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Case Report

Primary Pulmonary Chondrosarcoma: A Case Report and Literature Review

Junaid T. Yasin, Salah A. Daghlas, Aws Hamid, Ayman H. Gaballah

Department of Radiology, University of Missouri, Columbia, Missouri.

***Corresponding author:**
Dr. Ayman H. Gaballah,
Department of Radiology,
University of Missouri,
1 Hospital Drive 65212,
Columbia, Missouri.

gaballaha@health.missouri.edu

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ABSTRACT

Chondrosarcomas are tumors consisting of osseous or cartilaginous stroma. They are not an uncommon pathology; however, primary pulmonary chondrosarcomas arising in lung parenchyma are extremely rare, with few cases published in literature. Herein, we present a case with biopsy-proven primary pulmonary chondrosarcoma after exclusion of primary origin elsewhere. In the case presented in this report, we demonstrate the clinical presentations, pulmonary function tests, and the radiological findings of this rare tumor in a young male patient. Further, we present a brief review of existing literature for patients with similar pathology.

Keywords: Primary, Pulmonary, Chondrosarcoma

INTRODUCTION

Chondrosarcomas of primary lung parenchymal origin are an extremely rare pathology with most descriptions coming from a series of case reports and small case series. These tumors can present as centrally located, parenchymal masses. Diagnosis requires extensive workup to exclude alternative etiology and to confirm diagnosis. To date, 28 cases, not including the current study, of this pathology have been reported. Herein, we present a case with biopsy-proven primary pulmonary chondrosarcoma and a brief literature review of similar cases.

CASE REPORT

A 23-year-old Caucasian male presented to a cardiothoracic surgery clinic for follow-up of a lung mass. The mass was identified on chest X-ray and computed tomography (CT) chest studies that were performed for the evaluation of a non-productive cough, fever, and 10 pounds weight loss.

On physical examination, the patient exhibited mild expiratory wheezing bilaterally. Pertinent laboratory results showed hypercalcemia. Pulmonary function tests were indicative of an obstructive lung disease; however, a bronchoscopy examination was unremarkable. Chest X-ray and contrast-enhanced CT were performed, revealing the presence of a large mass with several intrinsic calcifications in the upper lobe of the right lung along the paramediastinum [Figures 1 and 2]. An ultrasound-guided procedure was conducted to localize the lesion and biopsy the surrounding lymph nodes and was indicative of low-grade cartilaginous neoplasm, presumed to be either chondroma or teratoma.

An exploratory right video-assisted thoracoscopic surgery was performed as primary treatment. The operation revealed an intraparenchymal central mass in the right upper lobe [Figure 3]

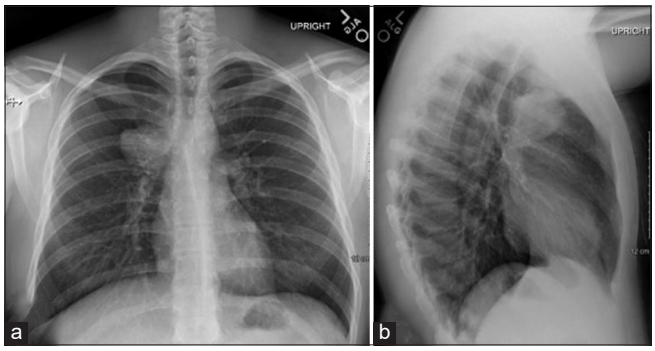

Figure 1: Chest radiograph frontal (a) and lateral (b) views of a 23-year-old male demonstrate rounded mass-like opacity (arrows) along the superior right paramediastinal border in the right upper lobe.

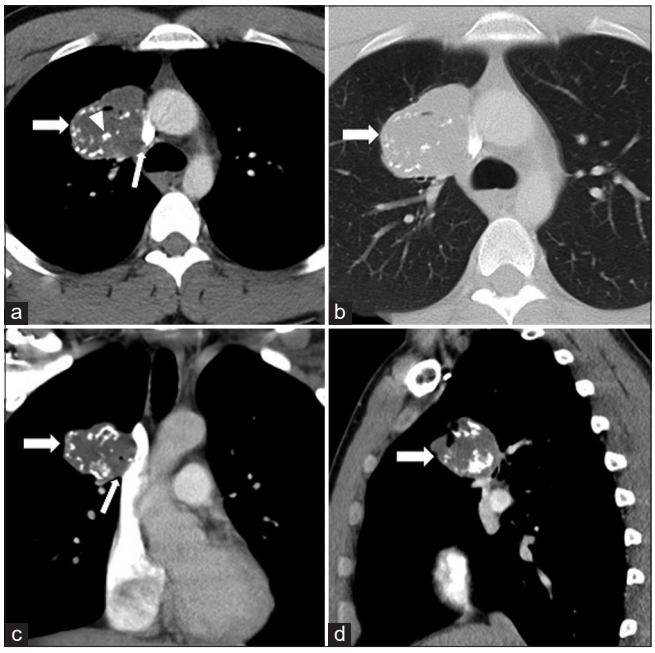

Figure 2: Contrast-enhanced computed tomography chest in axial (a and b), coronal (c), and sagittal (d) views of a 23-year-old male revealed a large soft tissue mass in the right upper lobe (thick arrows) along the right paramediastinal border abutting the adjacent mediastinal vascular structures (thin arrows), with multiple intrinsic calcifications (arrowhead). Surgery resection of the mass was performed and the lesion proved to be a low-grade primary lung chondrosarcoma.

adherent to the mediastinum adjusted to the main pulmonary artery, thus necessitating a lobectomy. Postsurgical CT scan of the chest and bone scan using Tc-99m diphosphate revealed no residual tumor or secondary lesions, respectively.

A histopathological review revealed a well-demarcated lobulated, rubbery, and cartilaginous tumor with a focal necrotic appearance and calcifications. The lesion had stromal hyalinization and prominent stromal vessels with mild atypia. Immunostains for pankeratin, PLAG1, and HMGA2 were negative. The presence of cellular crowding and binuclear chondrocytes [Figure 3] supported the final interpretation of the lesion as a low-grade (Grade 1) pulmonary chondrosarcoma.

DISCUSSION

Chondrosarcomas of primary lung parenchymal origin are extremely infrequent with most descriptions of the pathology coming almost exclusively from case reports, as depicted in Table 1. A literature search of PubMed using the query [(pulmonary OR lung) AND chondrosarcoma AND primary] yielded 14 studies with full-text available or sufficient information available in the abstract. Ten additional studies were found through a bibliographic review. Cases that did not originate from lung parenchymal tissue were excluded from the study. Such excluded cases include pulmonary artery, costal, and bronchial chondrosarcomas.

The incidence of primary pulmonary chondrosarcoma is difficult to estimate as this pathology is frequently misdiagnosed, requiring an extensive clinical history and diagnostic workup to exclude alternative primary origin; thus, it may be underreported in the literature. Patients with pulmonary chondrosarcomas frequently present with symptomatic pulmonary obstruction, dyspnea, and chest pain. The tumors are often centrally located and present with sharp borders. CT scans that demonstrate a well-demarcated, centrally located mass with calcifications may be indicative of pulmonary chondrosarcoma and this pathology should be included in the differential diagnosis of patients with similar presentations to ours [Figures 1 and 2].

The presence of calcifications is disputed as a case report claimed calcifications are classically present,[1] whereas other case reports indicate an absence of calcifications.[2] Histologically, pulmonary chondrosarcomas are characterized by ill-defined lobules containing differentiated hyaline cartilage with plump nuclei, binucleation, hyperchromasia, and coarse chromatin.[2] The presence of basophilic myxoid stroma with focal areas of hyaline cartilage may also be seen.[2] Immunohistochemical studies of the tumors are negative for keratin, CD31, and desmin and are S-100 positive.

To diagnose a primary pulmonary chondrosarcoma, a detailed clinical history and appropriate imaging are essential to exclude the following criteria: Prior history of skeletal neoplasms, a thoracic cage neoplasm, limb amputation, and teratoma.[2] Furthermore, the presence of the Carney triad (pulmonary chondroma along with gastrointestinal stromal tumor and extra-adrenal paragangliomas) should be excluded.[3]

Regarding treatment, complete surgical resection of the tumor is recommended as malignant cartilage

Table 1: Literature review of primary pulmonary chondrosarcoma cases

Study primary author, year	Age/ sex	Presentation	Location	Type	Treatment	Outcome
Balanzá, 2016[5]	69/M	Hemoptysis	RLL	Myxoid	Thoracotomy	Complete surgical resection. No evidence of recurrence for 6 months of follow-up
Jiang, 2016[4]	59/M	Hemoptysis, cough, dyspnea	RUL	Myxoid	Surgical removal, ifosfamide, doxorubicin	Distal metastases, Ongoing treatment
Endicott, 2015[6]	69/M	Acute-onset dyspnea	LUL	Myxoid	Sublobar resection	Complete surgical resection. No evidence of recurrence for 6 months of follow-up
Wang, 2014[1]	52/F	Intermittent cough, dyspnea	LUL	Dedifferentiated	Thoracotomy	Patient is alive after 36 months of follow-up
Mei, 2013[7]	20/F	Non-productive cough, chest pain	LLL	Mesenchymal	Pneumonectomy	Lost to follow-up
Kalhor, 2011[2]	1. 69/F	1. NA	1. NA	1. Hyaline	1. NA	1. NA
	2. 69/M	2. NA	2. NA	2. Myxoid	2. NA	2. Decreased, 45 days post-operative
	3. 53/M	3. NA	3. NA	3. Myxoid	3. NA	3. NA
	4. 51/F	4. NA	4. NA	4. Hyaline	4. NA	4. Patient is alive after 36 months of follow-up
Li, 2011[8]	51/F	Severe anemia	LUL	Myxoid	Thoracotomy, left upper lobectomy with systemic lymph node dissection	Smooth recovery, discharged in good condition without anemia after 2 weeks, follow-up every 3 months, no abnormalities 32 months post-operative
Boueiz, 2009[9]	57/F	Exertional dyspnea	RLL, posterior mediastinum	Myxoid	Thoracotomy, complete debulking, pleurodesis	Uneventful post-operative course, no recurrence of pleural effusion, declined adjuvant chemotherapy, and lost to follow-up
Shah, 2007[10]	60/M	Dry cough	LLL	Hyaline	Thoracotomy, recurrence/ metastasis treated with radiotherapy, ifosfamide, doxorubicin, surgical resection	Full recovery after treatment of distal metastases.
Steurer, 2007[11]	49/M	Chest pain, fatigue, weight loss	LUL	Dedifferentiated	Doxorubicin, ifosfamide, surgical resection	No evidence of further tumor lesions after 30 months of follow-up
Ichimura, 2005[12]	35/M	Incidental	LLL	Myxoid	VATS	Patient is alive after 72 months of follow-up
Huang, 2002[13]	40/M	Incidental	LUL	Mesenchymal	Thoracotomy	No evidence of recurrence/metastasis after 12 months of follow-up
Parker, 1996[14]	25/F	Abdominal pain	LUL	Hyaline	Surgical excision	Patient is alive after 14 months of follow-up
Hayashi, 1993[15]	73/M	Incidental	RML	Hyaline	Surgical resection	Metastases to skull, kidneys
Kurotaki, 1992[16]	45/F	NA	RLL	Mesenchymal	Lobectomy	Patient is alive after 12 months of follow-up
Stanfield, 1991[17]	78/M	Dull pain in the left chest and shoulder	LUL	Mesenchymal	Thoracotomy, resection of LUL, 2nd and 3rd ribs, and adjacent soft tissue	Normal post-operative recovery with a significant reduction in cancer-related pain, tumor identified in a subclavian nodule of the chest wall adjacent to surgical incision 2 months later
Watanabe, 1990[18]	67/M	Fever, cough	RLL	Myxoid/Hyaline	Lobectomy	No signs of recurrence after 44 months

(*Contd...*)

Table 1: (*Continued*)

Study primary author, year	Age/ sex	Presentation	Location	Type	Treatment	Outcome
Jazy, 1984[19]	1. 61/F 2. 70/M	1. Non-productive cough, right-sided pleuritic chest pain. 2. Left-sided chest pain, let scapular pain, left arm pain, left Horner syndrome, signs of cord compression with paraparesis and altered sensorium	1. RLL, lower posterior mediastinum. 2. LUL, extending into epidural space at the level of T2	1. NA 2. NA	1. Exploratory thoracotomy, radiation, incomplete resection, cyclophosphamide, doxorubicin 2. Decompressive laminectomy ×2, radiation, physical rehabilitation	1. Recurrence 5 months post-operative. Died of cardiac causes 4 months later 2. Progressive debilitation, death within 7 months of presentation.
Sun, 1982[20]	31/F	Wheezing, cough	RLL	Myxoid/Hyaline	Pneumectomy, radiotherapy	Deceased
Morgan, 1972[21]	23/F	Incidental	RLL	Myxoid	Thoracotomy, local excision	Discharged on post-operative day 9. Doing very well on follow-up 15 months post-operative.
Rees, 1970[22]	64/M	Cough with purulent sputum	LUL	Hyaline	Lobectomy	Patient is alive after 48 months of follow-up
Smith, 1960[23]	53/F	Chronic fatigue, frontal headache, palpitations, nocturia, ankle swelling	Right lung	NA	NA	NA
Lowell, 1949[24]	41/F	Fatigue, anorexia, lower extremity edema, weight loss, dyspnea, tachycardia, palpitations	LLL	Mesenchymal	No treatment, patient decided to forgo thoracotomy	Patient died of expanding the pneumothorax
Greenspan, 1933[25]	35/F	Chest pain	LUL	NA	None (diagnosed at autopsy)	Deceased

M: Male, F: Female, NA: Not available, RUL: Right upper lobe, RML: Right middle lobe, RLL: Right lower lobe, LUL: Left upper lobe, LLL: Left lower lobe, VATS: Video-assisted thoracoscopic surgery

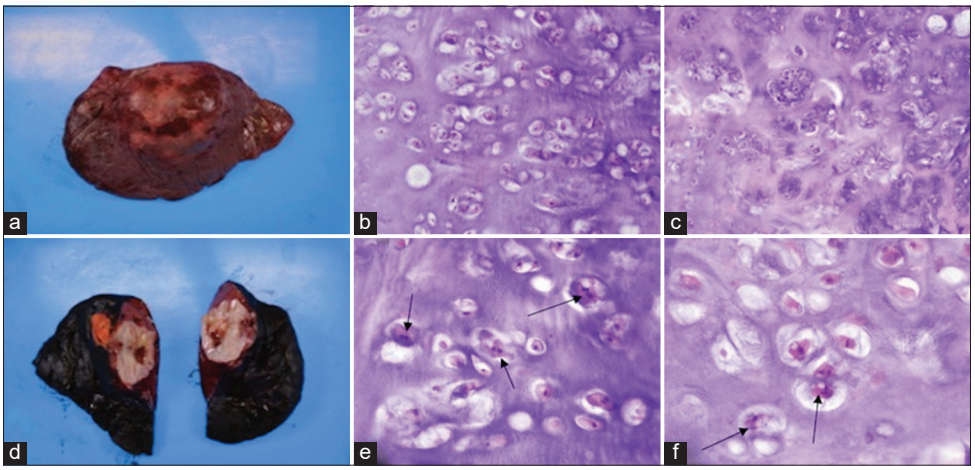

Figure 3: Resected 5 cm primary chondrosarcoma from a 23-year-old male from the upper lobe of right lung through exploratory video-assisted thoracoscopic surgery, (a and b) gross appearance of resected mass, (c-f) histopathological findings of primary pulmonary chondrosarcoma with H and E stain, arrows point toward binuclear chondrocytes with nuclear crowding, which suggest the presence of a low-grade chondrosarcoma.

may be observed with mixed epithelial/mesenchymal neoplasms.[2] Complete resection of the tumor may also prevent misdiagnosis based on improper biopsy specimen as teratomas may resemble pulmonary chondrosarcoma from biopsy location inaccuracy.[4]

A comprehensive review of existing literature on this tumor reveals that these tumors, when treated early, generally respond well to surgical treatment. These tumors have been known to metastasize to distal sites, such as the skull and kidneys, where adjuvant chemotherapy may be more beneficial. Further, investigation and study of the radiological findings in cases of primary pulmonary chondrosarcomas and reported outcomes are needed to establish reliable diagnostic criteria and treatment recommendations.

CONCLUSION

Primary pulmonary chondrosarcomas are a rare form of chondrosarcomas as seen in the dearth of cases reported in literature. We report a case which was prompted by the identification of a lung mass when the patient came in for chest X-ray and chest CT for fever, non-productive cough, and weight loss. Diagnostic workup of exclusion and histological analysis confirmed its origin from lung parenchymal tissue. It is necessary to pursue further study of radiological findings of primary pulmonary chondrosarcomas to develop guidelines and criteria to reliably establish its diagnosis.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Wang XY, Hu S, Guo LC, Chen J, Zhu M, Yao FR, *et al.* Primary chondrosarcoma presenting as an intrathoracic mass: A report of three cases. Oncol Lett 2014;8:1151-4.
2. Kalhor N, Suster S, Moran CA. Primary pulmonary chondrosarcomas: A clinicopathologic study of 4 cases. Hum Pathol 2011;42:1629-34.
3. Carney JA, Sheps SG, Go VL, Gordon H. The triad of gastric leiomyosarcoma, functioning extra-adrenal paraganglioma and pulmonary chondroma. N Engl J Med 1977;296:1517-8.
4. Jiang J, Shen Q, Ding W, Zhou J. Primary pulmonary chondrosarcoma and a fast-growing mass that accidentally mimicked teratoma. J Thorac Dis 2016;8:E947-51.
5. Balanzá R, Arrangoiz R, Cordera F, Muñoz M, Luque-de-León E, Moreno E, *et al.* Pulmonary extraskeletal myxoid chondrosarcoma: A case report and literature review. Int J Surg Case Rep 2016;27:96-101.
6. Endicott KM, Barak S, Mortman KD. A rare case of extraskeletal pulmonary myxoid chondrosarcoma. J Solid Tumors 2015;6:18.
7. Mei B, Lai YL, He GJ, Shou YN, Liu J. Giant primary mesenchymal chondrosarcoma of the lung: Case report and review of literature. Ann Thorac Cardiovasc Surg 2013;19:481-4.
8. Li XF, Zhou HB, Zhao XL, Dai F, Li T, Wang L, *et al.* Primary dedifferentiated chondrosarcoma of lung: Report of a case. Zhonghua Bing Li Xue Za Zhi 2011;40:127-8.
9. Boueiz A, Abougergi MS, Noujeim C, Bousamra A, Sfeir P, Zaatari G, *et al.* Primary dedifferentiated chondrosarcoma of

the lung. South Med J 2009;102:861-3.
10. Shah ND, Diwanji SR. Primary chondrosarcoma of the lung with cutaneous and skeletal metastases. Singapore Med J 2007;48:e196-9.
11. Steurer S, Huber M, Lintner F. Dedifferentiated chondrosarcoma of the lung: Case report and review of the literature. Clin Lung Cancer 2007;8:439-42.
12. Ichimura H, Endo K, Ishikawa S, Yamamoto T, Onizuka M, Sakakibara Y. Primary chondrosarcoma of the lung recognized as a long-standing solitary nodule prior to resection. Jpn J Thorac Cardiovasc Surg 2005;53:106-8.
13. Huang HY, Hsieh MJ, Chen WJ, Ko SF, Yang BY, Huang SC. Primary mesenchymal chondrosarcoma of the lung. Ann Thorac Surg 2002;73:1960-2.
14. Parker LA, Molina PL, Bignault AG, Fidler ME. Primary pulmonary chondrosarcoma mimicking bronchogenic cyst on CT and MRI. Clin Imaging 1996;20:181-3.
15. Hayashi T, Tsuda N, Iseki M, Kishikawa M, Shinozaki T, Hasumoto M. Primary chondrosarcoma of the lung. A clinicopathologic study. Cancer 1993;72:69-74.
16. Kurotaki H, Tateoka H, Takeuchi M, Yagihashi S, Kamata Y, Nagai K. Primary mesenchymal chondrosarcoma of the lung. A case report with immunohistochemical and ultrastructural studies. Acta Pathol Jpn 1992;42:364-71.
17. Stanfield BL, Powers CN, Desch CE, Brooks JW, Frable WJ. Fine-needle aspiration cytology of an unusual primary lung tumor, chondrosarcoma: Case report. Diagn Cytopathol 1991;7:423-6.
18. Watanabe A, Ito M, Nomura F, Saka H, Sakai S, Shimokata K. Primary chondrosarcoma of the lung--a case report with immunohistochemical study. Jpn J Med 1990;29:616-9.
19. Jazy FK, Cormier WJ, Panke TW, Shehata WM, Amongero FJ. Primary chondrosarcoma of the lung. A report of two cases. Clin Oncol 1984;10:273-9.
20. Sun CC, Kroll M, Miller JE. Primary chondrosarcoma of the lung. Cancer 1982;50:1864-6.
21. Morgan AD, Salama FD. Primary chondrosarcoma of the lung. Case report and review of the literature. J Thorac Cardiovasc Surg 1972;64:460-6.
22. Rees GM. Primary chondrosarcoma of lung. Thorax 1970;25:366-71.
23. Smith EA, Cohen RV, Peale AR. Primary chondrosarcoma of the lung. Ann Intern Med 1960;53:838-46.
24. Lowell LM, Tuhy JE. Primary chondrosarcoma of the lung. J Thorac Surg 1949;18:476-83.
25. Greenspan EB. Primary osteoid chondrosarcoma of the lung: Report of a case. Am J Cancer 1933;18:603-9.

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