A 33-year-old female presented with four years of shortness of breath. She denied sputum production, fever, chills, chest pain or hemoptysis. She was diagnosed with pulmonary embolism four years prior, treated with anticoagulation for six months. Physical examination was remarkable for sinus tachycardia. Echocardiogram along with pulmonary function tests and diffusion coefficient were normal. Chest Computed Tomography (CT) angiogram showed Inferior Phrenic Artery (IPA) to Pulmonary Artery (PA) communication. A dilated right IPA was seen supplying the vascular malformation in the Right Lower Lobe (RLL), measuring approximately 1.9x1.1 cm (Figure 1). Phrenic arteriogram showed a complex vascular malformation supplied by the right IPA branches and draining into a dilated branch of the RLL PA (Figure 2).

She underwent successful embolization of the IPA-PA fistula using arterial to arterial approach with onyx embolization material and coils.

Systemic-PA fistulas, an extremely rare, cause of filling defects in pulmonary arteries, should be suspected with atypical presentations and imaging findings. Diagnosis is confirmed by delayed contrast-enhanced CT or conventional pulmonary angiography. Trans catheter embolization is a feasible therapeutic option. No definitive treatment guidelines are established, but some recommend treatment to avoid complications including hemoptysis, pulmonary hypertension, and congestive heart failure [1-3].

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Conflicts of interest: None.

References
**Figure 1:** CTA chest showing dilated right inferior phrenic artery supplying the vascular malformation in the right lower lobe which is draining into the right lower lobe branch of the pulmonary artery.

**Figure 2:** Phrenic arteriogram showing a complex vascular malformation supplied by the right inferior phrenic artery branches and draining into a dilated branch of the right lower lobe pulmonary artery (A, B and C). Arteriogram post procedure showing obliteration of the arteriovenous fistula (D).