A 34-year-old, African-American woman with a history of sickle cell disease and a remote history of asthma presented to the inpatient service with hemoptysis and associated pleuritic chest pain for 24 h. The patient was awakened the morning prior to hospital admission by two episodes of hematemesis/hemoptysis. She noted an increase in coughing, with bloody sputum, and shortness of breath during the day and called emergency medical services. The patient was hospitalized 1 month prior to presentation for acute pain crisis and community-acquired pneumonia (Fig 1).

Physical examination on presentation was remarkable for a well-nourished woman in no acute distress. She was afebrile, vital signs were stable, and room air oxygen saturation was 100%. Blood-tinged sputum was noticed in the emergency department. Pulmonary examination revealed rhonchi and decreased breath sounds over the left lung field. The remainder of her examination was otherwise noncontributory at presentation. Laboratory data were significant for a WBC count of 8,200/μL, hemoglobin of 8.0 g/dL (baseline, 7.5 to 10 g/dL), platelets were 223,000/μL, international normalized ratio of 1.2, and partial thromboplastin time of 25.1 s. Posteroanterior/lateral chest radiography demonstrated a multilobar pneumonia with left pleural effusion. She was subsequently admitted to the hospital for pneumonia.

The patient continued to have hemoptysis and dyspnea on hospital day 2, and the pulmonary service was consulted. Chest CT was performed to evaluate the bilateral infiltrates (Fig 2), which showed occlusion of the left mainstem bronchus (LMS), atelectasis of the right upper lobe (RUL) and left lower lobe, and extensive mediastinal adenopathy (Fig 2). No pulmonary embolism was noted. The following morning, the patient underwent fiberoptic bronchoscopy (FOB) to investigate the etiology of the bronchial compression. During FOB, massive hemoptysis occurred from an unknown location resulting in respiratory failure. She was intubated to protect her.
airway and transferred to the medical ICU for stabilization. After intubation, the bleeding subsided and bronchoscopy was performed later that afternoon. The second FOB found 80 to 90% intrinsic obstruction of the LMS and 100% intrinsic obstruction of the RUL bronchus. The bronchoscope was unable to bypass the strictures at the RUL and LMS. Biopsy specimens were obtained at these sites for pathology analysis. There was diffuse inflammation of the mucosa, stigmata of recent bleeding, but no active bleeding noticed in all lung segments visualized. No further FOB was attempted due to the strictures of the RUL and LMS.

What is the diagnosis?

Figure 2. Chest CT with soft-tissue mass (4 × 5 cm) in the mediastinum extending to the subcarinal region. The origin of the RUL is tapered. Patchy consolidation is seen in the RUL and left upper lobe. Ground-glass opacities are noted in the RUL. A subpleural consolidation is seen in the right lower lobe.
Diagnosis: Endobronchial sarcoidosis with massive hemoptysis

The differential diagnosis for endobronchial stenosis and hemoptysis includes primary neoplasm of the lung, metastatic cancer to the lung, pulmonary vasculitis, and granulomatous diseases of infectious or inflammatory origin.1 Evaluation of the patient with hemoptysis begins by defining potential airway diseases, parenchymal abnormalities, and vascular sources of bleeding. A pulmonary vascular source can be arterial, venous, or capillary in origin. Arterial causes of hemoptysis include pulmonary hypertension, pulmonary embolus, and vascular neoplasms such as hemangiomatisis and Kaposi sarcoma.2 Good-pasture disease, pulmonary hemosiderosis, and isolated capillaritis are potential capillary reasons for hemoptysis.2 Venous causes are limited to cardiac and noncardiac sources of venous hypertension, such as pulmonary veno-occlusive disease.2 Bronchial, axillary, intercostal, and subclavian artery fistulas may all cause hemoptysis. These sources of hemoptysis were excluded in our patient by diagnostic angiography performed on hospital day 6.

Most parenchymal reasons for bleeding typically show infiltration on chest radiographs.1,2 Common infiltrative diseases are neoplasm, abscess, mycetoma, and pneumonia.2 FOB was performed to obtain tissue and cell samples in an attempt to better define the cause of the infiltrate in our patient. During FOB, intrinsic stenosis of the airway was noted. Airway disease is the most common source of hemoptysis, and may stem from bronchiectasis, bronchitis, endobronchial tumors, endometriosis, tuberculosis, or fungi.2

Granulomas of infectious origin must be excluded, as these diseases can manifest as parenchymal or airway pathogens.2 Tuberculosis is the most common granulomatous disease, infecting a third of the population of the world, and should be sought in immunocompromised patients, patients that live in close quarters such as the homeless or inmates, and patients with exposure to tuberculosis.3 Fungal pathogens such as histoplasmosis, blastomycosis, coccidioidomycosis, and aspergillosis must be considered and were excluded from the differential by silver stain and periodic acid-Schiff staining of biopsy specimens.2,4 Uncommon infectious pathogens include actinomycosis and Nocardia.2

The lung is a frequent site of metastasis due to its large blood flow and extensive capillary network.5 Primary lung cancer was excluded in our patient based largely on her age of 34 years,1 and later confirmed by bronchial biopsy. However, secondary or metastatic cancers to the lung in our patient’s age group are possible.1 Breast cancer is a common fatal malignancy in women, causing > 500,000 deaths per year worldwide secondary to metastatic disease.6 Pulmonary metastasis of cervical cancer can be as high as 6.1% in patients with extrapelvic disease.7 Breast and cervical cancer do not usually present as occult disease to the lung, but should be considered high in the differential of secondary neoplasms. Germ-cell tumors, both gonadal and extragonadal, have a tendency to metastasize to the lung and are common in childhood and young adults.8 Finally, in women of childbearing age, persistent trophoblastic disease must be considered.9 Histologic classification of trophoblastic disease has important prognostic value.10 Complete hydatiform moles commonly metastasize to the lung, while partial hydatiform moles rarely involve the lung.9

Pulmonary manifestations of vasculitis are rare, despite its association with connective tissue diseases.1,11 Systemic polyangiitis associated with Behçet syndrome, lupus, rheumatoid arthritis, microscopic polyangiitis (MPA), and polyarteritis nodosa can have radiologic findings and clinical signs of bronchial stenosis.11,13 MPA is a necrotizing, small-vessel vasculitis that deserves special mention due to its pulmonary involvement similar to Wegener granulomatosis and pathologic similarity to Churg-Strauss syndrome, both of which are important causes of endobronchial stenosis.11,13 Antineutrophil circulating antibodies of the cytoplasmic type are associated with Wegener granulomatosis, Churg-Strauss syndrome, and MPA.11 The lack of granulomatous inflammation distinguishes MPA from Wegener granulomatosis.11 The presence of eosinophilia and asthma separates Churg-Strauss syndrome from MPA.11 Our patient’s serology for antineutrophil circulating antibodies was negative, making the diagnosis of vasculitis unlikely.13

Citron and Scadding first described bronchostenosis related to sarcoidosis in 1957.2 Airway obstruction is the most common presenting pulmonary abnormality occurring at all stages of sarcoidosis and is associated with increased morbidity.14 Extrinsic compression by lymph nodes, granulomatous inflammation of the airway, and distortion of the airway by parenchymal fibrosis can all lead to airway obstruction.2 Chest CT scan is useful to define the cause and severity of obstruction.2 The CT scan of our patient illustrated intrinsic narrowing of the RUL and LMS, and biopsy specimens were obtained at these locations. Pathology analysis of the biopsy samples found noncaseating, epithelioid granulomas consistent with sarcoidosis (Figs 3, 4).

Hemoptysis, in patients with sarcoidosis, is an uncommon presenting sign, in spite of the high incidence of pulmonary involvement.15 In a review16 of patients with sarcoidosis, < 7% of the patients had...
hemoptysis. Bronchoscopy is only useful in identifying endobronchial lesions and rarely localizes the source of bleeding in patients without massive hemoptysis, defined as > 500 mL in 24 h.\textsuperscript{4,14,17}

Our patient continued to have a small amount of hemoptysis during her hospitalization; therefore, no further FOB was attempted. Blocking of the RUL and LMS with either Fogarty or Swan-Ganz catheters are temporizing possibilities, but as long as hemoptysis was not massive, these options were not implemented. The primary team elected to have definitive treatment of the hemoptysis performed by attempting bronchial artery embolization. Interventional radiology performed two bronchial artery angiograms for possible coil embolization on hospital days 4 and 6, but were not able to localize a source of bleeding (compatible with low-grade bronchial bleeding). Retrospectively, a pulmonary artery source of bleeding, secondary to Rasmussen aneurysm, can account for 5 to 10\% of hemoptysis cases. Angiography of the pulmonary artery was not performed. In cases with diffuse mucosal bleeding, treatment with laser or cryotherapy can be performed. These procedures were not available at our facility. Finally, on hospital day 7, she had a new bout of massive hemoptysis and succumbed to respiratory failure.

\textbf{References}