Evaluation and Surgical Excision of an Intraluminal Primary Pulmonary Myxoid Sarcoma of the Pulmonary Artery

Cynthia Harrison, MD,[1] Caroline Snyder,[1] Staci Chen, MD,[1] Andrew Vaynblat,[1] Jose G. Mantilla, MD,[2] Les James, MD,[1] Mikhail Vaynblat, MD[1]
[1]Cardiothoracic Surgery, NYU Grossman School of Medicine [2]Pathology, NYU Grossman School of Medicine

NYU Grossman School of Medicine
Department of Cardiothoracic Surgery
NYU Langone Health

INTRODUCTION

Primary pulmonary myxoid sarcomas (PPMS) are exceedingly rare, low-grade thoracic tumors. First described in 1999 and classified by the WHO in 2015, approximately 40 cases have been reported worldwide.

PPMS primarily arise within the lung parenchyma; only one prior case (Opitz et al.) describes a solitary intraluminal PPMS of the pulmonary artery (PA).

Epidemiology & Presentation
- Median age at diagnosis: 44 years (range 21–80)
- Symptoms: dyspnea, hemoptysis, chest pain, weight loss
- Many patients are identified incidentally on imaging

Histopathology
Monotonous spindle cells in a loose myxoid stroma. IHC typically shows SMA reactivity; ALK expression variably present. Defined by recurrent EWSR1::CREB1 fusion gene.

Management
Surgical excision is the mainstay of treatment with good long-term outcomes. Metastasis reported in 14.7% of cases; one fatality from cerebral metastasis.

This report presents the second case of intraluminal PA PPMS and the first detailed description of its operative management.

SURGICAL TECHNIQUE

Elective median sternotomy performed 2 months after initial ED presentation.

Approach
Median sternotomy. Small thymic lymph node excised and sent to pathology.

Cardiopulmonary Bypass
ACT-guided heparinization; ascending aorta and right atrium cannulated; antegrade and retrograde cardioplegia placed. Normothermic CPB — no cardiac arrest.

Tumor Resection
- Incision of proximal MPA revealed a large gelatinous pedunculated mass extending into the RPA; bifurcation and left PA were tumor-free.
- RPA dissected from posterior aorta and under SVC. Two arteriotomies connected for complete tumor visualization. Stalk (1.5 cm) excised with Metzenbaum scissors including underlying PA wall. MPA/RPA reconstructed with bovine pericardial patch (Cryolife 6×8 cm), 4-0 Prolene. CPB 89 min; EBL minimal.

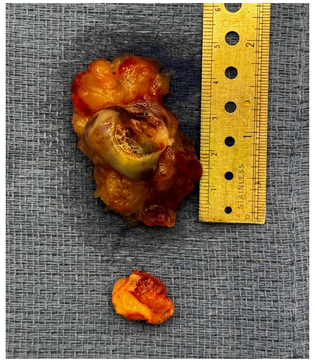

Figure 1. Excised Mass

PATHOLOGY & OUTCOMES

Final Pathology: Primary Pulmonary Myxoid Sarcoma

Histology
- H&E: Cytologically bland, monotonous spindle cells in cords and short fascicles within a loose myxoid stroma
- Mitotic activity: Sparse; no tumor necrosis

Immunohistochemistry
- SMA: Diffuse positivity
- ALK (clone D5F3): Strong, diffuse positivity (no ALK gene rearrangement)

Molecular
- NGS: EWSR1 (NM_005243.4, exon 7)::CREB1 (NM_004379.5, exon 6) fusion confirmed

Post-Operative Course
- Uncomplicated post-operative course
- Discharged home on POD #2

Follow-Up
- **2-week follow-up**: Complete resolution of chest pain, dyspnea, cough, and hemoptysis
- **Surveillance**: CT imaging every 3 months given low-grade tumor, complete resection, and lack of evidence-based adjuvant therapy
- Oncology follow-up for monitoring of FDG-avid pulmonary opacities and recurrence

CASE PRESENTATION

Patient
28-year-old woman, former smoker. PMH: Wilms' tumor, status post left nephrectomy in childhood.

Presentation
2-month history of right pleuritic chest pain, night sweats, and chills; 2-week history of hemoptysis, dyspnea, and fatigue. Initially evaluated for pulmonary embolism.

Diagnostic Workup
- **CTA chest**: 4.0 × 1.8 × 2.5 cm hypodense lobulated filling defect extending from the mid main pulmonary artery (MPA) into the right pulmonary artery (RPA); peripheral right lobe consolidations and nodules *(Fig 2)*
- **Echocardiogram**: Mild pulmonary hypertension (PASP 37 mmHg), otherwise normal
- **MRI**: 4 cm enhancing MPA/RPA mass consistent with neoplasm; multiple hemorrhagic infarcts in right lung
- **PET-CT**: FDG-avid PA mass (SUV 6.4); peripheral opacities (SUV 2.0–2.3); mediastinal/hilar nodes (SUV 2.3) *(Fig 2)*

Case Conference & Biopsy
Discussed at cardiothoracic surgery case conference and thoracic tumor board. EBUS-TBNA performed of the mass and mediastinal nodes.
- **Biopsy**: Spindle cells in myxoid stroma; diffuse SMA and ALK (D5F3) positivity
- **NGS**: EWSR1 (exon 7)::CREB1 (exon 6) fusion confirmed
- **Nodes** (stations 7, 4R, 11R): Negative for metastasis
- **Pre-op CTA**: Mass enlarged to 4.7 × 2.0 cm

Decision: Proceed with surgical excision.

FIGURES

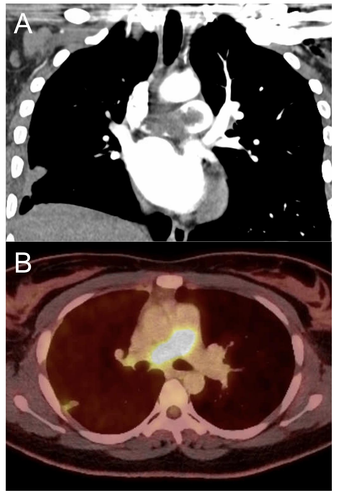

Figure 1. Preoperative Imaging
A) Axial CTA: 4.7 × 2.0 cm hypodense lobulated filling defect extending from the mid MPA into the RPA. B) Axial PET-CT: FDG-avid PA mass (SUV 6.4, red arrow) and FDG-avid posterolateral RLL opacity (SUV 2.3, white arrow).

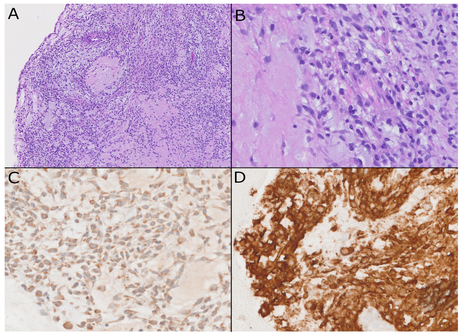

Figure 2. Pathology
(A, B). H&E: Bland spindle cells in myxoid stroma, sparse mitoses. (C) IHC SMA: Diffuse reactivity. (D) IHC ALK (D5F3): Strong, diffuse positivity.

DISCUSSION & CONCLUSIONS

PPMS & Angiomatoid Fibrous Histiocytoma
PPMS shares the EWSR1::CREB1 rearrangement with AFH, suggesting a shared biological spectrum. Diffuse ALK positivity (without ALK rearrangement) may mimic inflammatory myofibroblastic tumor on small biopsy.

Diagnostic Approach
CT, PET-CT, and EBUS-TBNA are effective for evaluating intraluminal PA masses and differentiating PPMS from PA intimal sarcoma, angiosarcoma, and inflammatory myofibroblastic tumor.

Surgical Considerations
- Pedunculated intraluminal tumor allowed complete excision via arteriotomy + patch repair — avoiding pneumonectomy/lobectomy
- Friable, gelatinous character requires careful dissection to prevent fragmentation
- Minimal PA wall involvement enabled curative resection

Conclusions
Only the 2nd reported intraluminal PA PPMS; the 1st with detailed operative description. Complete surgical excision is feasible with excellent short-term outcomes.

REFERENCES

1. Nicholson AG, et al. Histopathology 1999;35(4):313–8.
2. Travis WD, et al. J Thorac Oncol 2015;10(9):1240–1242.
3. Miao X, et al. J Cancer Res Clin Oncol 2024;150(3):108.
4. Kerper AL, et al. Am J Surg Pathol 2024;48(5):562–569.
5. Thway K, et al. Am J Surg Pathol 2011;35(11):1722–32.
6. Opitz I, et al. Histopathology 2019;74(3):526–530.
7. Shingyoji M, et al. Ann Thorac Surg 2013;96(2):e33–e35.
8. Cheah AL, et al. Am J Surg Pathol 2019;43(1):93–101.

Corresponding Author: Mikhail Vaynblat, MD | mikhail.vaynblat@nyulangone.org