CASE REPORT

Dennis H. Kraus, MD, Section Editor

TRACHEAL MUCORMYCOSIS PRESENTED AS AN INTRALUMINAL SOFT TISSUE MASS

Omer Wolf, MD,1 Ziv Gil, MD, PhD,1 Leonor Leider-Trejo, MD,2 Avi Khafif, MD,1 Philippe Biderman, MD,3 Dan M. Fliss, MD1

1 Department of Otolaryngology Head & Neck Surgery, Tel-Aviv Sourasky Medical Center, Sackler Faculty of Medicine, Tel-Aviv University, Tel-Aviv, Israel
2 The Pathological Institute, Tel-Aviv Sourasky Medical Center, Sackler Faculty of Medicine, Tel-Aviv University, Tel-Aviv, Israel
3 The Intensive Care Unit, Tel-Aviv Sourasky Medical Center, Sackler Faculty of Medicine, Tel-Aviv University, Tel-Aviv, Israel

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Abstract: Background. Mucormycosis is a potentially lethal disease caused by an opportunistic fungal infection. It occurs mostly in diabetic or immunosuppressed patients and usually involves the lungs or paranasal sinuses.

Methods. We report a rare case of a patient with diabetic ketoacidosis who presented with progressive cough and dyspnea. CT of the neck and chest showed an intraluminal soft-tissue mass extending from the first tracheal ring to the thoracic inlet, causing severe destruction of the trachea. Direct laryngoscopy and biopsy demonstrated hyphal invasion with architecture typical of invasive mucormycosis.

Results. The patient underwent resection of the upper trachea and prolonged amphotericin B therapy and is disease free at 24 months after surgery.

Conclusions. In patients with diabetes presenting with progressive hoarseness, dyspnea, and endobronchial mass, a fungal infection should be considered. In case of invasive tracheal mucormycosis, prompt diagnosis and early surgical resection may help improve survival. © 2004 Wiley Periodicals, Inc. Head Neck 26: 541 – 543, 2004

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Mucormycosis is an opportunistic fungal infection that occurs mostly in diabetic or immunosuppressed patients. The common clinical manifestation of mucormycosis is rhinocerebral, pulmonary, or disseminated disease. Pulmonary involvement is typically expressed by rapidly progressive pneumonia with diffuse infiltrates on chest x-ray.1 Very rarely, mucormycosis may involve the bronchi2–7 or the trachea.8,9 Such cases are characterized by mucosal ulceration and vascular invasion, leading to profound tissue necrosis and destruction of the normal bronchial architecture. Prognosis is poor, with a mortality rate of greater than 50%.10

This case describes the presentation, diagnosis, and successful treatment of a patient with severe invasive mucormycosis of the trachea.
CASE REPORT
A 61-year-old man with poorly controlled non–insulin-dependent diabetes mellitus presented with a 2-week history of rapidly progressive shortness of breath. On admission, he had severe respiratory distress and metabolic acidosis (pH 7.01, PCO₂ 28 mmHg, PO₂ 85 mmHg in 100% O₂, anion gap 26 mmol/L). Fiberoptic laryngoscopy revealed a subglottic lesion occluding the lumen. CT of the neck and chest demonstrated an intraluminal soft tissue mass extending from the first tracheal ring to the thoracic inlet (Figure 1, A). The patient underwent a low tracheostomy and biopsy, which showed tissue necrosis with cocci infiltration. Intravenous antibiotic therapy (ceftriaxone and metronidazole) was initiated but failed to achieve any improvement. Clinically, the patient continued to deteriorate. He suffered from a progressive dyspnea and required mechanical ventilation. CT repeated 10 days after admission showed progression of the disease and disappearance of the normal tracheal architecture (Figure 1, B). A second direct laryngoscopy was performed, and examination of the trachea revealed a soft, grayish-pink mass occluding the lumen, with the consistency of chewing gum. This time the biopsy demonstrated hyphal invasion (Figure 2) with architecture typical of mucormycosis (ie, nonseptate hyphae with frequent right angle branching). A revision of the pathologic slides of the first biopsy displayed the fungal elements in the deeper sections.

The patient underwent surgical removal of the necrotic tissue, including partial sternectomy and resection of the trachea from the cricoid cartilage to 4 cm above the carina. A diversion procedure was performed, and the tracheal stoma was sutured to the skin below the suprasternal notch. The inferior laryngeal inlet was obliterated at the level of the cricoid cartilage with a rotational sternocleidomastoid muscle flap, forming a blind stump below the larynx. Liposomal amphotericin B (1.2 mg/kg) was then administered for 6 weeks. The postoperative course was complicated with bilateral pneumonia and recurrent pleural effusions. Blood and body fluid cultures failed to show fungal or bacterial growth. The patient was discharged 2 months later in good condition. At 24 months after surgery, the patient is disease free and undergoing rehabilitation. He is unable to speak owing to the diversion of his trachea; however, he can eat and drink without difficulty.

DISCUSSION
We report a rare case of tracheal mucormycosis in a patient with diabetic ketoacidosis. His initial presentation required the differential diagnosis of a focal necrotic involvement of the trachea. Most such cases are related to a secondary injury of the trachea from tubing in prolonged mechanical ventilation or from jet-ventilation (eg, necrotizing ischemic tracheobronchitis). The primary involvement in our case precluded these considerations and raised the suspicion of a neoplastic disease (ie, infiltrating lymphoproliferative disease, chondrosarcoma, adenoid cystic carcinoma) or a granulomatous disease (ie, Wegener’s granulomatosis, tuberculosis). An infectious etiology other than tuberculosis would include invasive mycotic disease, which has been described occasionally with aspergillosis although rarely with mucormycosis. The key to diagnosis in such
cases lies in the biopsy of the lesion. The erroneous interpretation of the first biopsy and the mixed bacterial growth in culture from the necrotic specimen caused a delay in diagnosis during the first 10 days of admission, although a mycotic etiology was highly suspected (thus raising the question whether earlier empiric treatment with amphotericin B would have been justified).

Primary invasive tracheal mucormycosis is very rare. There are two previous reports on tracheal involvement of mucormycosis in the English literature. The first described a young diabetic female with a small and localized tracheal lesion, enabling successful resection of the lesion and primary anastomosis followed by administration of amphotericin B. The second involved an elderly diabetic man with profound tracheal fungal invasion who subsequently died of bilateral pneumonia before resection was attempted. These two cases and the patient presented here had severe uncontrolled diabetic ketoacidosis. All patients complained of sore throat, progressive hoarseness, and dysphagia, whereas our patient also presented with severe dyspnea.

After tracheal mucormycosis was diagnosed, management consisted of prompt radical surgical intervention followed by intravenous administration of amphotericin B for 6 weeks. Airway reconstruction was not attempted because of the extent and severity of the tracheal involvement and because of the critical condition of the patient. It is possible though that an early diagnosis of the pathogen would have allowed initiation of amphotericin B before surgery and a less extensive procedure.

**CONCLUSION**

Mucormycosis is an uncommon disease that rarely involves the trachea. Patients usually present with diabetic ketoacidosis and progressive dysphagia and dyspnea. Prompt diagnosis and early surgical resection of the necrotic tissues and antifungal treatment may help improve survival in these cases. Tracheal mucormycosis should be considered in the differential diagnosis of an endobronchial lesion in a diabetic patient.

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**REFERENCES**