A rare “mimicker” of lung malignancy

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Clinical presentation—A 50-year-old African-American woman was referred to our institution for workup of a right hilar mass. The patient reported that she had been evaluated 2 years prior and underwent a right upper lobar resection of a mass which turned out to be benign. At the time of presentation, she mainly complained of dyspnoea on exertion and occasional chest pain. On physical examination, she was seen to have increased fullness of her face, but oxygen saturations were 95% on room air. X-ray of the chest (Figure 1A) and CT scan of the chest (Figures 1B, 1C & 1D) are shown below. What is the diagnosis?

Figure 1A. Chest X-ray showing right hilar mass

Figure 1B. Chest CT scan showing calcified right hilar mass with mediastinal involvement and superior venacava encasement (arrow)

Figure 1C. Chest CT scan showing encasement of superior venacava and right pulmonary artery (arrow)

Figure 1D. Chest CT scan showing splenic calcification (arrow)
Answer & Discussion—Fibrosing mediastinitis. Chest CT scan showing a right hilar mass with calcification and circumferential involvement of the right main stem bronchus, pulmonary artery and superior venacava with consequent narrowing. Splenic calcifications (arrows) are present consistent with sequelae of histoplasma infection.

Proliferative fibrotic process in the mediastinum with associated airway or vascular compromise is called fibrosing mediastinitis, with morbidity and mortality related to the location and extent of fibrosis.

It is a rare disease, with a vast majority of cases (>90%) secondary to previous histoplasma infection, especially in the United States. Pathogenesis is believed to be secondary to leak of fungal antigens from lymph nodes into mediastinum causing a hypersensitivity reaction with excess fibrotic response. Diagnosis is based on clinical and radiological presentation with associated calcification.¹

There is no effective treatment. Antifungals have been found to have some potential benefit in case reports, while surgery to palliate symptoms by relieving airway and vascular compromise have been tried with variable success.²

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