Acta Scientific Otolaryngology (ISSN: 2582-5550)

Volume 5 Issue 8 August 2023

Case Report

Isolated Tracheal Mucormycosis - A Rare Entity

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DOI: 10.31080/ASOL.2023.05.0585

Received: June 12, 2023

Published: July 10, 2023

Abstract

Mucorymycosis is a fungal infection caused by rapidly invasive filamentous fungi of the Mucorale order. Typically, patients who are immunocompromised or have uncontrolled diabetes fall prey to this uncommon infection. Manifestations of Mucormycosis can range from a pulmonary infection to a lethal rhino-orbital-cerebral infection. The mortality rate of this disease is very high.

The clinical presentation of pulmonary and respiratory tract mucormycosis is non-specific. These include cough, dyspnea, haemoptysis, fever, and therefor can easily be missed if the index of suspicion is kept low. Our case represents one of the various respiratory manifestations of the disease, which usually presents as a pulmonary infection associated with a tracheo-bronchial spread. However an isolated tracheal involvement as seen in our case makes this a rare one.

Here we report a case of isolated tracheal mucormycosis in a previously undetected case of uncontrolled diabetes, successfully treated over a prolonged period. Our discussion spans early diagnosis, treatment, cure and rehabilitation. She was treated over a protracted course of hospital stay and out-patient care, requiring multiple hospital admissions- with both medical and timely surgical intervention being vital in the survival of the patient.

Early detection and aggressive evaluation are vital in making an accurate diagnosis so as not to delay treatment. The three cardinal elements of management of an invasive fungal infection are- control of risk factors, antifungal treatment and early surgical intervention.

Keywords: Laryngotracheal; Mucormycosis; Fungal; Tracheal; Mucor

Abbreviations

CECT: Contrast Enhanced Computed Tomography; POD: Post-op Day; RT: Ryle's Tube; VC: Vocal Cord; GA: General Anaesthesia; B/L: Bilateral; MOTT: Mycobacteria Other Than Tuberculosis

Introduction

Invasive fungal infections have been on the rise, especially in the COVID- Post COVID- era. Mucorymycosis is a fungal infection caused by rapidly invasive filamentous fungi of the Mucorale order. Typically, patients who are immunocompromised or have uncontrolled diabetes fall prey to this uncommon infection [1]. The incidence is higher in India, this has been attributed to poorly controlled diabetes [2]. Whereas in Europe, the most common risk factors are hematological malignancy and transplantation, in view of the immunosuppression associated with malignancy or treatment- this highlights the importance of having regular healthchecks, leading to the early detection and control of diabetes.

Manifestations of Mucormycosis can range from a pulmonary infection to a lethal rhino-orbital-cerebral infection [3]. Inhalation of spores of the organism is what leads to a pulmonary infection that is both rapidly spreading and associated with a high mortality. A pneumonic infiltration leading to a parenchymal disease is the most common manifestation, with involvement of larger airways like trachea and bronchi being rare [4,5].

Patients can present with very non-specific symptoms like cough, dyspnea, haemoptysis and fever [6]. Therefore, it is easy to miss the initial manifestations of the disease, making an early diagnosis difficult. In patients who are at high risk, e.g.: immunocompromised patients and diabetic patients, a high index of suspicion is warranted so as not to miss this life threatening disease.

An isolated tracheal mucormycosis, without parenchymal involvement altogether, is even more uncommon. Here we report a case of isolated tracheal mucormycosis in a previously undetected case of uncontrolled diabetes, successfully treated over a prolonged period- from diagnosis, to treatment, cure and rehabilitation. She was treated over a protracted course of hospital stay and out-patient care, requiring multiple hospital admissions- with both medical and timely surgical intervention being vital in the survival of the patient.

Case Report

A 32 year old female presented to us with a history of severe throat pain since 20 days- associated with severe odynophagia even for saliva. She also had fever which was high grade, which reduced with antipyretics. She also complained of pain while touching/ moving her neck. She also noticed a change in her voice since 7 days. On clinical examination- she had severe tenderness on palpation of the laryngeal framework. There were no palpable neck nodes. She was initially treated with oral antibiotics with no relief of symptoms. She was admitted in view of severe throat pain. A flexible video laryngoscopy done on the day of presentation showed an edematous left vocal cord, with a left cord palsy. In order to assess this further, a CECT Neck was done which showed mild diffuse edema and minimal narrowing of the supraglottic and glottic regions-likely representing infective/inflammatory process. There was no demonstrable mass lesion along the course of recurrent laryngeal nerve. Multiple small air pockets were seen in the visceral space within and around the thyroid gland obscuring the thyroid parenchyma without surrounding inflammatory changes or collection, features suggestive of an emphysematous thyroiditis (Figure 1).

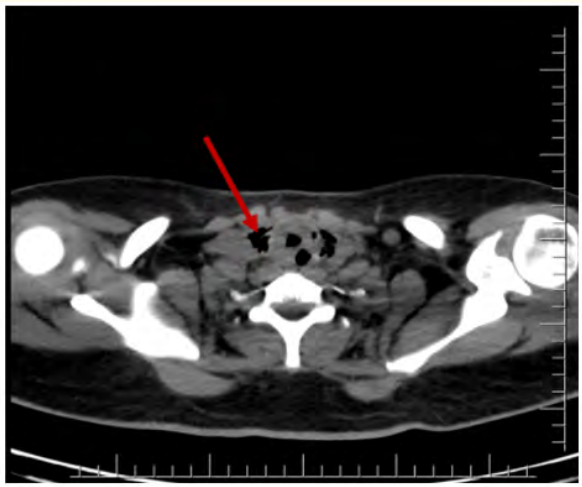

Figure 1: CT: Mild diffuse edema and minimal narrowing of the supraglottic and glottic regions, multiple small air pockets in the visceral space within and around the thyroid gland (red arrow).

She was found to have deranged blood sugar values (RBS 523mg/dl, FBS 323 mg/dl, HbA1c 16.3%) and hence a physician opinion was sought in view of uncontrolled diabetes mellitus and advice followed. She was started on a course of Inj Augmentin empirically.

On day 2 of admission, she developed noisy breathing and a repeat flexible videolaryngoscopy showed a fixed left VC and right VC paresis: Bilateral abductor palsy (Figure 2). She had a narrow glottic chink, and subglottic edema. She was then taken up for emergency tracheostomy. During dissection of soft tissue, necrotic tissue and secretions were visualized in the pretracheal region same removed and sent for AFB stain , culture , gene xpert MOTT, fungal stain and culture, aerobic culture, gram stain. A tracheoscopy was done that showed necrotic slough in subglottic area (Figure 3), which was also sent for pathological and microbiological evaluation. Sensitivity guided antibiotic treatment was given and she was started on a course of Inj Meropenem (14 days), Inj Piptaz (7 days), along with pain management. She was started on RT feeds in view of worsening odynophagia.

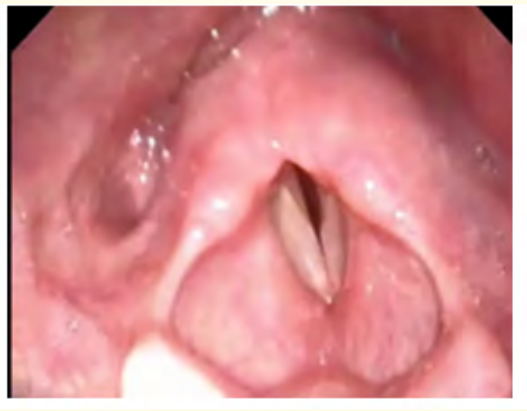

Figure 2: B/L abductor palsy.

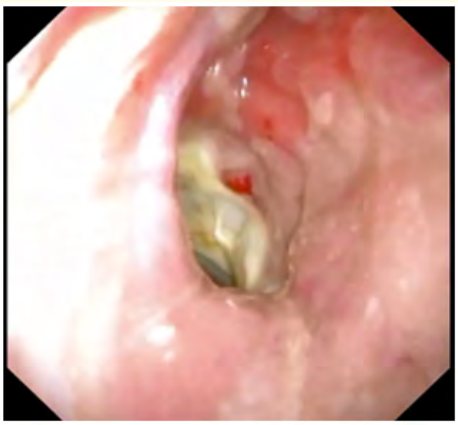

Figure 3: Tracheal wall: slough, necrotic tissue.

Tissue KOH microscopy showed fungal elements suggestive of mucormycosis and was started on liposomal amphotericin after test dose and Tab Posaconazole 300 mg BD for first day followed by OD. She was then taken up for a formal tracheal debridement on POD 3. On the next day, her throat pain had drastically reduced. She was slowly started on oral feeds and her RT was removed. Histopathology report showed necrotic tissue with fungal structures (Figure 4) suggestive of mucor species. Serial flexible evaluation was done showed left VC fixed and right VC mobile not compensating. Flexible passed through stoma- slough seen in the infrastomal region, partially removed. Lower airway was healthy. She received a cumulative dose of 5 grams of liposomal amphotericin B. She was discharged after 26 days with the tracheostomy tube and IV line insitu. She was prescribed Inj Dalacin and to continue Tab Posaconazole.

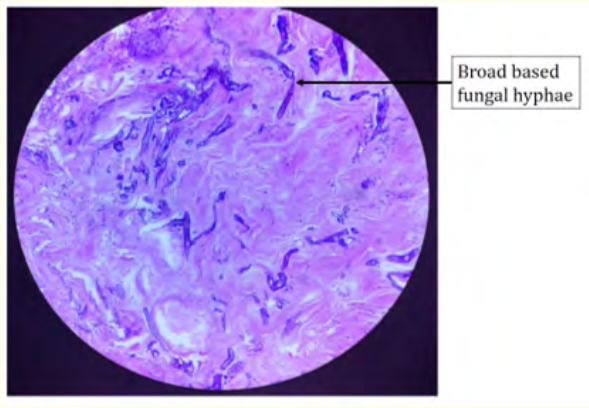

Figure 4: HPE: Fungal filaments with features suggestive of mucor species.

During her follow up visits, she was noted to have granulation tissue around her tracheostomy site, hence she was readmitted and taken up for tracheoscopy under GA- suprastomal granulation tissue was excised, tracheostoma widened and tracheostomy tube removed and a No 10 T tube was inserted as suprastomal tracheal wall collapse (Figure 5) was noted intra-op. Flexible videolaryngoscopy done on POD 9 -showed left VC palsy, right VC mobile. She was discharged on POD 10, with T- tube *in situ*.

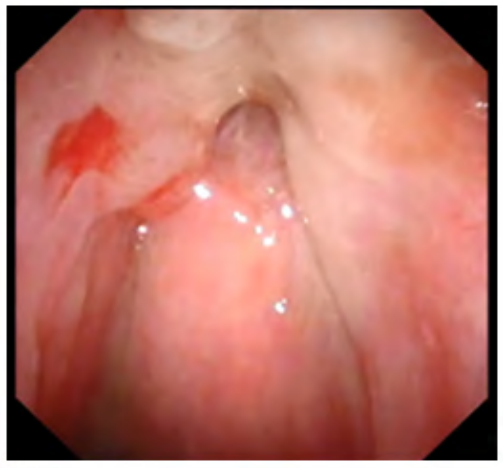

Figure 5: Supra stomal collapse of trachea.

On follow up after 2 weeks flexible laryngoscopy showed T tube touching inferior surface of vocal cord with vocal cord edema

and glottic granulations. She was admitted and taken to the OR- T tube was removed, small granulation tissue noted in anterior and posterior walls of subglottis (suprastomal) granulations removed and sent for HPE. Tracheostomy tube 7 with cuff inserted- this was later changed to an uncuffed No 6 portex tube before discharge. On subsequent OPD follow-up, her vocal cords healed well and she was decannulated. Her voice improved as her right cord was compensating well. As of today, she is doing well.

Discussion

Mucormycosis, although rare, has a high mortality and hence poses a significant threat to immunocompromised patients. Although in India, even the incidence of the disease in immunocompetent people is higher at 3-26%. This is 18–19% globally. The higher incidence in India has been attributed to uncontrolled diabetes mellitus which is rampant in this country [3,7-9]. 'Unmasked diabetes' as seen in our patient, contributes to a significant proportion of people diagnosed with mucormycosis. This is due to the lack of routine health checks at regular intervals, as reported by several studies – 24% in South India [10], 43% in North India [11] and 40% in Western India [12]. Thereby leaving the diabetes undetected and the patient prone to severe infections like mucormycosis. The uncontrolled sugars also make the management and control of the disease far more difficult. It also prolongs the recovery time, as eventually the infection itself contributes to the rising blood sugar values, thereby becoming a self-perpetuating problem.

Diagnosis at an early stage can be challenging due to the rapid spread of fungal hyphae. Our case represents one of the various respiratory manifestations of the disease, which usually presents as a pulmonary infection associated with a trachea-bronchial spread. However an isolated tracheal involvement as seen in our case makes this a rare one. The clinical presentation of pulmonary and respiratory tract mucormycosis is non-specific. These include cough, dyspnea, haemoptysis, fever. This is similar to the presentation of our patient, apart from the fact that she also had severe odynophagia and tenderness on palpation of the laryngeal framework externally.

Direct examination, biopsy and culture form the pillars of diagnosing this disease, as the presentation can be non-specific and hence aggressive evaluation essential for an early diagnosis and treatment. An incidentally noticed vocal cord palsy on flexible videolaryngoscopic examination is what drove us to a more aggressive evaluation, finally culminating in an early diagnosis of the disease in our patient and therefore, a timely management. Infact, had the vocal cord palsy not presented itself as it did- there was a high chance of missing the diagnosis early on, leading to further spread of the disease.

Recent recommendations strongly support an early surgical treatment for mucormycosis in addition to systemic antifungal treatment [13]. Tracheostomy was done for our patient within 2 days of presentation and intraoperative samples were sent for tissue KOH and biopsy- which confirmed the diagnosis. Angioinvasion and thrombosis of vessels is a hallmark of mucormycosis, leading to necrosis of tissues [14,15]. Therefore, tissues infected with mucor characteristically appear crusted and necrotic. Immune cells as well as antifungal medication are unable to penetrate necrotic tissue, making medical management alone inadequate. Liposomal amphotericin B (L-AmB) is the recommended first-line treatment for mucormycosis at a dose of 5–10 mg/kg/day [13,16]. Posaconazole is recommended as salvage [17] and maintenance therapy. It is not recommended as first-line therapy. As adjunctive modalities, deferasirox has been tried but it showed no significant benefit [18]. Hyperbaric oxygen therapy, although not recommended in routine treatment did show some benefit in diabetic patients [13]. Our patient was treated with Inj Liposomal Amphotericin B, IV Meropenem, IV Piptaz, apart from aggressive control of blood sugars. Her progress was monitored with serial video laryngoscopic examinations and multiple timely tracheoscopic debridements.

Fallahi., *et al*. described two cases of airway obstruction caused by a necrotic fungal mass in the larger airways. This presentation of acute respiratory failure must also be kept in mind in patients with pulmonary mucormycosis [19]. Ansari., *et al.* reported a case of tracheal mucormycosis with associated inflammation of the oesophagus, therefore the possibility of oesophageal involvement and tracheao-oesophageal fistula formation must be considered, especially in patients with severe odynophagia. Insertion of an RT early, so as to keep the patient nil-by-mouth is also to be considered as a part of management [20].

Chaddha U., *et al.* (2017) reported a case of tracheal mucormycosis with necrosis within the trachea [21], and Munjal M., *et al.* (2021) reported a case of laryngeal mucormycosis, with vocal cord edema [22]. Both patients presented with non-specific symptoms like dyspnea and mild dysphagia with hoarseness of voice. Both patients were treated with Inj Amphotericin B and underwent a tracheostomy, without any other surgical intervention. Therefore, it is possible to get away without multiple debridements in certain patients as long as the airway is protected early and adequate antifungal treatment is given.

Although there were several moments where we felt her condition was deteriorating- she continued to slowly, but progressively improve. We were able to downsize her tracheostomy tube and later insert a T-tube, and discharge her in oral Posaconazole maintenance. Post – discharge, she continued to improve drastically, regaining her voice, her tracheal mucosa healed well and we were able to decannulate her.

Conclusion

In conclusion, mucormycosis is an aggressive and life threatening infection, with a poor survival rate and a challenge to treat. The 3 cardinal elements of management are- control of risk factors, antifungal treatment and early surgical intervention, simultaneously. We attribute the survival of our patient to the aggressive evaluation, early diagnosis, timely airway intervention and proper medical management of the patient. Laryngotracheal mucor should always be ruled out in immuncompromised patients who present with vague airway symptoms- even as vague as 'throat pain'.

Conflict of Interest

None.

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