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Case Report

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Pulmonary spindle cell carcinoma: case report and literature review

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Abstract

Spindle cell carcinoma is a rare and malignant variety of squamous cell carcinoma. It is a tumor that is constituted by a double cell proliferation: a carcinomatous sarcomatous epithelial cells of spindle cells and another sarcomatous of spindle cells. It can affect any part of the body; however, it is more commonly found in upper airway and digestive tract. It affects men more frequently between the sixth and seventh decade of life. It has an aggressive behavior with a tendency of recurrence. Alcohol and tobacco have been identified as the most important risk factors. The histopathological diagnosis is complicated and it is often necessary to resort to immunohistochemical techniques and the use of the electron microscope. Given the infrequent strain, it is considered important to publish the information collected from a case in our environment to compare it with what is currently described in the literature.

Introduction

Sarcomatoid neoplasms located in the lung and pleura are extremely rare, with an estimated incidence between 0.3 and 1.3% of all malignant lung neoplasms. The belong to a group of little differentiated non-small cell carcinoma that contain sarcoma, fusiform or giant cell components. According to the World Health Organization, it is classified into pleomorphic carcinoma, spindle cell carcinoma, giant cell carcinoma, carcinosarcoma and pulmonary blastoma.[1] The subtype of spindle cell carcinoma usually affects the oral cavity, larynx, breasts, kidney, uterus, conjunctiva, prostate, other organs, being rarely affected the lung.[2]

Affected patients are predominantly males (4:1) with a history of smoking, with an average age of approximately 65 years. The most common symptoms are cough and hemoptysis. The course of the disease is always aggressive, and the best indicator of prognosis is the stage.[3] The prognosis of patients with this entity is poor, with a 5-year survival rate of approximately 20%.[4]

The histopathological diagnosis plays the most important role in the characterization and classification of these tumors. A diagnosis can be suspected with small samples, but the definitive diagnosis is only possible with the complete resection of the tumor due to its heterogeneity and its histological pleomorphism.[4,5] It can be a challenge to make a diagnosis, especially with small biopsies, because sarcomatoid mesothelioma, metastatic melanoma, and high-grade sarcomas may show overlapping morphological features. It is for this reason that immunohistochemistry is often used to evaluate the sample of thoracic tumors together with specific markers, becoming of great potential value.[6]

In the present study, a case of a spindle cell carcinoid diagnosed after cryobiopsy was studied, being reported by histopathology with immunohistochemistry.

Case Report

A 60-year-old man who is referred from the emergency room to the hospitalization due to a pulmonary tumor. As a background of importance, he denies chronic degenerative diseases, allergies, surgeries, traumas or transfusions. He reports having been hospitalized on one occasion for heroin poisoning, which he has been using for more than 10 years and suspended approximately 5 years ago. He is a smoker who consumed 8 cigarettes a day for 10 years, but this was also suspended 5 years ago. The patient engages in social alcoholism without drunkenness, and positive COMBE because he had been in rehabilitation centers and in prison.

His current condition started 1 month before admission with nonproductive cough, acute, with no preferences of time, no precipitants, and no hemoptoic or hemoptysis, and evolved to productive in less than a week of hyaline and paroxysmal characteristics, nocturnal diaphoresis, and persistent fever predominantly at night. It is not quantified and accompanied by dyspnea mMRC 2, with improvement in rest and pain of the pleuritic type at infrascapular level, right from insidious onset, without irradiation with weight loss of 7 pounds in 10 days. In his local health center, it is suspected that the patient has tuberculosis, although the bacilloscopy results are negative. The patient is referred to second level of assistance. On the physical examination at the time of admission, decreased right-amplexion and decreased ipsilateral amplexation are noticed, as well as dullness to percussion and abolition of right-sided breathing noises. In the left hemithorax, thick rales at inspiration were identified, rest without alteration. Chest radiography was performed in which total radiopacity of the right side is observed, which correlates with pleural effusion of probable parapneumonic origin (Figure 1).

The laboratory studies reported Hb of 11.1 g / dL, HCT 34.7%, MCV 78.4 fL, MCH 25.2 pg, platelets of 508x103, total leukocytes

of 18,340 uL, with neutrophils of 17,550 uL (95.7%), lymphocytes 460 uL (2.5%) monocytes 290 uL (1.6%), eosinophils of 0%, and basophils of 40 uL (0.2%). This is why third-generation cephalosporin and macrolide are initiated for suspected community-acquired pneumonia with pleural effusion. The following approach was to perform a contrast computed tomography of the thorax, which reported a tumor with central necrosis in the right hemithorax of 3.5 inches x 4.3 inches, infiltrating the right pulmonary bronchus from its origin in the main carina, with peripheral enhancement, practically collapsing the right hemithorax in its entirety, with left hemithorax without evident alterations, and mediastinum with retraction towards affected hemithorax as well as infiltration to fat, presence of adenopathies reporting 1.1 inches as the largest diameter located sub-carinal. The esophagus showed a loss of fat separation interface in the middle third, which is why it is considered infiltration of the same. Bone tissues showed only degenerative changes and soft tissues with hypotrophy. The department of diagnostic radiology suggested the diagnosis of bronchogenic cancer, possibly of large cells, thus requiring histopathological confirmation. Macrolide was suspended and clindamycin was initiated for probable post obstructive pneumonia. During his hospital stay prior to his bronchoscopy, the patient was found to have a transcutaneous oxygen measurement of 89% without supplementary oxygen, which improved with the administration of supplementary oxygen by nasal tips at 0.5 L/min. Continuing with the same symptomatology, the patient was stable. A bronchoscopy was performed with diagnostic cryobiopsy, showing extrinsic compression of the middle third of the trachea, 100% obstruction of the right main bronchus with extension to the main carina and permeabilization of the intermediate bronchus, persisting with obstruction of the lobar bronchi and partial recanalization of the basal trunk. Therefore, bronchial lavage was sent to culture and cytology, as well as cytology and immunohistochemistry biopsy. Crops reported negative and the pathology reports indicated Fusocellular malignant neoplasm compatible with spindle cell carcinoma with focally positive Cytokeratin AE1 / AE3, Negative TTF and Negative P63 (Figures 2-4).

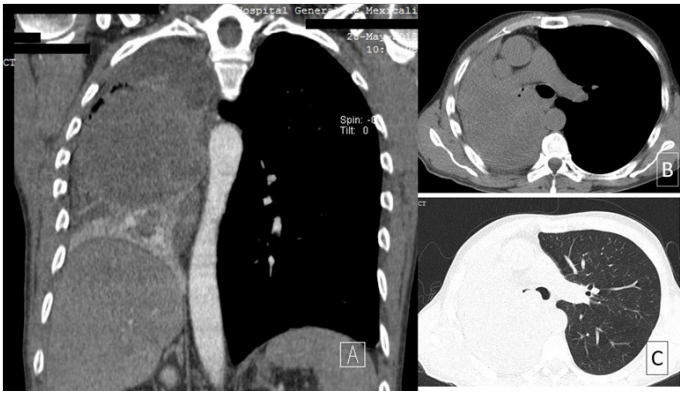

Figure 1 A) Coronal CT-Scan of the chest, mediastinal window B) Axial Slide of a CT-CT-Scan of the chest, mediastinal window C) Axial Slide of a CT-CT-Scan of the chest, Lung Window.

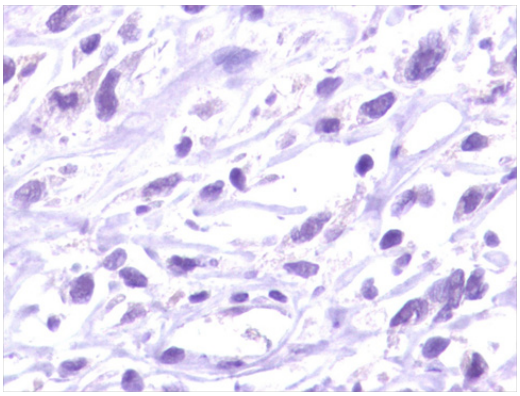

Figure 2 Negative *Immunohistochemical* expression of P63.

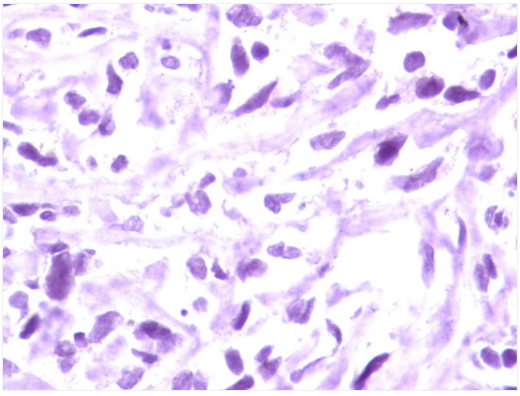

Figure 3 Negative *Immunohistochemical* expression of TTF-1.

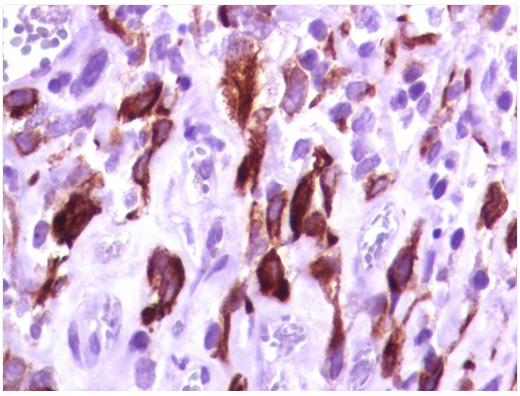

Figure 4 Positive Cytokeratin AE1/AE3.

Discussion

Sarcomatoid carcinoma is the name given by WHO to unify pleomorphic carcinoma, spindle cell carcinoma, giant cell carcinoma,

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carcinosarcomas and blastomas.[4] These tumors are extremely rare with an estimated incidence of 0.3-1.3% of all lung malignancies.[7]

This study reports a case of spindle cell carcinoma of a 60-year-old male patient with a history of smoking for 10 years, which is related to what was written in the literature where the average age is 60 to 70 years of age, the great majority of males in a 4/1 relationship and in smokers.[8]

Symptoms depend on the location of the tumor. It has been described in cases of proximal tumors with cough, hemoptysis in 50% of cases, progressive dyspnea and recurrent pneumonias due to bronchial obstruction more frequently, however, it can also be present. These patients present with chest pain in 25% of the cases of pleomorphic carcinoma.[5]

The imaging studies in the described case were of great help because we managed to identify a generalized radio-opacity on the right side in the case of radiography, as well as a hyperdensity in the computed tomography occupying all the aforementioned hemithorax. It was apparently being observed a necrotic center as well as affection of the costal wall and infiltration to the right main bronchus from its origin in the main carina with peripheral enhancement practically collapsing right hemithorax in its totality, as well as affection to soft tissues. Sarcomatoid carcinomas are frequently large tumors with an average size between 5-8 cm, and in this case was 9cm x 11cm (slightly above the average). More frequently in a peripheral location and in upper lobes, pleomorphic carcinomas and blastomas are the ones that most affect the periphery of the lung. With involvement of the costal wall, the pleomorphic is seen in up to 25% of cases. A central location with or without endobronchial involvement can be seen in all sarcomatoid carcinomas, but is mainly in the carcinosarcomas and less frequently in the pleomorphic ones. These tend to be well circumscribed and without any capsules, often seeing hemorrhage and necrosis, as the latter is seen in the case.[1,5]

A bronchoscopy was performed with diagnostic cryobiopsy where endobronchial involvement extrinsic compression was observed from the middle third of the trachea obstructing 100% of the trachea. These characteristics are commonly present in all the sarcomatoid carcinomas of bronzed yellow color and of soft consistency upon manipulation. During the procedure, samples were taken of bronchioalveolar lavage and cultures for bacteria and fungi analysis.[1,3]

In this case, pathology reported fusocellular malignancy compatible with spindle cell carcinoma by sample taken with cryobiopsy. An immunohistochemistry analysis revealed focally positive Cytokeratin AE1 / AE3, Negative TTF and Negative P63. The use of AE1 / AE3 has been described as the most sensitive compared to Cytokeratin OSCAR and CAM5.2 as a marker for pulmonary sarcomatoid carcinoma. CAM 5.2 also has a high sensitivity, but it has been reported as diffusely positive in less than half of the cases, which can lead to false negatives in small biopsies. The best routine option is the AE1 / AE3 markers alone or in combination with Cytokeratin OSCAR.[6]

In this case, having the histopathological result with immunohistochemistry, we proceeded to refer to oncological hospital for follow-up, being the management of choice in patients with sarcomatoid carcinoma. Depending on the extent of the disease, surgical resection, as well as chemotherapy and radiotherapy, are possible treatment options. Despite surgical therapy, survival is much lower compared to other non-small cell lung tumors even in stage I or II, so extensive surgery must be carefully planned. Without clear evidence that chemotherapy prolongs survival,[8] risks and benefits of surgery are analyzed and extensive surgery must be carefully planned. The prognosis in these patients is poor with a 5-year survival rate of 21.3%.[4]

Conclusion

Sarcomatoid carcinomas are neoplasms with an unfavorable and extremely rare evolution. A timely diagnosis is of vital importance to begin surgical management accompanied by chemotherapy and radiotherapy. However, even if the prognosis is made early, the survival rate is lower than the rest of the non-small cell lung tumors.

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