



# Myxopapillary Ependymoma Metastasis Mimicking Pulmonary Embolism: An Illustrative Case

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## Abstract

Myxopapillary ependymomas (MPEs) are rare spinal cord tumors with low rates of metastasis outside of the neuraxis. Gross total resection of MPEs can significantly improve progression-free survival; however, adjunctive treatment remains unstandardized. A 29-year-old female with a history of spina bifida occulta surgical correction and lower back pain presented with dyspnea and tachycardia. A large pulmonary artery mass was discovered consistent with pulmonary thromboembolism. It was subsequently determined to be an intravascular metastasis secondary to sacral MPE. Standardization of MPE treatment and clinical suspicion of spinal neoplasm in the setting of chronic back pain with undetermined origin are of value.

## Keywords

- ▶ myxopapillary ependymoma
- ▶ metastasis
- ▶ pulmonary embolism

## Introduction

Myxopapillary ependymomas (MPEs) are rare neoplastic tumors arising from ependymal glial cells of the spinal cord; incidence is 0.5 to 0.8 per 100,000 people/year.<sup>1</sup> MPEs are the most common tumor of the cauda equina region, and the average age of onset is in the third or fourth decade of life.<sup>1</sup> MPEs are considered benign; however, they were upgraded to Grade II neoplasms in the 2021 World Health Organization (WHO) classification of Tumors of the Central Nervous System.<sup>2</sup> Local recurrence occurs commonly following surgical resection, even after decade-long quiescence, and long-term monitoring and care are recommended.<sup>1,3</sup> Treatment for MPE is not standardized. In general, it is thought that gross total resection is an optimal course of treatment; however, the efficacy of adjuvant therapy remains controversial.<sup>1,4</sup> Total tumor resection with maintenance of capsular margins is

associated with a reduced recurrence rate compared with subtotal resection with violated margins.<sup>5</sup> Many studies demonstrate that adjuvant radiotherapy following resection is associated with increased progression-free survival (PFS).<sup>6–8</sup> However, other studies do not report significant differences in 5-year PFS, recurrence rate, or overall survival benefit with adjunctive radiotherapy or chemotherapy compared with surgical resection alone.<sup>5,7–9</sup>

In addition, MPEs rarely metastasize outside of the cerebrospinal axis, and pediatric patients show higher rates of central nervous system dissemination and extramedullary presentation than adults.<sup>1,10–12</sup> A few published cases that show MPEs spread outside the neuroaxis into other organ systems, most commonly to the lymphatic system.<sup>13,14</sup>

Here we describe a case in which a primary spinal MPE metastasized to the pleura, lung parenchyma, and intravascular space and mimicked the presentation of pulmonary embolism.

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### Patient History and Presentation

A 29-year-old female with a history of spina bifida occulta that was corrected during childhood presented initially for lower back pain, which was attributed to a recent pregnancy and noneventful vaginal delivery. At initial visit, the patient described significant sacrum pain radiating to the right buttock. Pain was reportedly somewhat responsive to osteopathic manipulative therapy (OMT) and was determined to be musculoskeletal in nature. Subsequently, the patient was discharged with a physical therapy consultation, recommendation of rest, and nonsteroidal anti-inflammatory drugs (NSAIDs) for pain relief.

Eight months following the initial presentation, the patient presented again for worsening sacral pain and tenderness and additionally reported 3 days of urinary incontinence and numbness in her right thigh and groin. At this time, symptoms were no longer responsive to physical therapy, OMT, or NSAIDs. Lumbar spine, sacrum, and coccyx X-rays revealed no osseous abnormality, though a small calcification in the right pelvis was noted. The following week the patient consulted with a sports medicine physician where she noted new onset constipation; she was prescribed a suppository and additionally started on baclofen for ongoing pain.

One month later, the patient presented to urgent care with dyspnea, left-sided chest pain, and heart palpitations. She was found to be tachycardic (131 beats per minute [BPM]), normotensive with normal respiratory rate (20 breaths per minute) and oxygen saturation (SpO<sub>2</sub>: 98%). Wheezing could be heard in the right upper lung field. Chest X-ray revealed a 3.0 cm right middle lobe pulmonary nodule (►Fig. 1). D-dimer (1.48 µg fibrinogen equivalent unit /mL), C-reactive protein (1.30 mg/dL), serum protein (9.2 g/dL), serum globulin (4.2 g/dL), and white blood cell count ( $12.6 \times 10^3/\mu\text{L}$ ) values were elevated. Considering the possibility of a thromboembolic event, the patient was advised to transfer immediately to the emergency department (ED). At triage, she was found to be tachycardic (130 BPM), normotensive with a lowered SpO<sub>2</sub> (94%). Upon admission, a contrasted chest, abdomen, and pelvis computed tomography (CT) imaging study was performed in which a large, invasive sacral mass was discovered.

After a 2-day admission, the patient was discharged on anticoagulation (apixaban) therapy. Meanwhile, further oncological testing was performed. Following CT-guided biopsy of the sacral mass (►Fig. 2A), the patient was diagnosed with an epidural MPE WHO Grade II. To confirm extraspinal metastasis, spine positron emission tomography (PET) scan and a magnetic resonance imaging scan and lung biopsy were scheduled.

### Imaging

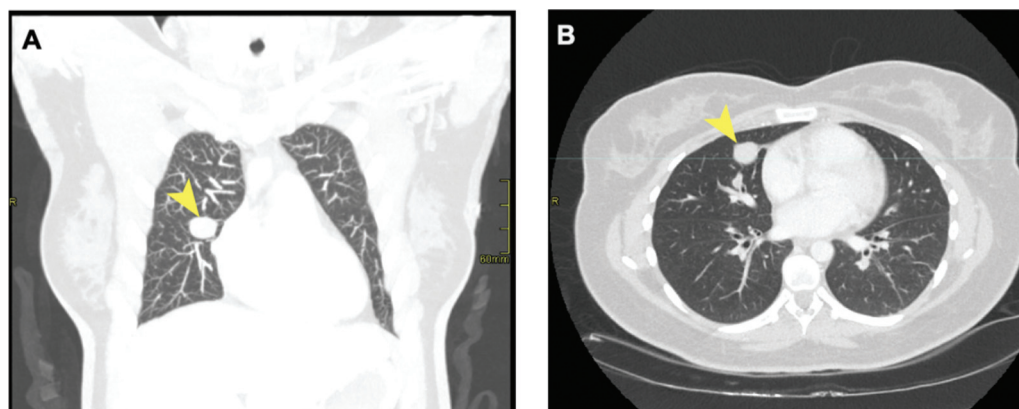
Thorax CT revealed large bilateral pulmonary emboli with multiple pleural tumor deposits. Spine and pelvis CT revealed a large sacral mass with soft tissue involvement and extension superiorly to L4–5 level (►Fig. 3). Bilateral neoplastic infiltration of piriformis muscle was also observed (►Fig. 3B). A whole-body scan identified an enhancing lobular mass of the sacrum. A 2.4 cm mass located in the right internal mammary lymph nodes was thought to represent adenopathy.

### Histopathology

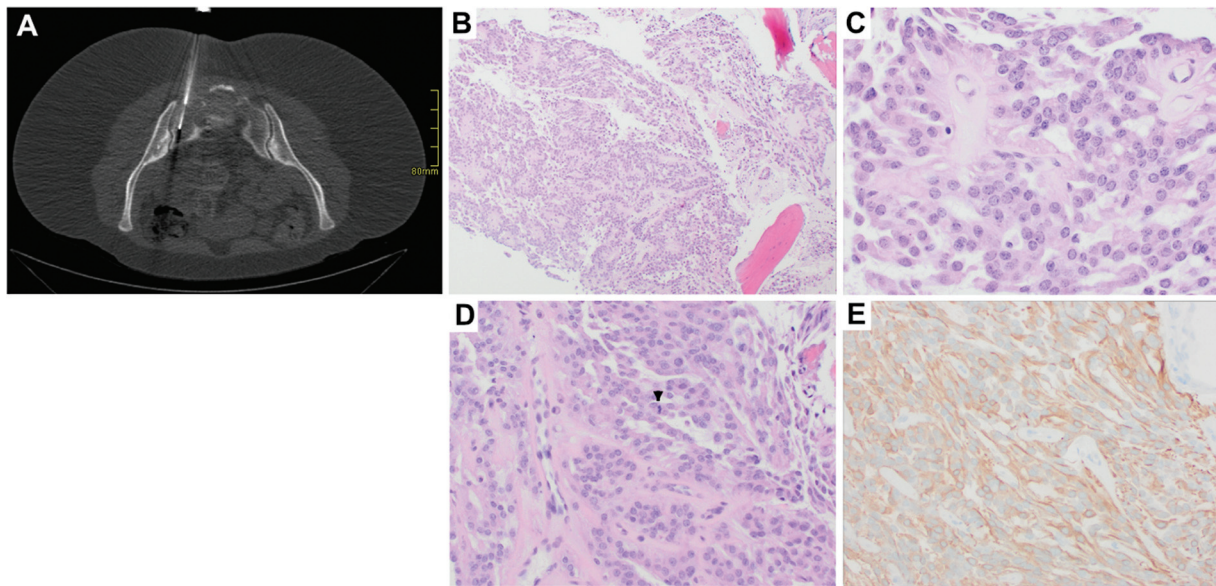
A sacrum CT-guided biopsy was conducted following the patient's first admission (►Fig. 2A). Hematoxylin and eosin staining demonstrated a proliferation of cells with round to oval nuclei with focal radial arrangement around blood vessels, on a background of microcystic myxoid material (►Fig. 2B, C). Mitotic activity was conspicuous, with up to 4 mitotic figures/mm<sup>2</sup> (►Fig. 2D). The lesion demonstrated diffuse immunoreactivity for glial fibrillary acidic protein with abundant glial fibrillary processes (►Fig. 2E). No high-grade features, such as Ki67, were found.

### Clinical Course

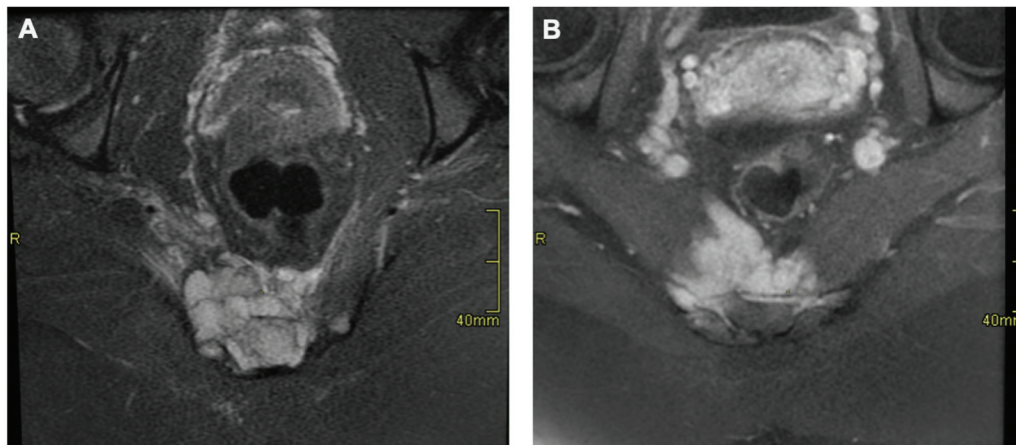
Following initial discharge, the patient was referred to Oregon Health & Science University (Neurological Surgery). Detailed history and physical exam were noted. Discussions with the patient were centered around information regarding total resection of the sacral tumor that would result in a near-complete sacrectomy with a risk to bowel and bladder function. A limited resection was also discussed with the patient. The case was presented at a



**Fig. 1** Computed tomography imaging at initial presentation. (A) Coronal and (B) axial computed tomography chest studies showing prominent 2.6 cm pulmonary nodule (yellow arrowhead) in the right middle lobe at initial emergent presentation.



**Fig. 2** Histopathology of sacral biopsy. (A) Axial computed tomography imaging demonstrating posterior course of sacral biopsy probe. (B) Sacral biopsy. Tumor infiltrating between bony trabeculae, x40. (C) Monomorphic cells with round nuclear contours, radially arranged around blood vessels, with perivascular fibrillar processes, x200. (D) Mitotic activity was conspicuous (arrowhead, mitotic figure), x100. (E) The tumor was diffusely positive for glial fibrillary acidic protein, with abundant fibrillary, x200.



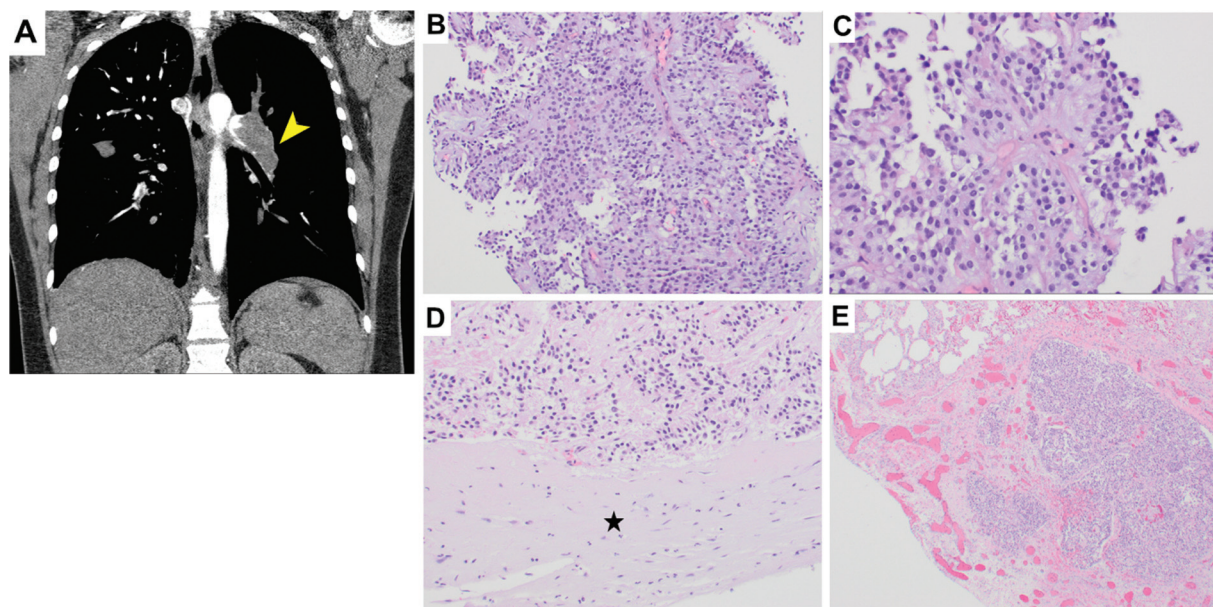
**Fig. 3** Magnetic resonance imaging of sacral myxopapillary ependymoma. Axial imaging of (A) T2-weighted and (B) T1-weighted with contrast and fat sat demonstrating T2 hyperintense soft tissue mass centered on the sacrum and coccyx measuring 8.5 × 5.9 × 4.8 cm.

multidisciplinary tumor board conference with a tentative plan for surgery made pending repeat imaging and pleural biopsy. A subsequent pleural biopsy demonstrated near identical morphologic features to the original sacral biopsy (► Figs. 2C, 4B, C).

Unfortunately, the patient was unable to complete a pre-operative PET scan and coronavirus disease 2019 test 2 weeks later, due to an episode of dizziness, shortness of breath, and loss of consciousness. The patient was transferred to the ED and found to be in cardiogenic shock from a suspected recurrent pulmonary embolism. Chest CT showed an enlarged saddle pulmonary embolism with complete obstruction of the left pulmonary artery and near complete obstruction of the right pulmonary artery (► Fig. 4A). Given concern for bilateral pulmonary embolism, she was administered tenecteplase (intravenous 50mg) and started on heparin drip with

persistent hemodynamic instability that required vasopressor support. Bilateral pulmonary artery emboli were found, mechanical suction thrombectomy was deemed technically unfeasible, and bilateral EKOS catheters (Boston Scientific Corporation, Marlborough, Massachusetts, United States) were placed for targeted thrombolysis. In addition, there were complete bilateral thrombotic occlusion of the external iliac vein and complete occlusion with a patent inferior vena cava (IVC) below the renal veins; an intrarenal IVC filter was placed.

Due to poor prognosis and limited treatment options at this stage, the patient and her family decided to halt further treatment and transition to palliative measures instead of ventilatory support. The patient passed away 3 days later. As such, subsequent chest and lumbosacral spine X-ray images are not available.



**Fig. 4** Pleural biopsy and autopsy. (A) Coronal chest computed tomography demonstrating large central embolus completely occluding the left pulmonary artery (yellow arrowhead) and nearly complete in the right pulmonary artery. (B) Pleural biopsy, x100 and (C) x 200. (D) Postmortem pulmonary artery tumor embolism (star, vessel wall), x100. (E) Postmortem representative of pulmonary metastasis, x20.

### Autopsy

At autopsy, metastatic tumor was found within both the right and left main pulmonary arteries (► **Fig. 4D**), along with extensive pulmonary parenchymal metastases (► **Fig. 4E**). Notably, no gross metastasis was observed in the brain or any other organ.

### Discussion

To our knowledge, this is the first reported case of MPE metastasis causing severe pulmonary disease mimicking pulmonary embolism. Previous case studies have reported MPE metastasis to lung parenchyma and pleura without intravascular metastases.<sup>14–16</sup> Previously reported MPE cases with extraneural metastasis are summarized in ► **Table 1**.<sup>17–23</sup>

A proposed mechanism of MPE metastasis is that tumor cells infiltrate the pleural space via hematogenous, direct or lymphatic spread.<sup>19,24</sup> Metastasis is not consistently reported

in the lungs; metastasis has been reported in the cervix, lymph nodes, paraspinal muscles, and abdominal structures.<sup>19–21,23</sup> In cases of lung metastasis, there are no reports of a substantial mass in the pulmonary arteries.<sup>17–19,22</sup>

Although MPE is considered slow growing with low metastatic potential, several factors may complicate an accurate and timely MPE diagnosis. MPEs primarily originate in the sacrum and cauda equina region, and often lead to nonspecific symptoms of lower back pain. MPEs are not appreciated on X-ray and in this case symptom concern (e.g., saddle paresthesia) was not raised until an advanced disease stage. Initially, there was no palpable mass or neurological deficit and the patient developed symptoms of concern (saddle paresthesia and urinary incontinence) after significant tumor growth. More timely investigation of back pain, in this case, could have potentially led to an expedited clinical workup and lesion identification prior to metastasis. Providers are encouraged to consider spinal neoplasm in the setting of chronic back pain of an unknown origin.

**Table 1** Myxopapillary ependymomas (MPEs) with extraneural metastasis; a literature summary (years 1999–2022)

Author, year	Age (years)	Sex	Origin	Histology	Metastasis
Graf et al 1999 <sup>19</sup>	15	Male	Cauda equina	MPE	Lungs, liver, lymph nodes, pleura, chest wall
Rickert et al 1999 <sup>22</sup>	55	Female	Cauda equina	MPE	Lungs
Vega-Orozco et al 2011 <sup>23</sup>	22	Male	Cauda equina	MPE	Inguinal lymph node
Fujimori et al 2013 <sup>18</sup>	28	Male	Cauda equina	MPE	Lungs
Güzin et al 2016 <sup>20</sup>	34	Female	Sacrum	MPE	Cervix
Batich et al 2019 <sup>17</sup>	30	Male	Sacrum	MPE	Lung
Mastantuoni et al 2022 <sup>21</sup>	39	Male	Cauda equina	MPE	Paraspinal muscles

Resection remains the gold standard treatment of MPEs.<sup>6,7</sup> However, treatment for metastatic MPEs is not standardized nationwide. Chemotherapy, radiation, and in one instance immunotherapy after initial resection of the primary lesion have been used previously.<sup>14,25</sup> Prompt initiation of systemic and radiation therapy should be considered when full resection is not possible. Providers should be aware of life-threatening complications such as lung metastasis. In this case, the patient presented emergently for shortness of breath on two occasions, initially thought to be a pulmonary thromboembolism; however, a neoplastic embolism could have been considered given the patient's history of sacral pain.

MPEs, classically "benign" gliomas, can spread outside of the neuraxis and cause associated sequelae of extraspinal metastases. We hope this case raises awareness of an unique intravascular tumor embolus that resulted from pulmonary metastasis of a MPE. Maintaining clinical suspicion of spinal neoplasm in the setting of chronic back pain with undetermined origin is warranted. Finally, the authors further hope this case galvanizes consensus around a standardized treatment protocol for patients with a MPE.

#### Ethical Approval

Institutional Review Board approval is not required for a case report.

#### Patients' Consent

Institutional standard of care surgical informed consent was obtained.

#### Authors' Contributions

The manuscript has been read and approved by all the authors and requirements for authorship have been met. Each author attests that the manuscript represents authentic work.

#### Funding

None.

#### Conflict of Interest

None declared.

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