VOLUME-9, ISSUE-5, MAY -2020 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper | Clinical Science

PLEUROPULMONARY SYNOVIAL SARCOMA: PRESENTATION OF AN EXCEPTIONAL CASE

Valdiviezo Allauca Nataly Sofía*	MD. General Practitioner *Corresponding Author/Riobamba
Chávez Berrones Mayra Silvana	Medical resident at "Hospital de Especialidades de las Fuerzas Armadas"/Quito-Ecuador
Viviana Karina Moyano Paz y Miño	Medical resident at "Hospital de Especialidades Carlos Andrade Marín"/Quito-Ecuador
Gabriela Paulina Valle Pazmiño	Medical resident at "Hospital de Especialidades de las Fuerzas Armadas"/Quito-Ecuador
Alexis Santiago Mejía Arias	Medical resident at "Hospital de Especialidades de las Fuerzas Armadas"/Quito-Ecuador
María Belén Pinduisaca Verdugo	Medico Rural at "Centro de Salud Sangolqui"/ Quito-Ecuador

ABSTRACT Pleuropulmonary synovial sarcoma (SSPP) is a morphologically, clinically, and genetically distinct entity and can be seen in any location. It is a mesenchymal spindle cell tumor with variable epithelial differentiation. There is no standardized therapy, generally the treatment is surgical. We present the case of a 40-year-old patient who presents with nonspecific symptoms and an imaging study reveals a pleural effusion, without response to treatment. Extension, paraclinical and open lung biopsy studies were performed, which reported positive for malignancy and immunohistochemical studies diagnosed monophasic synovial sarcoma. Despite adjuvant treatment, it evolves unfavorably and dies.**Objective:** Learn about Pleuropulmonary Synovial Sarcoma through the presentation of an exceptional case, so that readers are trained to identify the key points of the pathology (main epidemiological, clinical and pathological characteristics) despite its rare presentation.**Method:** This is a retrospective study of monophasic pleuropulmonary synovial sarcoma, emphasizing its clinical, radiological and histological characteristics.**Conclusión:** SSPP is an extremely rare primary lung tumor with a description based on a limited number of cases. The diagnosis can only be established after clinical and imaging studies have ruled out any other possible location that justifies lung extension. Likewise, a detailed immunohistochemical study is required for its definitive diagnosis. Treatment is surgical and its prognosis is poor with an overall survival rate of 50% at 5 years.

KEYWORDS : Synovial sarcoma, Pleural pathology, Immunohistochemistry

INTRODUCTION

Pleuropulmonary synovial sarcoma (SSPP) has been an entity described in the literature for 15 years. It belongs to the group of synovial sarcomas, malignant tumors of spindle cell mesenchymal origin characterized by a variable epithelial differentiation, and cytogenetically by a specific chromosomal translocation t(X; 18) (p11.2; q11.2)[2]

It represents one of the most frequent malignant tumors of the soft tissues, accounting for approximately 8% of all of them. It is a rare aggressive neoplasm that generally affects people between the ages of 15 and 50. Although the lung is the most common site of soft tissue sarcoma metastasis, pleural metastases from synovial sarcoma are very rare[3].

Despite its name, synovial sarcoma does not arise from synovial tissue. As with most soft tissue sarcomas, the tissue from which it originates is unknown and its risk factors are not well established; therefore, there is no possibility of prevention. It can be asymptomatic, but it is often suspected due to the appearance of dyspnea or pleuritic chest pain.[4]

The best therapeutic option is not clear since it varies from resection, chemotherapy (with doxorubicin and ifosfamide), radiotherapy and even hyperthermic therapy in inoperable tumors, having in any case a rather poor prognosis[2]

We present as a case a middle-aged adult patient, who presented with nonspecific symptoms of pain in the left gluteal region, and subsequently presented MMRC4 dyspnea. With a chest tomography image of a bilateral pleural effusion that did not respond to thoracentesis and permanent thoracostomy, it was decided to perform lung decortication and take an open biopsy, which after the histological diagnosis of Monophasic Synovial Sarcoma initiates chemotherapy treatment and in the absence of response finally passes away

METODOLOGÍA

This is a retrospective study of a case of monophasic pleuropulmonary synovial sarcoma, emphasizing its clinical, radiological and histological characteristics.

The information and images obtained belong to the medical personnel in charge of the case whose reinforcements rest in the statistical package Excel, Word y JPG.

CASE PRESENTATION

This is a 40-year-old female patient, resident in Pichincha, Ecuador, by profession an Electronic Engineer, with only personal pathological history of Anovulation. Patient with previous evaluation for a clinical picture characterized by stitch-like pain located in the left gluteal region irradiated to the left lower limb diagnosed with ciatalgia, so NSAIDs were

prescribed without improvement. 5 days ago, the picture was accompanied by MMRC 4 dyspnea and pleuritic-type chest pain. 2 hours ago dyspnea arrives at orthopnea with this symptomatology goes to a specialized hospital.

Upon arrival at the Pneumology service, a chest x-ray is performed (photo1) and a contrasted chest tomography.

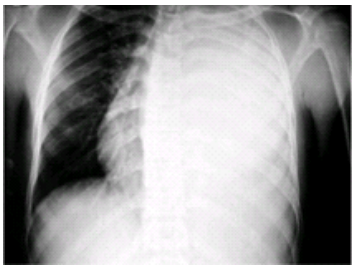

Photo 1. Admission chest radiograph, homogeneous left hemithorax opacity

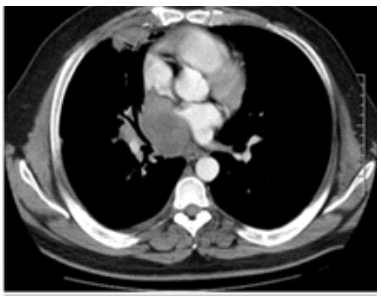

Photo 2. Contrast-enhanced CT shows a soft tissue attenuation mass that invades the pulmonary veins and the left atrium.

Imaging studies decided to perform evacuatory thoracentesis and diagnose, where 2000 ml sallow liquid, mononuclear exudate type, negative liquid cytology for malignancy, in management with placement of a chest tube was observed. Due to persistent septed effusion despite thoracotomy, it was decided to perform drainage, left decortication, and lung biopsy to take samples to rule out Pulmonary Neoplasia, Meigs Syndrome, Pulmonary tuberculosis or other causes.

Extension exams were carried out CA 19-9 7.57, AFP 3.06, CYFRA 2.09, CA-125 25.19 CEA 1.03. In addition, lung decortication was performed and an open biopsy was taken at the lingual level of the right upper lobe, with the presence of 300 cc serous fluid, very thickened parietal pleura, mediastinal and diaphragmatic left pulmonary adhesion, disseminated tumors in the parietal, thoracic, mediastinal pleura. and diaphragmatic that involves the pericardium and the lung. The positive biopsy result for malignancy and for immunohistochemical profile concludes Monophasic Synovial Sarcoma (photo 2)

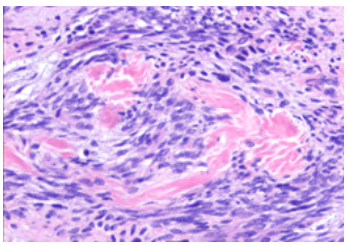

Photo 2. Monophasic Synovial Sarcoma. Spindle cell proliferation in a fibrous hyaline stroma.

Oncology service treatment started with ifosfamide + doxorubicin, he received only 1 cycle, later due to severe acute respiratory failure patient dies.

DISCUSSION

The SSPP is a morphologically well-defined neoplasm whose most common location resides in soft tissues, in the vicinity of the large joints. Despite its current name, based on old descriptions that are now out of date, there is no evidence that synovial sarcoma originates or differs from synovial tissue and its extra-articular location attests to this; furthermore, immunophene-typical and molecular studies confirm the phylogenetic differences of neoplasia and synovial tissue. However, it is a well-established nomenclature that struggles to stay current.5

The morphological spectrum of synovial sarcoma is broad, with four main variants or phenotypes having been described: biphasic, monophasic fibrous, monophasic epithelial (the least frequent) and the poorly differentiated.6-7

The most frequent clinic is cough and hemoptysis, the latter due to the presence of cystic formations and necro-hemorrhagic lesions of the tumor. 25% of cases are asymptomatic. [8]

They present as single nodules or lung masses, usually with well-defined and lobed margins. They can grow very quickly, reaching a large size (greater than 10 cm.) In presentation, heterogeneous. They can also show invasion of neighboring structures by contiguity or be accompanied by an ipsilateral pleural effusion, sometimes very abundant, making it difficult to determine its origin.[9]

In our case, the tumor manifested with veiling of the left pulmonary field, a rare form of presentation.

Treatment is surgical, chemotherapy and adjuvant radiation therapy can reduce and delay the appearance of distant metastases. The response to chemotherapy is moderate, around 50% of cases, with regimens of isophosphamide and doxorubicin. The prognosis is poor, some patients survive up to 20 years; however the average survival is 5-6 years. [10]

Factors that predict a worse prognosis in patients with synovial sarcomas are: tumor size (> 5 cm), male sex, advanced age (> 20 years), extensive tumor necrosis, histological grade. [11]

The case we present shows lung location. This location is infrequent, it is known from the studies carried out on this pathology that affects more men than women in a young population; Our case refers to an elderly female patient, so its epidemiology makes it an even more exceptional case. However, the prognosis of the patient was poor as the literature indicates due to the extension of the tumor and advanced age, which makes it inoperable, so that our patient dies days after having finished her first chemotherapy cycle.

CONCLUSIÓN

Pleuropulmonary synovial sarcomas are a rare entity among primary lung tumors. In our hospital, a patient with this diagnosis was not previously treated, it has been reviewed in the literature at the Ecuador level, being the first clinical case described in our town, this could be because only 3% occupy other neoplasms at the level 12, reason why we ratify its nature. It has been described that the prognosis of the patients depends a lot on whether they meet the criteria of being removable or ineffective because surgery plays an essential role, as well as the various therapeutic modalities that have been discussed.

REFERENCES

1. Pontillo H, Uebistazu J, Gadea C, González R, Mosquera J, Briceño A. Sarcoma Sinovial Monofásico primario de Pulmón. Reporte de un caso y Revisión de la literatura. Revista Venezolana de Oncología. Rev. venez. oncol. v.20 n.3 Caracas ago. 2008

2. Estorsa M, Rinaldia P, Gasparb C, Sánchez F, Esturic R, Galbis J. Neumotórax de repetición como presentación de sarcoma sinovial pleuropulmonar. Servicio de Cirugia Toracica, Hospital Universitario La Ribera, Alzira, Valencia, España. Revista de Patología Respiratoria. Vol. 15. N 12 p 61-63. (Abril 2012).
3. Ferrari A, Collini P. Sarcoma Sinovial. Aglobal initiative for everyone affected by sarcomas. Disponible en http://sarcomahelp.org/translate/es-sarcoma-sinovial.html
4. Gómez A, Campano L, 1, García R, Núñez A, Díaz T, Ávila R, A.B. Enguita Valls3, V. Villena Garrido1, C. Marrón Fernández2. Sarcoma sinovial primario en pared costal. REVISTA DE PATOLOGÍA RESPIRATORIA Vol 22 , Número 1, Enero-Marzo 2019
5. Iwata T, Nishiyama N, Izumi N, Tsukioka T, Suehiro S. Metastatic monophasic synovial sarcoma of the pleura. Ann Thorac Cardiovasc Surg. 2007; 13: 258-61.
6. Torres F, Vázquez F, Torres J. Sarcoma sinovial bifásico pulmonar. Presentación de un caso y revisión de la literatura. Rev. chil. enferm. respir. 2011; 27: 203-207 vol.27 no.3 Santiago set. 2011. Disponible en http://dx.doi.org/10.4067/S0717-73482011000300005
7. Rassard M, Le Dousal V, Hacene K, Terrier P, Ranchere D, Guillou L, et al.. Prognostic factors in localized primary sinovial sarcoma: a multicenter study of 128 adults pacientes.. J Clin Oncol. , 19 (2001), pp. 525-34 http://dx.doi.org/10.1200/jco.2001.19.2.525 | Medline.
8. Essary LR, Vargas SO, Fletcher CD.Primary pleuropulmonary synovial sarcoma: reappraisal of a recently described an atomic subset.Cancer. , 94 (2002), pp. 459-6 http://dx.doi.org/10.1002/cncr.10188 | Medline
9. Fraser RS, Muller NL, Colman N, Pare PD. Anterior mediastinal masses. In: Fraser and Pare's diagnosis of diseases of the chest. 4th ed. Philadelphia, Pa: Saunders, 1999; 2875–2937
10. Lajas A, Serrano F, Rodríguez M, Díez M. Sarcoma sinovial pleural primario: diagnóstico diferencial con el carcinoma pulmonar microcítico. Archivos de Bronconeumología. Vol.54. Núm. 9.pg 488-489. Septiembre 2018
11. Trassard M, Le Doussal V, Hacene K, Terrier P, Ranchère D, GuillouL, et al. Prognostic factors in localized primary synovial sarcoma: amulticenter study of 128 adult patients. J Clin Oncol 2001;19:525-534.
12. Dra. Patricia Cueva Ayal MSc., Dr. José Yépez Maldonado MSc. (2006-2010). SOCIEDAD DE LUCHA CONTRA EL CANCER, SOLCA NUCLEO DE QUITO. Epidemiología del Cáncer en Quito 2006-2010 by SOLCA. https://issuu.com/solcaquito/docs/epidemiologia_de_cancer_en_quito_20