarcomatoid, or sarcomatoid elements [4]. The WHO 2004 classification defined PSC as poorly differentiated non-small cell carcinoma that have a histological appearance suggesting mesenchymal differentiation [5]. Diagnostic criteria and terminology didn’t change in the WHO 2015 classification, except for the molecular testing recommendation [6].

Men are more commonly affected with a higher male-to-female ratio. The mean age for diagnosis is 50 - 80 years with a median age of 51.4 years. Smoking is the main risk factor for SC. The patient in our case also had a long history of cigarette smoking. It commonly arises as a large solitary, peripheral mass in the upper lobes like the other smoking-related NSCLC. In our case it was located in the apex [7-8].

The clinical presentation is not specific as it is the case in our patient, usual symptoms are cough, hemoptysis, chest pain, shortness of breath, fever and weight loss. The clinical signs and symptoms may be related to tumor localization. Hemoptysis is mostly seen in cases of proximal or central tumors, peripheral tumors may be asymptomatic or may present with chest pain[1-9]. Paraneoplastic syndromes due to SC have not been reported so far, although they are seen in around 5–8% of NSCLC [1-10].

Imaging modalities like CT scan, MRI, and PET scan seem to be less efficient in diagnosing SC of the lung as it resembles and mimics malignant pleural mesothelioma and pulmonary aspergillosis on imaging [11].

On gross examination, the morphology of the tumour varies. SC are usually well circumscribed grey tan masses measuring > 5 cm in diameter hard or rubbery in consistency. The cut surface varies from white gray to tan-yellow in color with frequent areas of hemorrhage, necrosis and/or cavitation [1-12].

Hematoxylin and eosin (H & E) staining is key to diagnosis. It shows a carcinomatous and sarcomatous double tumoral contingent. It is important to provide multiple levels and and serial sections in order to minimize the chance of missing the dual contingent. In our case, the minor carcinomatous contingent showed after several levels of HE staining. The carcinomatous component is often squamous, sometimes adenocarcinomatous, and rarely composite. In our case, the epithelial component was a moderately differentiated adenocarcinoma type associated with a sarcomatous component [1-2-6].

The final diagnosis is attained through IHC as it is sometimes difficult to distinguish SC from true sarcomas, if no carcinomatous areas are recognized on the H & E stained sections. It helps to better highlight the different cell components of the tumour. The carcinomatous elements stain positive for cytokeratin, EMA, and carcinoembryonic antigen (CEA). The sarcomatous or sarcomatoid component, stains positive for vimentin. In our case, Immunohistochemistry stained positive for vimentin in the sarcomatoid contingent and for cytokeratin 7 in the glandular contingent of CK7. It showed lack of expression of tumor cells of CD45, CD30, EMA, Napsin, Melan A, TTF1, Napsine, P63 and CK20, which was in accordance with previous reports [1-2-6-7].

As it is a rare malignancy, there are no clearly defined guidelines for the treatment of SC of the lung. Radical surgical resection remains the best option, especially for localized tumors. Radiotherapy and chemotherapy are being used as adjuvant therapy or in cases where the patient is a poor surgical candidate because there seems to be a little benefit*. Chemotherapy used for the treatment of SC is the same as used for NSCLCs.

The prognosis of this tumor is poor, with a survival at 2 years <10% because it is an aggressive tumor with advanced local stage and metastasis at the time of diagnosis; and refractory to chemoradiation therapy. It has a worse outcome than conventional NSCLC. The early identification of the tumor followed by surgical and adjuvant therapy might lead to prolonged survival [1-6-7-10]

CONCLUSION

Sarcomatoid carcinomas of the lung are uncommon aggressive cancers with a poor prognosis. Large samples, multiple level HE stain and immunohistochemistry are of a big help to set the right diagnosis.

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