

Fig 3. Photomicrograph (original magnification $\times 400$) of carcinoid tumor shows positive cytoplasmic staining for insulin-like growth factor-1.

of acromegaly are rare. This is because hormone levels are low and have fairly short half-lives [3, 4]. These tumors usually present with pulmonary symptoms such as cough, wheezing, hemoptysis, and recurrent pneumonias [5]. Most cases of acromegaly are the result of ectopic production of GHRH, and only rarely GH directly [4]; however, this patient presented with a bronchial carcinoid with associated acromegaly due to secretion of IGF-1.

Physiologic GH secretion is controlled by a complex interaction of factors, including GHRH, ghrelin, somatostatin, estrogen, glucocorticoids, and IGF-1 [1]. Most acromegaly is caused by pituitary somatotrope adenomas that secrete excessive amounts of GH. These patients present with features of frontal bossing, mandibular enlargement, prognathism, soft tissue swelling, a deep or hollow-sounding voice, arthropathies, carpal tunnel syndrome, and increased hand and foot size or enlargement of organs, such as cardiomegaly or macroglossia [1]. Signs of insulin resistance are also present, owing to the antagonistic effects of GH. Clinical and laboratory features caused by extrapituitary tumors are indistinguishable from those resulting from pituitary adenomas [2]. Regardless, surgical removal of these tumors is the primary treatment modality [1-3, 5].

Histology and location of the tumor influence the choice of surgical procedure [5]. Carcinoid tumors are classified as typical or atypical. Typical features include mitotic activity in fewer than 2 cells per 10 HPF and absence of focal necrosis. Atypical features include greater mitotic activity and punctuate necrosis [6]. Metastasis to regional lymph nodes occurs in less than 15% of typical bronchial carcinoids, but may be present in 30% to 50% of atypical tumors.

Peripheral tumors with typical features are preferably removed with a large wedge or segmental resection, whereas more radical procedures, such as lobectomy with lymph node sampling, bilobectomy, sleeve resection, or pneumonectomy, are often chosen for central or

atypical carcinoids [5]. Overall, long-term postoperative survival is 83% to 94% for typical carcinoids and 31% to 74% for atypical carcinoids [5]. In cases of unresectable disease, long acting somatostatin analogues, such as octreotide, may be used to suppress the production of GHRH, GH, and IGF-1 for control of symptoms [2, 3, 7].

Our patient demonstrated only elevated levels of IGF-1 with normal level of GH. He was treated with surgical resection with resolution of symptoms and normalization of his biochemical abnormalities.

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An Unusual Case of Aspergillus Fibrosing Mediastinitis

Sean C. Wightman, BA, Anthony W. Kim, MD,
Laurie A. Proia, MD, L. Penfield Faber, MD,
Paolo Gattuso, MD, William H. Warren, MD, and
Michael J. Liptay, MD

Division of Thoracic Surgery, Section of Infectious Disease,
Department of Pathology, Rush University Medical Center,
Chicago, Illinois

Fibrosing mediastinitis due to *Aspergillus* is rare, particularly in the immunocompetent host. Fibrosing mediastinitis due to *Aspergillus* species in the immunocompetent patient can be indolent and may be treated with antifungal therapy rather than surgery. We present a 78-year-old non-smoking, nondiabetic woman with chronic fibrosing medi-

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Address correspondence to Dr Liptay, Division of Thoracic Surgery, Rush University, 1725 West Harrison St, Suite 774, Chicago, IL 60612; e-mail: michael_liptay@rush.edu.

astinitis due to *Aspergillus*. Multiple attempts at securing a tissue diagnosis were inconclusive. Ultimately, *Aspergillus* infection was diagnosed by a video-assisted thoracoscopic surgical biopsy. The patient was started on oral voriconazole, and she remains clinically stable with radiographic improvement. A prolonged, perhaps life-long, course of antifungal therapy is planned.

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Fibrosing mediastinitis due to *Aspergillus* is rare, particularly in the immunocompetent host. Fibrosing mediastinitis due to *Aspergillus* species in the immunocompetent patient can be indolent and may be treated with antifungal therapy rather than surgery. We present a 78-year-old nonsmoking, nondiabetic woman with chronic fibrosing mediastinitis due to *Aspergillus*, and we describe the clinical course and medical management of this uncommon infection.

A 78-year-old woman without a significant past medical history presented to an outside hospital with cough and hemoptysis 5 years ago. Treatment with an oral antibiotic for presumed community acquired pneumonia relieved her symptoms, but a right lower lobe infiltrate persisted on chest roentgenogram. A computed tomographic (CT) scan demonstrated right lower lobe pneumonitis with lymphadenopathy, and 2 months later a repeat CT scan was unchanged. However, a third serial CT scan performed 5 months after the initial scan revealed a hilar mass, a two centimeter lymph node in the right hilar region, and a nodule in the right lower lobe surrounded by patchy infiltrates (Fig 1). A positron emission tomographic scan and a CT scan demonstrated increased activity in the right hilar mass, along with mild activity in the right lower lobe. One year later the patient was referred to our institution, and a flexible bronchoscopy with biopsy was inconclusive. Histopathology showed reactive epithelial cells without evidence of neoplasm; no cultures were sent at that time. Three months later, the patient underwent a fine-needle aspiration and core needle biopsy of the right lung mass. These findings were also nondiagnostic; histopathology showed no neoplastic cells or granulomas, and special stains for microorganisms, including fungi, were negative.

During this time, the patient remained asymptomatic without fever, cough, or chest pain. However, serial CT scans demonstrated progression in size of the ill-defined, lower right hilar mass with encasement of the right lower lobe bronchus, inferior pulmonary vein, and encroachment on the left atrium. Endoscopic ultrasound with biopsy was attempted but was deemed too risky due to the encasement of the left atrium. A repeat CT scan showed a mass (4.8 cm) along the posterolateral wall of the left atrium. This mass extended anteriorly, encompassing the ascending aorta, compressing the superior vena cava, and severely encompassing and narrowing the right mainstem bronchus. The

Fig 1. An infiltrating mass in the right infrahilar region. The arrow shows an edge of the mass. (DA = descending aorta; LA = left atrium; RA = right atrium.)

paratracheal and subcarinal lymph nodes were enlarged, and the right lower lobe nodule was still present (Fig 2).

The patient ultimately underwent a video-assisted thoracoscopic surgical biopsy of the mediastinal mass. Histopathologic examination was negative for malignancy, but revealed granulomatous inflammation. A Gomori methenamine silver stain highlighted fungal hyphae. Fungal culture of the mediastinal mass grew a mold on two of three plates after 3 days of incubation, which was later identified as *Aspergillus flavus*. On further questioning, the patient reported being a nonsmoker who lives alone in a suburb outside of Chicago. She denied any obvious evidence of mold in the bathroom, attic, or basement of her home. She was not aware of any construction in her neighborhood but does like to garden outdoors.

Comment

Aspergillus, a mold found throughout the environment, may cause invasive disease in the immunocompromised host, but it is a rare pathogen in the immunocompetent individual [1-3]. The most common manifestation of invasive aspergillosis is pneumonia, because the lungs serve as the main portal of entry for inhaled spores [4]. Although there are many causes of fibrosing mediastinitis, most cases are usually due to the immunologic reaction to the antigens of *Histoplasma*, also found throughout the environment. Mediastinal aspergillosis may result from a primary *Aspergillus* pneumonia that spreads to adjacent mediastinal structures [5]. Most of the reported cases of *Aspergillus* mediastinitis are caused by *Aspergillus fumigatus* and occur in the pulmonary arteries, the heart, or the aorta [1].

The first reported case of *Aspergillus flavus* causing granulomatous mediastinitis was in 1981. A 22-year-old

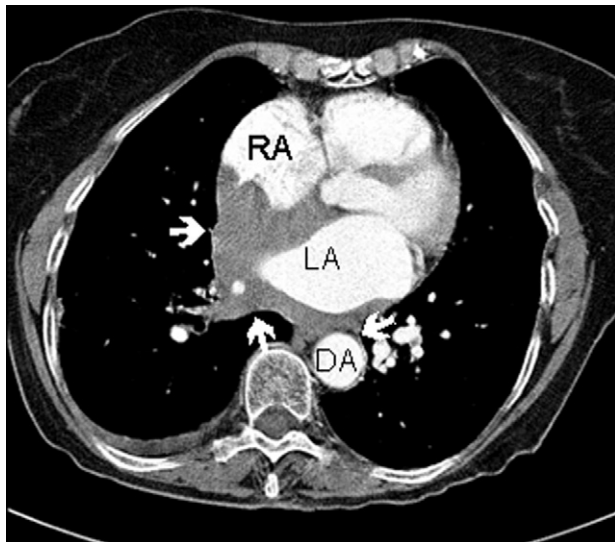


Fig 2. The shown hilar infiltrate extends into the interatrial septum, encompasses the right atrium, and also partially encases the ascending aorta. The arrows show edges of the mass. (DA = descending aorta; LA = left atrium; RA = right atrium.)

immunocompetent man presented with cough, fever, and a right hilar mass. Mediastinal biopsies revealed *Aspergillus flavus*. The patient was treated with amphotericin B and 5-fluorocytosine, but expired after esophageal and superior vena caval compression [3]. Another case published in 1984 described chest wall invasion by *Aspergillus flavus* in a 24-year-old healthy, immunocompetent man. After *Aspergillus flavus* was confirmed on biopsy, amphotericin B therapy resulted in complete resolution of the infection [6].

It is interesting to note that our patient had an atypical course of chronic mediastinal aspergillosis as evidenced by chest roentgenograms and CT scans obtained within 5 years. Although the infection progressed during the 5 years, the patient remained essentially asymptomatic during that time. It is plausible that *Aspergillus* was the cause of her initial right lower lobe pneumonia, because the infiltrate evolved into a right lower lobe nodule and then subsequently progressed to a right hilar mass with ipsilateral adenopathy. Because our patient is presumed to be immunocompetent, it is possible that her immune system was able to partially control the infection during this period of time. This is distinctly different from the typical pattern of *Aspergillus* infections in immunocompromised patients, which is usually rapidly progressive [2]. Despite our patient's lack of symptoms, the growth of the mediastinal mass was troublesome and a definitive tissue diagnosis was needed. Using the video-assisted thoracoscopic surgical approach, adequate tissue specimens were obtained directly from the mass to reveal fungal hyphae and *Aspergillus flavus* in the tissue culture.

Optimal treatment of *Aspergillus* mediastinitis is unclear because of the infrequent number of reported cases. The few published reports have shown the benefit of combined surgical and medical therapy [7]. Our patient

did not receive surgical debridement owing to the extensive involvement of mediastinal structures. She was started on oral voriconazole, 200 mg twice daily, with monitoring of serum concentrations. Voriconazole, an extended spectrum triazole antifungal, is recommended as a first-line therapy for infections caused by *Aspergillus* [4]. The patient continues to do well clinically and radiographically, with no evidence of disease progression after 6 months of treatment with voriconazole. The exact duration of antifungal treatment is unknown, but is likely to be necessary indefinitely.

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Thoracic Outlet Syndrome in a Patient With Poland Syndrome

Levent Özçakar, MD, Engin Çakar, MD,
Mehmet Zeki Kıralp, MD, Alparslan Bayram Çarlı, MD,
Oğuz Durmuş, MD, and Ümit Dinçer, MD

Department of Physical Medicine and Rehabilitation, Gülhane Military Medical Academy Haydarpaşa Training Hospital, İstanbul, Department of Physical Medicine and Rehabilitation, Hacettepe University Medical School, Ankara, Turkey

We report a 20-year-old man with Poland syndrome who suffered from weakness, pain, numbness, and discoloration in the left upper extremity. He was eventually diagnosed as also having thoracic outlet syndrome. The concomitance of these two disorders is discussed with a special emphasis on the underlying mechanisms.

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Poland syndrome is a rare congenital disorder that refers to the absence of pectoralis major and minor muscles, ipsilateral breast hypoplasia, and the absence of

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Address correspondence to Dr Özçakar, Fakülteler Mahallesi Yeni Acun Sokak 11/2, Cebeci, Ankara, 06100, Turkey; e-mail: lozcakar@yahoo.com.