Pulmonary Inflammatory Rhabdomyoblastic Tumor: Case Report

doi: https://doi.org/10.32635/2176-9745.RBC.2023v69n1.3363

Tumor Rabdomioblástico Inflamatório Pulmonar: Relato de Caso

Tumor Rabdomioblástico Inflamatorio Pulmonar: Informe de Caso

Pedro Pinheiro Barros[1]; Henry Martins Soares Fortes[2]; Arthur dos Santos Sena[3]; Victória dos Santos França[4]; Davi de Castro Barros Nogueira[5]; Wolfgang William Schmidt Aguiar[6]

ABSTRACT

Introduction: Primary lung sarcomas are rare malignant tumors with an estimated incidence of around 0.5% of all lung neoplasms. Of their typifications described in the literature, two especially, leiomyosarcomas and rhabdomyosarcomas, have similarities in their genetic, morphological and immunohistochemical profile, which led them to be classified with the same name: inflammatory rhabdomyoblastic tumor. This type of tumor usually affects soft tissues in the extremities and trunk, and predominates among young and middle-aged men. Due to the small number of cases of rhabdomyoblastic tumors reported in the literature, both their diagnosis and treatment are poorly described. **Case report:** Female patient, 19 years old, who had three episodes of respiratory infection in the lower lobe of the right lung in a period of 18 months. Computed tomography of the chest showed obstruction of the intermediate bronchus and areas of bronchiectasis in the right lower lobe. In view of this, a lower middle bilobectomy was performed through robotic surgery. The patient was discharged from hospital three days after the operation. Immunohistochemistry revealed low-grade inflammatory rhabdomyoblastic tumor. **Conclusion:** This report described a case of a rare lung tumor, submitted to a surgical technique not yet reported for this type of pathology.

Key words: lung neoplasms; sarcoma; leiomyosarcoma; rhabdomyosarcoma.

RESUMO

Introdução: Os sarcomas primários do pulmão são tumores malignos raros com incidência estimada em torno de 0,5% de todas as neoplasias pulmonares. De suas tipificações descritas na literatura, duas especialmente, os leiomiossarcomas e os rabdomiossarcomas, apresentam semelhanças em seu perfil genético, morfológico e imuno-histoquímico, o que os levou a receberem uma mesma classificação: tumor rabdomioblástico inflamatório. Esse tipo de tumor costuma acometer tecidos moles em extremidades e tronco, e predomina em meio a homens jovens e de meia-idade. Em razão do pequeno número de casos de tumores rabdomioblásticos relatados na literatura, tanto seu diagnóstico quanto seu tratamento são pouco descritos. **Relato de caso**: Paciente, sexo feminino, 19 anos, apresentou três episódios de infecção respiratória no lobo inferior do pulmão direito em um período de 18 meses. A tomografia computadorizada do tórax evidenciou obstrução do brônquio intermediário e áreas de bronquiectasias no lobo inferior direito. Diante disso, realizou-se bilobectomia inferior-média, conduzida por cirurgia robótica. A paciente recebeu alta hospitalar três dias depois do pós-operatório. A imuno-histoquímica revelou tumor rabdomioblástico inflamatório de baixo grau. **Conclusão:** Este relato apresentou um caso de tumor pulmonar raro, abordado por uma técnica cirúrgica ainda não relatada para esse tipo de patologia.

Palavras-chave: neoplasias pulmonares; sarcoma; leiomiossarcoma; rabdomiossarcoma.

RESUMEN

Introducción: Los sarcomas pulmonares primarios son tumores malignos raros con una incidencia estimada en torno al 0,5% de todas las neoplasias pulmonares. De sus tipificaciones descritas en la literatura, dos en especial, los leiomiosarcomas y los rabdomiosarcomas presentan similitudes en su perfil genético, morfológico e inmunohistoquímico, lo que los llevó a recibir la misma clasificación: tumor rabdomioblástico inflamatorio. Este tipo de tumor suele afectar a los tejidos blandos de las extremidades y el tronco, y predomina en hombres jóvenes y de mediana edad. Debido al escaso número de casos de tumores rabdomioblásticos reportados en la literatura, tanto su diagnóstico como su tratamiento están pobremente descritos. **Informe del caso:** Paciente de sexo femenino, 19 años que consultó por tres episodios de infección respiratoria en el lóbulo inferior del pulmón derecho en un período de 18 meses. La tomografía computarizada de tórax mostró obstrucción del bronquio intermedio y áreas de bronquiectasias en el lóbulo inferior derecho. Ante esto, se realizó una bilobectomía media baja mediante cirugía robótica. La paciente recibió el alta hospitalaria tres días después de la operación. Se realizó inmunohistoquímica y se definió el diagnóstico de tumor rabdomioblástico inflamatorio de bajo grado. **Conclusión:** Este informe presenta un caso de tumor pulmonar raro, tratado mediante una técnica quirúrgica aún no reportada para este tipo de patología.

Palabras clave: neoplasias pulmonares; sarcoma; leiomiosarcoma; rabdomiosarcoma.

[1-4]Universidade Federal do Triângulo Mineiro (UFTM), Hospital de Clínicas. Uberaba (MG), Brazil.
E-mail: rgzimmer91@gmail.com. Orcid iD: https://orcid.org/0000-0002-4042-2975
E-mail: isadora.lyrio@hotmail.com. Orcid iD: https://orcid.org/0000-0003-4976-7897
E-mail: franpotros@hotmail.com. Orcid iD: https://orcid.org/0000-0001-5686-9985
E-mail: dri_amartins@hotmail.com. Orcid iD: https://orcid.org/0000-0002-0936-5474
Corresponding author: Rafael Gonçalves Zimmer. Rua João Alvares Correia, 111 – apto. 91 – Vila Mariana/Chácara Klabin. São Paulo (SP), Brazil. CEP 04115-030.
E-mail: rgzimmer91@gmail.com

INTRODUCTION

Primary lung sarcomas are rare malignant tumors with estimated incidence of 0.5% of all lung neoplasms[1,2]. Of its typifications described in the literature, leiomyosarcoma, fibrosarcoma, hemangiopericytoma and rhabdomyosarcoma are the most common intrathoracic types, with leiomyosarcoma corresponding to nearly 30% of all primary lung sarcomas[3,4]. This neoplasm can derive from structures formed by smooth muscles as the bronchus[4]. The malignancy grade should be evaluated with histopathology based in the number of mitosis, cellularity and degree of differentiation. High-grade tumors disseminate fast, are highly aggressive with extensive vascular and lymphatic invasion, while low-grade are larger, grow slowly and metastasize less frequently[5].

However, recent studies found similarities of genetic, morphologic and immunohistochemical profiles of leiomyosarcoma and rhabdomyosarcoma due to low mitotic activity, presence of necrosis and expression of typical markers of skeletal muscle[6]. Consensually, said groups of primary lung tumors should be classified as inflammatory rhabdomyosarcoma tumor[7].

The clinical presentation of these tumors are related to the growth of lung masses and the compressive effect on adjacent structures and may evolve as dyspnea, sibilance, stridor, dry or productive cough, hemoptysis and repetition pneumonia[8]. On the other hand, at the chest tomography, the class of tumors presents predominantly with regular border masses and radiologic signs associated with compressive effects as signs of air imprisonment, mucoid and consolidation impaction. This report presents a case of an inflammatory rhabdomyoblastic tumor of the lower lobe of the right lung in a young female patient.

The Institutional Review Board of "*Complexo Hospitalar HUOC/PROCAPE*" approved the case report, number 5,471,300 (CAAE: (submission for ethical review) 58713322.7.0000.5192) in compliance with Resolution 466 dated November 12, 2012[9] of the National Health Council.

CASE REPORT

Female patient, 19-years old, with history of three episodes of lower right lobe lung respiratory infections during 18 months, sought consultation with pneumologist for etiologic investigation of repetitive infections. At the consultation and while symptoms were developing, denied suggestive symptoms of consumptive syndrome and other common pathologies of the respiratory tract as dyspnea, cough and thoracic pain. No family history of neoplasms was reported and during physical exam, presented decreased vesicular breath sounds in the middle and lower third of the right hemithorax without the presence of sounds at auscultation.

A chest tomography was performed, revealing obstruction of the intermediate bronchus and areas of bronchiectasis at the right lower lobe with signs of mucoid impaction. She was referred to thoracic surgery for respiratory endoscopy with endobronchial biopsy and bronchoalveolar lavage to elucidate the etiology of the obstruction; the procedure has also shown semi-occlusive vascularized tumor at the intermediate bronchus submitted to biopsy and anatomopathology which revealed a leiomyoma. A resection of the tumor was discussed but due to the destruction of the lower lobe, it was decided to perform a middle-lower bilobectomy by robotic surgery whose surgical piece was sent for analysis; no complications occurred during the procedure and the patient was discharged asymptomatic three days later.

The histopathological exam of the surgical piece (Chart 1) revealed a fusiform cell neoplasm with elongated nuclei, eosinophilic cytoplasm and fascicular growth, with concomitant inflammatory infiltrate, including foamy histiocytes and rare Touton giant cells. Expression of smooth muscle markers was identified as well with occasional mitosis but without identification of necrosis.

After immunohistochemistry (Chart 2) and discussion among pathologists, the diagnosis of low-grade inflammatory rhabdomyoblastic smooth muscle tumor without atypia and infrequent mitosis expressing actin and desmin was reached. Due to its indolent nature associated with free margins resection and clinical evolution without complications, annual follow-up with chest tomography was the conduct agreed; one year later, a complete remission of the disease occurred with good clinical status and good residual lung capacity.

DISCUSSION

Usually, inflammatory rhabdomyoblastic tumors tend to occur in deep soft tissues of the extremities and chest predominantly in young to middle-aged males[8]. Immunohistochemically, these tumors expressed CD163-positive reactive histiocytes with phenotype of skeletal muscle and express myogenin, PAX-7 and MyoD1, typical markers of this muscle[10]. This configuration is confirmed by the genetic profile regulated by crucial genes to differentiate from skeletal muscle[2], in addition to genomic haploidization retaining chromosomes 5 and 22[7].

Its morphology is characterized by a fibrous capsule containing peripheral lymphoid aggregates, spindle-to-epithelioid cells with rhabdomyoblastic immunophenotype and a dense histiocytic infiltrate, well-circumscribed

Chart 1. Histopathological medical report

Material: Right lung	
Antibodies	**Results**
Human anti-cytokeratin	Negative
Melan-A-human	Negative
Myogenin	Negative
S100	Negative
Desmin	Positive multifocal
SOX10	Negative
Smooth muscle actin	Positive multifocal
CD246 (ALK1)	Negative
CD68	Positive
HMB-45	Negative
Diagnostic	
Low grade inflammatory smooth muscle tumor. **Note:** Fusiform cells neoplasm with elongated nuclei, eosinophilic cytoplasm and fascicular growth. Concomitant inflammatory infiltrate including foamy histiocytes and rare Touton giant cells. There is expression of markers of smooth muscle. Occasional mitosis occur. Necrosis is not identified. Recife, October 4, 2021. **Addendum:** Very rare lesion described in short casuistic in the literature. According to the current accepted terminology, the diagnosis would be low-grade inflammatory leiomyosarcoma. Recently, however, a new nomenclature was proposed, "inflammatory rhabdomyolysis tumor", since most would be clinically indolent with co-expression of skeletal muscle markers	

Chart 2. Immunohistochemistry

Material: Right lung	
Marker (antibody)	**Results**
SMA (smooth muscle actin) 1A4	Positive in areas
Desmin (intermediate filament muscle cells) (D33)	Positive in areas
S-100 Protein (polyclonal)	Negative
CD34 – hematopoietic cells and pericytes antigen (QBEnd 10)	Negative
Ki67 – Cell proliferation antigen (MIB1)	Positive in 2% of the cells
CD117/C-Kit (rabbit polyclonal)	Positive in moderate number of mastocytes
Cytokeratins 40, 48, 50 and 50,6 kDa (AE 1/AE3)	Negative
CD45 (LCA, leukocyte common antigen) (2B11+PD7/26)	Positive in moderate number of lymphocytes
CD68 (PG-M1)	Positive diffuse in histocytes
Conclusion	
Immunohistochemical profile associated with histopathology is compatible with inflammatory myofibroblastic tumor.	

with little infiltration to adjacent soft tissues[11]. These characteristics show a low growth and indolent behavior, however, a progression of these tumors to high-grade rhabdomyosarcoma[12] has been described with aggressive clinical behavior and histologically characterized by pleomorphic spindle cells with hyperchromatic nuclei and scant cytoplasm with common necrosis and desmin positivity at immunohistochemistry[13].

Due to the paucity of rhabdomyoblastic tumors reported and lack of long-term follow-up studies, its treatment and diagnosis are poorly described in the literature[4,14]. Since the diagnosis is difficult to reach based only in clinical data requiring further detailed histopathological and immunohistochemistry exams to determine the characteristics of the tumor, surgical resection is an important diagnostic option with good accuracy and successful treatment as described by Janssen et al.[15], the first authors to document a treatment for this type of lung tumor. The surgical treatment of choice for the present case because of the level of involvement was lower-middle bilobectomy by robotic surgery. To the best of the authors' knowledge so far, there are no reports in the literature about the utilization of this treatment for inflammatory rhabdomyoblastic tumor.

CONCLUSION

Pulmonary inflammatory rhabdomyoblastic tumors should be investigated by compatible clinic and suggestive radiologic findings. Due to its indolent nature they can be surgically and satisfactorily managed through robotic with oncologic safety.

ACKNOWLEDGMENT

To Dr. Paula Abreu e Lima for her involvement in the diagnosis.

CONTRIBUTIONS

All the authors contributed substantially to the study design, analysis and/or interpretation of the data, wording and/or critical review. They approved the final version to be published.

DECLARATION OF CONFLICT OF INTERESTS

There is no conflict of interests to declare.

FUNDING SOURCES

None.

REFERENCES

1. Qin BD, Jiao XD, Zang YS. Primary pulmonary leiomyosarcoma: a population-based study. Lung Cancer. 2018;116:67-72. doi: https://doi.org/10.1016/j.lungcan.2017.12.015
2. Gladish GW, Sabloff BM, Munden RF, et al. Primary thoracic sarcomas. Radiographics. 2002;22(3):621-37. doi: https://doi.org/10.1148/radiographics.22.3.g02ma17621
3. Etienne-Mastroianni B, Falchero L, Chalabreysse L, et al. Primary sarcomas of the lung: a clinicopathologic study of 12 cases. Lung Cancer. 2002;38(3):283-9. doi: https://doi.org/10.1016/s0169-5002(02)00303-3
4. Enzinger FM, Weiss SW, Meeroff NG. Tumores de tejidos blandos. Buenos Ayres: Médica Panamericana; 1985.
5. Hajdu SI. Differential diagnosis of soft tissue and bone tumors. Philadelphia: Lea & Febiger; 1986.
6. Lee JC, Li WS, Kao YC, et al. Toward a unifying entity that encompasses most, but perhaps not all, inflammatory leiomyosarcomas and histiocyte-rich rhabdomyoblastic tumors. Mod Pathol. 2021 Jul;34(7):1434-8. doi: https://doi.org/10.1038/s41379-021-00797-8
7. Folpe AL. Response to Lee et al: Toward a unifying entity that encompasses most, but perhaps not all, inflammatory leiomyosarcomas and histiocyte-rich rhabdomyoblastic tumors. Mod Pathol. 2021;34(7):1439. doi: https://doi.org/10.1038/s41379-021-00819-5
8. Cloutier JM, Charville GW, Mertens F, et al. "Inflammatory Leiomyosarcoma" and "Histiocyte-rich Rhabdomyoblastic Tumor": a clinicopathological, immunohistochemical and genetic study of 13 cases, with a proposal for reclassification as "Inflammatory Rhabdomyoblastic Tumor". Mod Pathol. 2021;34(4):758-69. doi: https://doi.org/10.1038/s41379-020-00703-8
9. Conselho Nacional de Saúde (BR). Resolução nº 466, de 12 de dezembro de 2012. Aprova as diretrizes e normas regulamentadoras de pesquisas envolvendo seres humanos. Diário Oficial da União, Brasília, DF. 2013 jun 13; Seção 1:59.
10. Michal M, Rubin BP, Kazakov DV, et al. Inflammatory leiomyosarcoma shows frequent co-expression of smooth and skeletal muscle markers supporting a primitive myogenic phenotype: a report of 9 cases with a proposal for reclassification as low-grade inflammatory myogenic tumor. Virchows Arch. 2020;477(2):219-30. doi: https://doi.org/10.1007/s00428-020-02774-z
11. Geiersbach K, Kleven DT, Blankenship HT, et al. Inflammatory rhabdomyoblastic tumor with progression to high-grade rhabdomyosarcoma. Mod Pathol. 2021;34(5):1035-6. doi: https://doi.org/10.1038/s41379-021-00791-0
12. Bourgeau M, Martinez AP. Histiocyte-rich rhabdomyoblastic tumor: a report of two cases and a review of the differential diagnoses. Virchows Arch. 2021;478(2):367-73. doi: https://doi.org/10.1007/s00428-020-02857-x. Erratum in: Virchows Arch. 2020;477(5):763. doi: https://doi.org/10.1007/s00428-020-02873-x
13. Kindblom LG, Angervall L. Nasal polyps with atypical stroma cells: a pseudosarcomatous lesion. A light and electron-microscopic and immunohistochemical investigation with implications on the type and nature of the mesenchymal cells. Acta Pathol Microbiol Immunol Scand A. 1984;92(1):65-72.
14. Kerr DA, Thompson LDR, Tafe LJ, et al. Clinicopathologic and genomic characterization of inflammatory myofibroblastic tumors of the head and neck: highlighting a novel fusion and potential diagnostic pitfall. Am J Surg Pathol. 2021;45(12):1707-19. doi: https://doi.org/10.1097/PAS.0000000000001735
15. Janssen JP, Mulder JJ, Wagenaar SS, et al. Primary sarcoma of the lung: a clinical study with long-term follow-up. Ann Thorac Surg. 1994;58(4):1151-5. doi: https://doi.org/10.1016/0003-4975(94)90476-6

Recebido em 26/9/2022
Aprovado em 1/2/2023

Scientific-Editor: Anke Bergmann. Orcid iD: https://orcid.org/0000-0002-1972-8777