



ESTUDO DE CASO: TUMOR FIBROSO SOLITÁRIO DA PLEURA

Case report: solitary fibrous tumor of pleura

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Abstract: The present article is an analysis and a study on the Solitary Fibrous Tumor of the Pleura (SFTP), which is a neoplasm of origin in the pluripotent cells of the fibrous mesenchyme. The neoplasia is not exclusive to the pleura, it also affects extra thoracic tissues such as muscles, soft parts, pelvis and abdomen. It is a rare tumor type pathology, affecting approximately 2.8 patients in 100,000, corresponding to 5% of all pleural neoplasms. It presents more than 80% of tumors characterized as benign. It has unknown etiology and is not associated with genetic factors or life habits. Biopsy and immunohistochemical analysis are essential for the definitive diagnosis of the disease, as well as imaging test. The treatment of choice is complete surgical excision. The study also analyzes the case of a 46-year-old male patient who was admitted to the Montes Claros hospital in January 2018. Therefore, the study addresses all the disease characteristics that have developed in this patient, as well as the clinical, diagnostic and therapeutic discussions of the Solitary Fibrous Tumor.

Keywords: Solitary Fibrous Tumor; Pleura; Treatment.

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Resumo: O presente artigo faz uma análise e estudo de caso acerca do Tumor Fibroso Solitário de Pleura que é uma neoplasia de origem nas células pluripotentes do mesênquima fibroso. A neoplasia não é exclusiva da pleura, acomete também tecidos extratorácicos como músculos, partes moles, pelve e abdômen. É uma patologia de tipo de tumor raro, afetando aproximadamente 2,8 pacientes em 100.000, correspondendo a 5% de todas as neoplasias pleurais. Apresenta mais de 80% dos tumores caracterizados como benignos. Possui etiologia desconhecida, não sendo associado a fatores genéticos, nem a hábitos de vida. A biópsia e a análise imuno-histoquímica são essenciais para o diagnóstico definitivo da patologia, assim como os exames de imagem. O tratamento de escolha é a excisão cirúrgica completa. O estudo faz ainda uma análise do caso de um paciente do sexo masculino com idade de 46 anos, que deu entrada no hospital de Montes Claros em janeiro de 2018. O estudo aborda, portanto, as características da doença, que desenvolveram neste paciente, bem como as discussões clínicas, diagnósticas e terapêuticas do Tumor Fibroso Solitário.

Palavras – chave: Tumor Fibroso Solitário; Pleura; Tratamento.

INTRODUCTION

The solitary fibrous tumor of the pleura (SFTP) is a neoplasia of origin in the pluripotent cells of fibrous mesenchyme¹⁻². The first description of the disease in the pleura was performed in 1930 by Klemperer and Rabin as the most common site of affection, previously, the tumor was known as pleural fibroma or benign fibrous mesothelioma²⁻³.

The tumors of the pleura according the World Health Organization (WHO) are classified by morphological aspects in three histogenetic classes: mesothelial tumors, mesenchymal tumors, and lymphoproliferative disorders¹.

SFTP is a primary cancer of the pleura, of insidious growth and oligosymptomatic, but presents a high rate of recurrence after surgical resection⁴. Two-thirds of this tumor have their origin in the visceral pleura and one third in the parietal pleura⁵. SFTP is not exclusive to the pleura, it also affects extra thoracic tissues such as muscles, soft parts, pelvis and abdomen.

It is a rare tumor type pathology, affecting approximately 2.8 patients in 100,000, corresponding to

5% of all pleural neoplasms². As it an uncommon neoplasm, it is poorly described in the literature, with most of the knowledge about the disease from case reports and case series. The incidence is similar among men and women, and the peak occurs from 50 to 80 years. Its etiology is unknown, and there is no connection with asbestos or smoking⁶. The majority follows a benign course, but a subset will recur or metastasize⁷.

SFTP is asymptomatic in the early stages of the disease, however with the evolution of the disease nonspecific signs and symptoms are observed that can direct the clinical reasoning; for other diseases such as pneumonia, congestive heart failure disease and also other more common types of neoplasms affecting the respiratory system. The most commonly observed initial symptoms are: chest pain, dyspnea, cough and hemoptysis. A large part of these symptoms is due to local compression caused by the tumoral growth. The diagnosis is made through the evaluation of the immunohistochemical profile and the SFTP treatment is based on surgical resection³.

This case report has as background the need to expand the

scientific knowledge about the SFTP, which is a rare neoplasm that affects other organs, but which has a shortage of publications about the same. In addition, it contributes with information on the clinical management, the development, diagnosis and therapy.

The objective of this study is to report the case of a patient with Solitary

CASE REPORT

Patient V.P.L, male, 46 years old, faioderm, non-alcoholic and non-smoker, was admitted to the hospital in the city of Montes Claros on January 25th of 2018, with a history of chest pain on the right and a dry cough started 45 days ago. It was detected on chest x-rays and computed tomography scan of the chest, a lung mass with approximately 6 cm in diameter in the right Para cardiac region without communication with the bronchial tree. It was requested, then immunohistochemistry which showed the diagnosis of Solitary Fibrous Tumor of Pleura.

On physical examination, the patient was ruddy, hydrated, without adenomegaly, eupneic, with good tissue

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Fibrous Tumor of the Pleura.

This study was conducted in accordance with the precepts established by Resolution 466/12 of the National Health Council of the Ministry of Health, in accordance with the approval of the Research Ethics Committee of UNIMONTES, upon embodied opinion no 2.599.222.

perfusion and without edema, panniculus and well distributed adnexa according to gender and age. Normal rhythmic and normal phonetic heart sounds in two times (BNRNF2T), flaccid abdomen, absence of palpable masses or visceromegaly, reduced vesicular murmur in the right lung base, with dullness sound to percussion. Vital Signals: B.P: 100x60 mmHg, HR 76 bpm, F.R: 18 Irpm, Axillary temperature: 37 °C.

In the first tomography, requested subsequently to the chest X-ray showed pulmonary mass with density of soft parts occupying almost entirely the middle lobe of the right lung, without fibro atelectatic aspect and not affecting adjacent osseous structures. Another characteristic of the lesion is to hold a mass effect on

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regional structures. There was no evidence of pleural effusion and lymph nodes in the mediastinum.

During the investigation, a fibro bronchoscopy was performed which revealed a nodular formation in the posterior wall distal in the left main bronchus. The patient was then subjected to the lower right lobectomy with voluminous resection of the pulmonary mass. He did not have postoperative complications. To the new X-ray, the presence of pneumothorax, mild pleural effusion with atelectasis on the right was detected.

The anatomopathological study revealed the following aspects: in macroscopy the material was composed by the right lower lobe of the lung measuring approximately 18.0 x 15.5 x 18.0cm, covered by serous predominantly smooth, presenting area of rupture of the serosa by whitish firm elastic mass that invades the pulmonary parenchyma. The lesion measures at least 13.0 x 8.0 x 3.5cm. Upon the cuts the lesion affects more than 60% of the pulmonary parenchyma. The remainder of the spongy parenchyma is mixed with brown and whitish areas. It accompanies several portions, similar to nodular pulmonary lesion, measuring 25.0 x 16.0 in x 8.0 cm.

In the microscopy, it revealed

that the lower right lobe displays undifferentiated spindle cell sarcoma characterized by spindle cell neoplasm consisting of elongated cells, nuclei with prominent nucleoli forming verticular arrangements and permeated by vessels of varying sizes; size 13,0 x 8.0 x 3.5cm; Mitotic index: 16/10CGA; necrosis present in 20% of the lesion; histological grade 3; not evaluated surgical margins; anamopathological staging: pT4a. The immunohistochemical study (CD34 +, MIB1 TLEI - EMA - WT-1 - Podoplanin - Protein S-100, Ki67 +) requested further confirmed the diagnosis of Solitary Fibrous Tumor of the Pleura.

Chemotherapy was started around 2 months after the right-side lobectomy. In the first regimen, Ifosfamide (Envolox®) (D1 to D5), Doxorubicin Hydrochloride (Adriplastina®) (D1 to D3) associated with Mesna®.

Around 2 and a half months after chemotherapy was started, new CT scan of the chest was requested, which showed a slight reduction in the dimensions of the voluminous heterogeneous paravertebral mass which measured approximately 14.2 x 11.8 cm and passed to measure approximately 12.2 x 8.6 cm. In the same examination

it was observed the appearance of erosions in the right costal arch and the adjacent vertebral body. It also presented stability in the size of the lesion along the right anterolateral wall of the thorax, measuring approximately 3.9x 2.3cm. However, it was observed the appearance of other heterogeneous masses, being the anterior paravertebral region in the thoracoabdominal transition, measuring 8.6 x 6x7cm,

DISCUSSION

The V.P.L. patient presented Solitary Fibrous Tumor of the Pleura, classified histogenetically as mesenchymal type, according to the morphological aspects of WHO division. The most common reported site of SFTP is the pleura, and the extra pleural sites are rare foci of the disease⁸. The case described corroborates this information, demonstrating that even being a rare neoplasm, the respiratory system, more specifically, the pleura is the main focus of the disease.

SFTP has a benign behavior in up to 88% of cases, and there is no

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along the anterior thoracic wall, measuring 8.1 x 5.5 cm the largest ones, in addition to other minor injuries in the anterior and posterior hepatophrenic spaces. Sparse nodular thickening in the pleura to the right is also associated with it. Emergence of small pleural at the right side. The lesions determine mass effect on the regional structures, notably on the right and left atria.

association with genetic factors, occupational hazards and previous exposure to asbestos⁹.

Most people with diagnosis of SFTP are more than fifty years old. The peak of the pathology is between the 5th and 8th decade. The signs and symptoms of SFTP are very specific to the disease, being the majority of the symptoms classified as general⁶⁻¹⁰. In 46.6% of the cases, the diagnosis was causal and without any association with the presence of systemic manifestations¹¹. However, in 20% of the cases the main complaints are cough, dyspnea, chest pain and hemoptiase⁶. In the description of the case report it is evident that the patient shares the

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same epidemiological and symptomatic characteristics. The age of diagnosis of the presented case presented, happened at 46 years, age next to the peak of the neoplasia. And the patient in the first consultation presented as the main symptom, dry cough.

The investigation of the disease, occurs through the patient's clinical symptoms, associated with the imaging exams, x-rays and computed scan are the most commonly used. But, the diagnostic confirmation only happens through the anatomopathological and immunohistochemical analysis. In some analysis of imaging examinations, it was seen that 80% of the computed tomography scans, demonstrated a single tumor and 20% had multiple tumors, the average size of the tumors was approximately 7.35 cm¹¹. The patient of the report presented in the beginning a tumor of significant size, approximately 14 cm, almost two times greater than the tumors found in studies of images. In addition, the case demonstrates that subsequently, the emergence of new masses was observed, which makes the individual reports to be within a small percentage of the group that presented multiple lesions.

From the histopathologic point of view, 55% of the analyzes showed no atypia, 25% showed necrosis and mitosis more than 4 per field in 30% of the results. The immunohistochemical profile is determined by a combination of positive markers such as vimentin and CD34, BCL2 and CD99 with negativity of PCK and D S100. In 85% of cases there was positivity for CD34, BCL2 and 40% for CD99 (40%), and in 100% of the keratins and protein S100 were negative¹¹.

According to the anatomopathological evaluation of the patient V.P.L. there were fusiform, atypical, moderate cells with presence of mitotic and necrosis contents. Regarding the immunohistochemical analysis there was positivity for the markers CD34 and Ki67 and negativity for S-100 protein.

The positivity of CD34 is required for differential diagnosis with mesothelioma of fusiform cells. From this diagnostic marker diseases are excluded such as: monophasic synovial sarcoma, leiomyoma, malignant tumor of the nerve sheath and fibrosarcoma⁵. The negativity for S-100 protein is important to exclude tumors of the nervous system¹¹.

The treatment of choice for the TFSP is surgery. In some situations, when the tumor invades the pulmonary parenchyma, it is necessary to perform a lobectomy⁵⁻¹¹. In 9-19% of the cases, the local recurrence is observed even after the surgical procedure, despite the complete excision, in 19% metastases occur outside the primary focus and in 0-27% the death is associated to the tumor¹¹. As described by clinical case,

FINAL CONSIDERATIONS

The Solitary Fibrous Tumor of the Pleura is a rare neoplasm, but that evolves with clinical presentation similar to signs and symptoms of the majority of the diseases of the doctor's every day. Because of this, it is important to extend the registration and survey forms of this disease, which are scarce, with the purpose of providing a more objective and accurate diagnostic investigation. Reducing spending on unnecessary exams and increasing the real chances of cure, due to the fact of being carried out the diagnosis of the disease so early. The present study also aims to foster discussion on the topic. It is recommended that scientific research

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the patient underwent surgery, however, due to the large size of the mass and the involvement of adjacent structures, there was not total resection, only partial, with total excision of the right lower lobe and part of the middle lobe of the right lung. The patient underwent subsequently to chemotherapy with the goal of reducing the size and infiltration of the tumor. The patient was well and in a good state of health.

with more robust designs be conducted, with the aim to determine if there are other treatment options, besides the surgical, impacting on the improvement of the quality of life of people affected by this neoplasm.

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