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Metastatic Pulmonary Intimal Sarcoma Presenting as *Cauda Equina* Syndrome

First Report of a Case

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Study Design. Case report and clinical discussion.

Objective. To describe a rare case of metastatic pulmonary intimal sarcoma presenting as *cauda equine* syndrome.

Summary of Background Data. Pulmonary artery sarcoma is a rare tumor, frequently misdiagnosed as pulmonary embolism, and although it is highly aggressive, metastasis to bone is very rare. In our case, the tumor metastasised to the vertebral column, which lead the patient to present with *cauda equine*-type symptoms.

Methods. We report the clinical and imaging features in a 38-year-old female presenting with back pain and weakness of the legs. Although initially misdiagnosed as a primary bone tumor of the vertebral column and a concurrent pulmonary embolism, the final diagnosis of pulmonary artery sarcoma with spinal metastasis, presenting as *cauda equina* syndrome was made.

Results. At surgery, the mass was excised with the pulmonary valve and replaced with a homograft. The histology demonstrated an undifferentiated intimal sarcoma of the pulmonary artery. When stable enough the patient underwent a posterior L4–S1 decompression laminectomy after which she regained the power in the legs. The histology also showed a metastatic undifferentiated intimal sarcoma likely to have come from the pulmonary artery. The patient went on to receive a more formal vertebrectomy, front and back repair, and radiotherapy. Her 1-year follow-up computed tomography (CT) chest showed evidence of further metastases.

Conclusion. To our knowledge, this is the first case of metastatic pulmonary intimal sarcoma presenting as *cauda equine* syndrome described in the literature.

Key words: pulmonary artery sarcoma, *cauda equina* syndrome, computerized tomography, pulmonary angiogram. **Spine 2008;33:E516–E520**

Case Study

A 38-year-old female presented to her GP with a 3-week history of lower lumbar back pain radiating into both buttocks and the left leg. She also reported some minor weight loss. There were no symptoms of nerve root compression or autonomic dysfunction. Relevant medical history included an episode of pleurisy the previous year, which had resolved with a course of antibiotics. At that time a systolic heart murmur was incidentally noted, although not formally investigated. More recently she had consulted a rheumatologist complaining of knee and left elbow pains and been given a diagnosis of synovitis of both knees and olecranon bursitis. She had been a smoker of 15 cigarettes per day for 20 years. Examination revealed a normal lumbar lordosis with moderate tenderness around the lower lumbar spine. There was no neurologic deficit. Examination of the cardiovascular system confirmed a systolic murmur loudest at the left sternal edge but heard throughout the chest. Examination of the breasts, thyroid, and abdomen was unremarkable.

Radiographs of the lumbar spine demonstrated a destructive lesion of the L5 body with partial collapse, although the adjacent disc spaces were preserved (Figures 1A, B). An infective or inflammatory pathology was considered a strong differential diagnosis given the history of pleurisy, synovitis, and olecranon bursitis, although a neoplastic lesion could not be excluded confidently without further investigation. As a result, the patient was referred to a regional Bone & Soft Tissue Tumor Service where it was recommended that she undergo a whole spine magnetic resonance imaging (MRI) and a CT-guided biopsy of the L5 vertebral body. The MRI demonstrated a low/intermediate T1W and high T2W/STIR (Figure 2A) signal intensity lesion in the body of L5 associated with partial vertebral collapse. The posterior vertebral margin was convex resulting in a moderate narrowing of the spinal canal (Figures 2 A, B). Similar smaller signal intensity lesions were also present within the T6 and T10 vertebrae. CT-guided biopsy yielded a spindle cell lesion with a background of chronic inflammatory change, and therefore a neoplastic lesion could not be confidently excluded.

Subsequent bone scan demonstrated intense increased activity at L5 and moderately increased activity in the mid and lower thoracic spine corresponding to the lesions demonstrated on the MRI scan. Further low-grade activity in the left shoulder blade and maxilla was noted and felt to be degenerative in origin (Figure 3). CT of the chest, abdomen, and pelvis was performed to search for a potential primary lesion or further metastases. Although there was no evidence of primary or secondary lesions elsewhere, a large filling defect thought to be a mass straddling the pulmonary trunk and extending into

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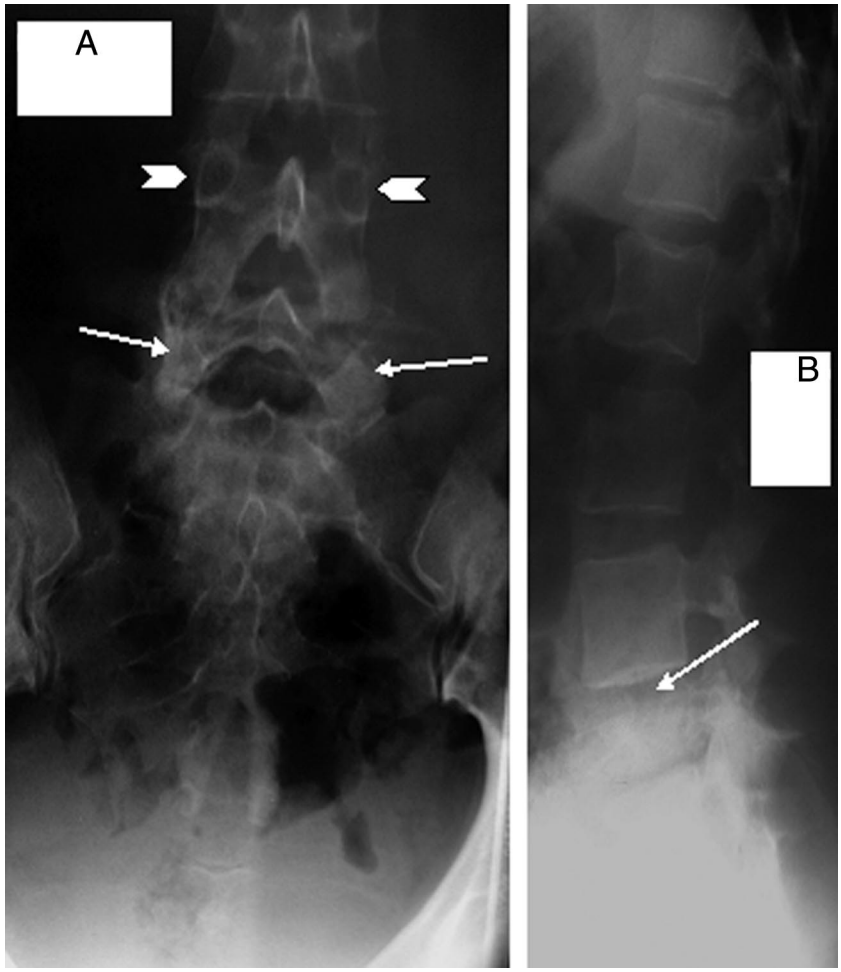

Figure 1. AP (**A**) and lateral (**B**) radiographs of the lumbar spine. The L5 vertebral body is sclerotic with ill-defined margins (arrow in **B**) and reduced height. Note that the pedicles of L5 in the AP view are dense and poorly defined (arrows in **A**). Compare with the normal pedicles of L4 (arrowheads). Pedicular sclerosis is a useful sign indicating vertebral metastases.

the right side of the heart was noted (Figure 4). This was initially thought to be consistent with a large pulmonary embolus secondary to immobility. These findings were discussed with a cardiologist and the patient subsequently commenced on treatment dose of low molecular weight heparin. However, she had little in the way of respiratory symptoms, and it was therefore suggested that the pulmonary lesion could represent a primary neoplastic process.

The patient was now finding it increasingly difficult to mobilize due to weakness of both legs. Examination of the peripheral nervous system revealed weakness of ankle dorsiflexion and extensor hallucis longus bilaterally together with absent ankle reflexes, consistent with the development of a *cauda equina*-type picture. We sought advice from the regional cardiothoracic unit in the context of the pulmonary mass. Her spinal surgery was postponed until she had undergone more cardiac investigation. An electrocardiogram showed T-wave inversions in leads V1–3 indicative of right heart strain. An echocardiogram demonstrated a moderately impaired and dilated right ventricle with a mass in the main pulmonary artery extending out from the left pulmonary artery resulting in raised right heart pressures. They were unable to conclude whether this mass was a simple embolus or a neoplastic lesion. A significant pulmonary hypertension and right ventricular hypertrophy however, meant that this stenotic vascular lesion had been present for several months. A lung perfusion scan revealed the absence of perfusion in the right upper lobe and abnormal perfusion of the left upper lobe. Compounded by the possibility that the mass might be neoplastic, it was decided to relieve the obstruction. At surgery the mass was found to extend through the pulmonary valve resulting in regurgitation. The pulmonary valve was therefore excised with the pulmonary trunk and replaced with a homograft under cardiopulmonary bypass. The histology from the pulmonary artery mass demonstrated an undifferentiated initimal sarcoma of the pulmonary artery (Figure 5).

She made an uneventful recovery from this surgery and when stable enough was transferred back to our high dependency unit. She then underwent an urgent posterior L4–S1 decompression laminectomy after which she regained the power in the legs. The L5 vertebral body histology also showed a metastatic undifferentiated intimal sarcoma likely to have come from the pulmonary artery. Consequently she underwent an anterior L5 vertebrectomy and a front and back repair of L4–S1 with vertebral body cage at L5 (Figure 6). She made an uneventful recovery after which she was referred to a re-

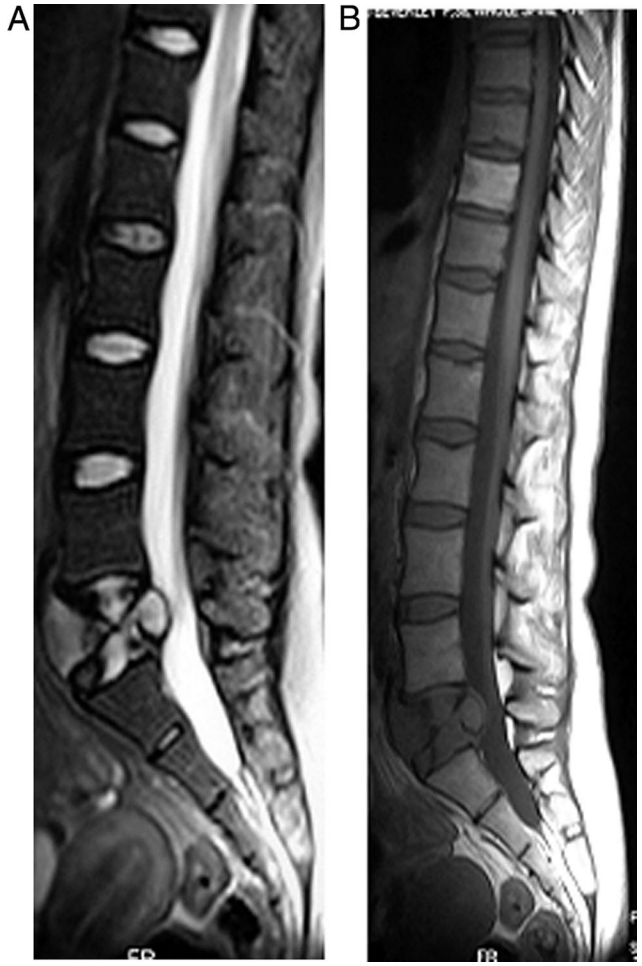

Figure 2. **A**, MRI of the lumbar spine. Sagittal T2W image demonstrates destruction of the L5 vertebra. The posterior vertebral margin shows retropulsion (arrow in **A**) and is causing spinal stenosis which is well seen in the axial T2W image (arrow in Figure **B**).

gional oncology service for radiotherapy to the lumbar spine and the asymptomatic lesions in the thoracic spine.

Her latest chest CT a year after her original surgery shows several new lung metastases in addition to a new bony lesion in the blade of the right iliac bone expanding out into the soft tissues on either side. She is now to receive chemotherapy to shrink the lesions and is being followed up by the bone and soft tissue sarcoma team.

■ Discussion

Primary sarcomas of the great vessels are extremely rare mesenchymal tumors, which are usually found in the pulmonary artery, the aorta (most often abdominal), and inferior *vena cava*. They are highly lethal and most commonly diagnosed at postmortem.[1] There is a slight female predominance,[2,3] through a broad age range, generally affecting adults between the ages of 22 and 81, with a mean age of 48 years for pulmonary and 62 years for aortic intimal sarcoma.

Pulmonary artery sarcomas (PAS) usually arise from the intimal layer of the pulmonary trunk and spread either anterogradely into the pulmonary artery branches

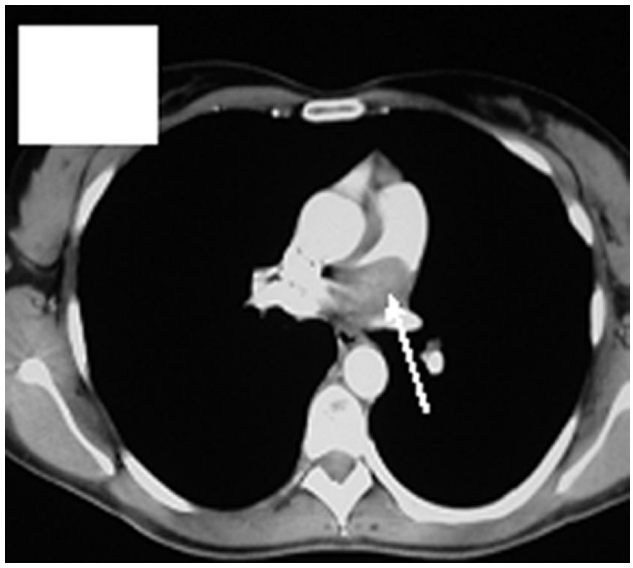

Figure 3. Contrast-enhanced CT of the pulmonary arteries. Cross section through the chest at the level of the infundibulum. A poorly enhanced filling defect is demonstrated in the main pulmonary artery (arrow).

or retrogradely into the pulmonary valve and right ventricle.[4] Metastases are most commonly found in the lungs, although deposits in the pancreas, kidney, brain, lymph node, and skin have been reported.[4,5] Metastases to bone, however, are extremely rare and our case appears to be the first where the neoplasm spread to the vertebral column resulting in *cauda equina* syndrome. Symptoms and signs at presentation correspond to the location of the primary tumor and the metastases.[6] PAS usually presents with dyspnoea, chest pain, cough, and/or hemoptysis. However, because of the insidious nature of PAS, patients sometimes also present with "constitutional" symptoms (*e.g.*, clubbing, weight loss, anorexia). Blood tests results are not very helpful (*e.g.*, with a mild anemia and moderately raised erythrocyte sedimentation rate) and echocardiograms will show evidence of raised right ventricular pressure if the tumor has spread through the pulmonary valve and into the right ventricle resulting in a degree of pulmonary hypertension. This retrograde extension of the sarcoma is a frequently mistaken for pulmonary thromboembolism (PED)[1,2] leading to inappropriate anticoagulation or thrombolysis.

CT pulmonary angiogram is the primary imaging modality used to investigate these patients. However, differentiating filling defects due to PED (common) from PAS is very difficult and sometimes not possible. Features favoring a diagnosis of PAS are location of the filling defect in the proximal pulmonary artery,[7] complete occlusion of the pulmonary vascular lumen, expansion of the pulmonary trunk/pulmonary artery, and extension of the filling defect lesion outside the pulmonary artery lumen.[8] PAS will usually show up as a heterogenous density, sometimes demonstrating calcification and can enhance with intravenous contrast medium.[9,10] A lack of clinical

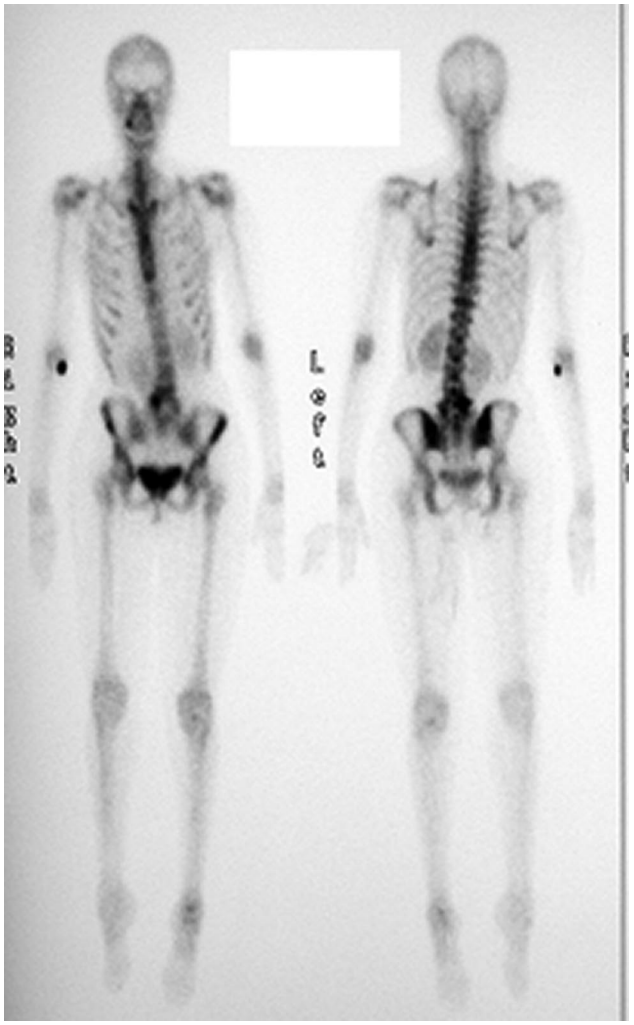

Figure 4. Whole body technetium bone scan–static image acquired at 4 hours. This demonstrates a focus of increased activity (black arrow) in L5 with associated scoliosis. The subtle increased activity in T6 (white arrow) was shown to represent a focus of marrow replacement in T6 on MRI (not shown). The focus of activity at the right elbow corresponds to tracer extravasation at the injection site.

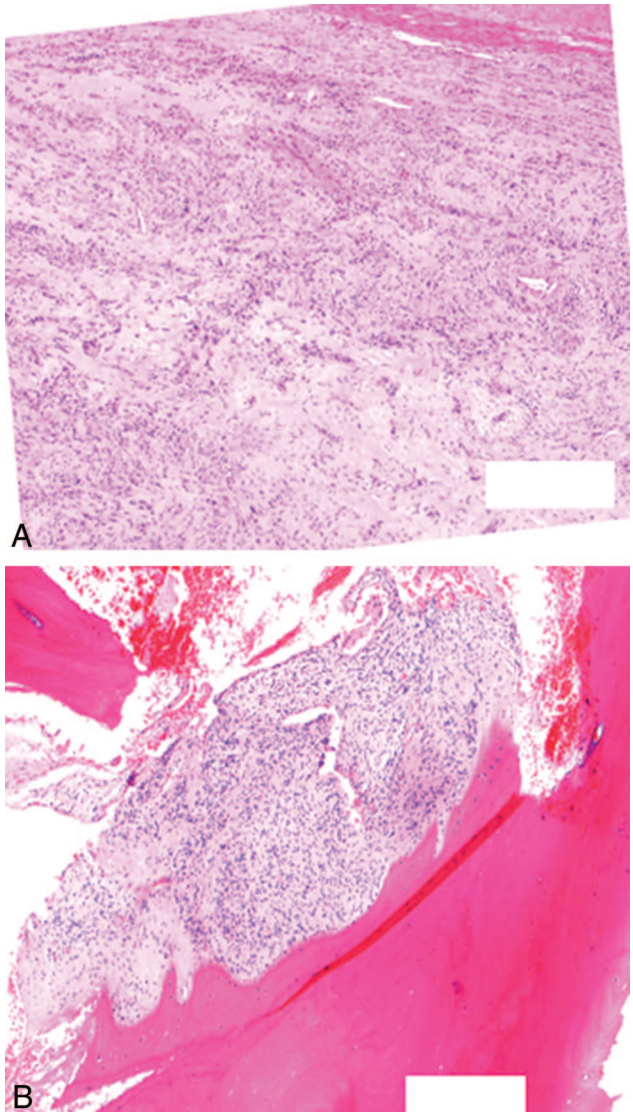

Figure 5. **A**, Intimal sarcoma of the pulmonary artery. Small spindle cells with a moderate cellularity, set in a vascular fibromyxoid stroma. The tunica media of the pulmonary artery is seen on the top right (H & E stain, 4× magnification). **B**, Intimal sarcoma metastatic to L5 vertebra. A spindle cell neoplasm showing the same morphologic features of the primary pulmonary artery tumor, permeates the lamellar cortical bone of the vertebra (H & E stain, 4× magnification).

response to long-term anticoagulation, supports the diagnosis of a neoplastic lesion in place of thromboembolism.[11,12] The advent of multislice CT and MRI have also enable better tissue and anatomy characterization, thus enabling earlier diagnosis and differentiation from other pathologies, namely, PED.[13–16] Although this would theoretically permit the initiation of earlier curative treatment, there is no evidence to date to suggest that earlier diagnosis followed by earlier intervention leads to improved overall survival.

Conventional pulmonary angiography can also be used to image these tumors. Typical "to and fro" motion of the filling defect on a stalk within the pulmonary artery was said to be a characteristic sign of this tumor on conventional pulmonary angiogram.[17] However, as a result of the recent advancements in CT technology, conventional pulmonary angiography is being used far less. In fact, CT pulmonary angiogram confers a significant advantage in terms of both speed and availability.

The mainstay of treatment for PAS is surgical resection (remaining the only potentially curative modality) followed by chemoradiotherapy. An approximate 20% response rate can be expected with an adjuvant chemotherapy regimen involving an anthracycline antibiotic and an alkylating agent, however, the value of this regimen is unclear.[17] The prognosis is poor with a mean survival of 12 months after onset of symptoms and 1- and 2-year survival rates of 22% and 7%, respectively. However, survival figures have improved since 2000 according to the literature.[1,2] This has been attributed to more frequent use of CT and MRI in the diagnosis of suspected PED and a greater awareness of PAS among physicians.[8,18] Use of adjuvant chemoradiotherapy may also have contributed to the improvement in patient

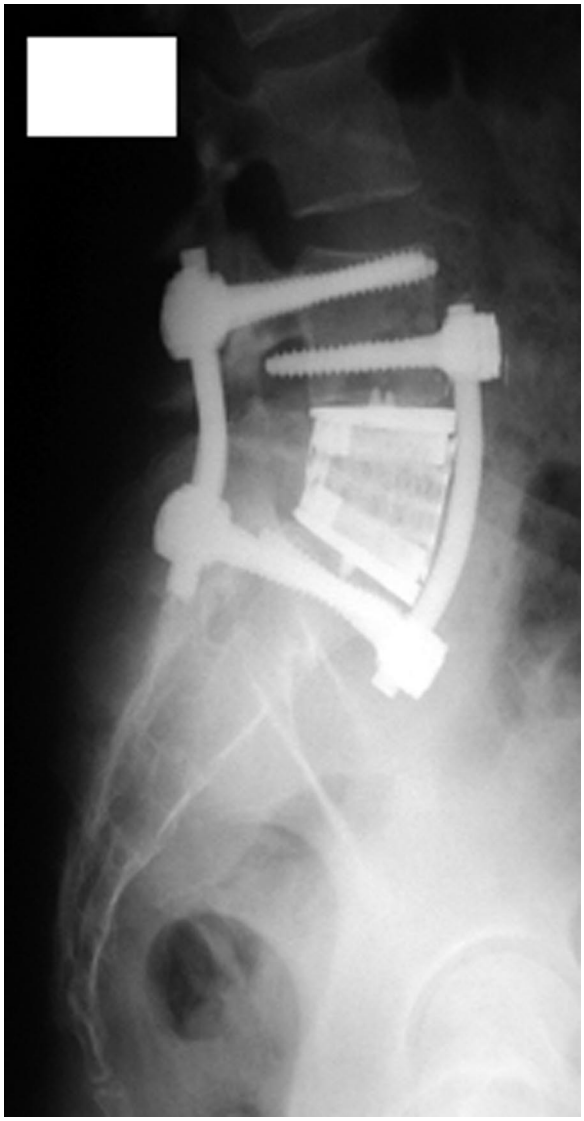

Figure 6. Post operative lateral radiograph shows pedicular screws and anterior body fixation in L4 and S1 with an interbody cage at L5.

survival although the number of patients is too small to confirm this.

In conclusion, we report a novel presentation of a rare tumor. PAS should be suspected in patients who show certain typical CT features of PED but whose symptoms fail to improve with prolonged anticoagulation. There may be clinical indicators or imaging features to suggest PAS over PED and clinicians and radiologists should be aware of this rare diagnosis. With an earlier diagnosis, patient survival may improve although prognosis with a late surgical intervention remains poor.

■ Key Points

- Pulmonary artery sarcoma is a rare primary tumor that is frequently misdiagnosed as pulmonary embolism.
- Pulmonary artery sarcoma should be suspected in patients who show certain typical CT features of pulmonary embolism but whose symptoms fail to improve with prolonged anticoagulation.
- There may be clinical indicators or imaging features to suggest pulmonary artery sarcoma over pulmonary embolism and clinicians and radiologists should be aware of this rare diagnosis.
- With an earlier diagnosis, patient survival may improve although prognosis with a late surgical intervention remains poor.

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