

Invasive Fungal Pharyngitis in a Pediatric Bone Marrow Transplant Recipient

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Abstract

Objectives:

1. Describe an atypical presentation of invasive fungal disease.
2. Discuss treatment options for invasive fungal pharyngitis.

Introduction: Invasive fungal disease of the head and neck is a potentially fatal infection most commonly seen in immunocompromised patients. Prognosis is poor even in the setting of combined surgical and medical treatment, with over 50% mortality rates seen in invasive fungal sinusitis.

Methods:

Review of the first successfully treated pediatric case of invasive fungal disease of the pharynx.

Results:

A 10-year-old female with severe aplastic anemia 14 days after matched sibling bone marrow transplant developed throat pain, and CT neck showed a rim-enhancing parapharyngeal mass. The patient was taken to the OR for evaluation and I&D of peritonsillar abscess, without return of purulence intraoperatively. Cultures grew *Rhizopus* species on postoperative day 2, and amphotericin B and posaconazole were initiated. After discussion with family, infectious disease and transplant services, extensive debridement and tracheostomy was performed. Intraoperatively, extensive necrosis of the palate and tongue base without invasion of the prevertebral fascia was observed. This patient had achieved successful neutrophil engraftment and all additional immunosuppressive medications were discontinued to allow for faster immune recovery. After a 4.5 month ICU and 2 month inpatient floor course of intravenous antifungal therapy and repeated negative biopsies, the patient was discharged without evidence of recurrent disease.

Conclusions:

Invasive fungal disease is most common in the sinonasal region, but alternative sites of disease must be considered in immunocompromised patients presenting with atypical symptoms. A multidisciplinary team approach and immediate aggressive treatment may be lifesaving.

Introduction

- Invasive fungal disease is a feared complication in immunocompromised patients, with mortality >50%.
- In adults, extra-nasal sites have been described, including larynx, trachea, thyroid, periorbita, and adenoids
- There are no reports of invasive fungal disease outside of the paranasal sinuses in the pediatric population.



Figure 1: Axial CT showing right parapharyngeal collection

Discussion

- Treatment strategy was based upon existing literature for invasive fungal sinus disease. Unclear duration of antifungal treatment with positive surgical margins.
- High index of suspicion must be maintained for extra-nasal sites of invasive fungal disease in immunocompromised patients. Delay in diagnosis has been associated with poor prognosis in adult studies.
- Treatment of invasive fungal pharyngitis may lead to velopharyngeal insufficiency, airway obstruction and dysphagia, with rehabilitation and reconstruction required after resolution of the infection.

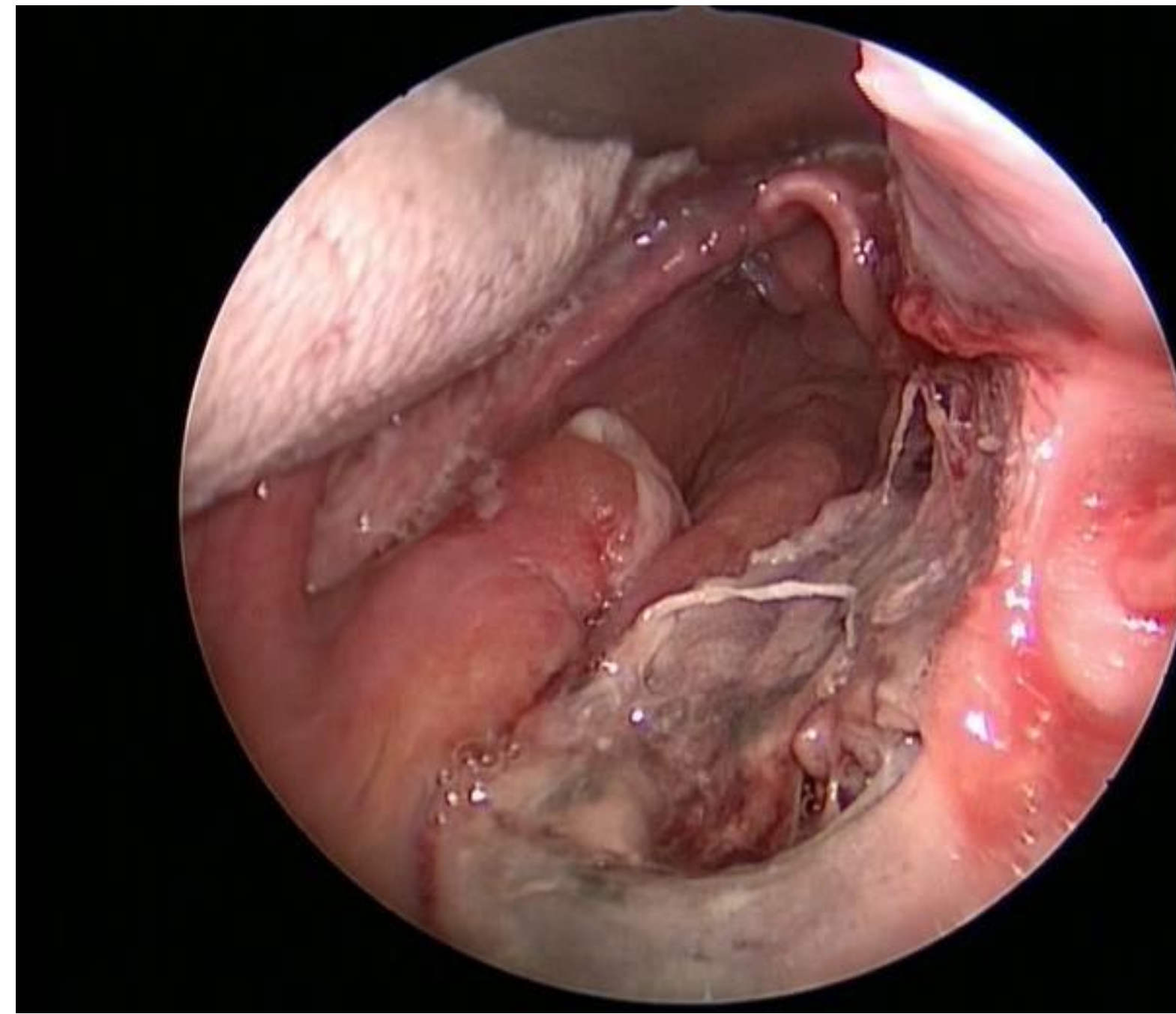


Figure 2: Right pharynx prior to resection

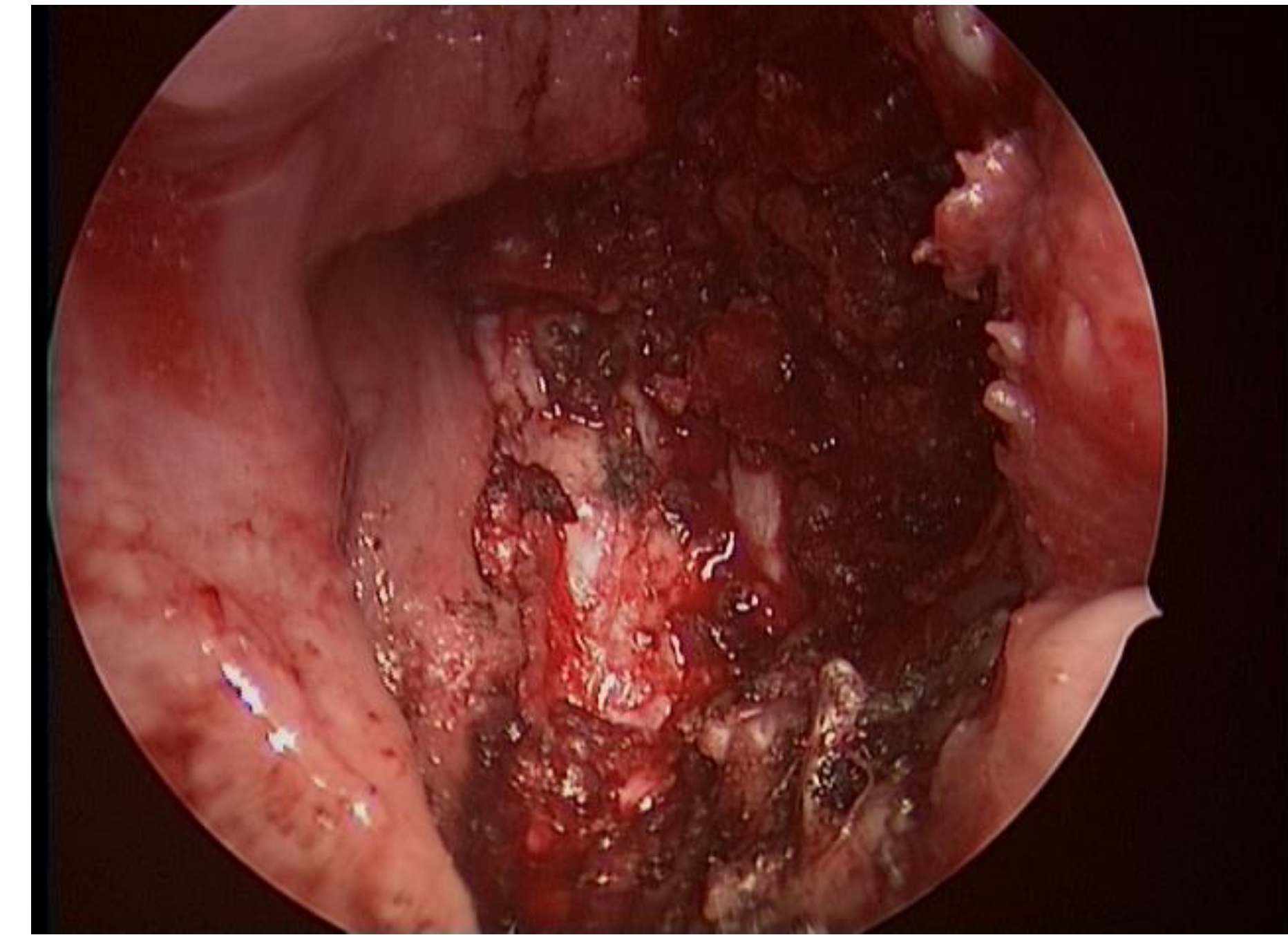


Figure 3: Right soft palate and pharyngeal defect

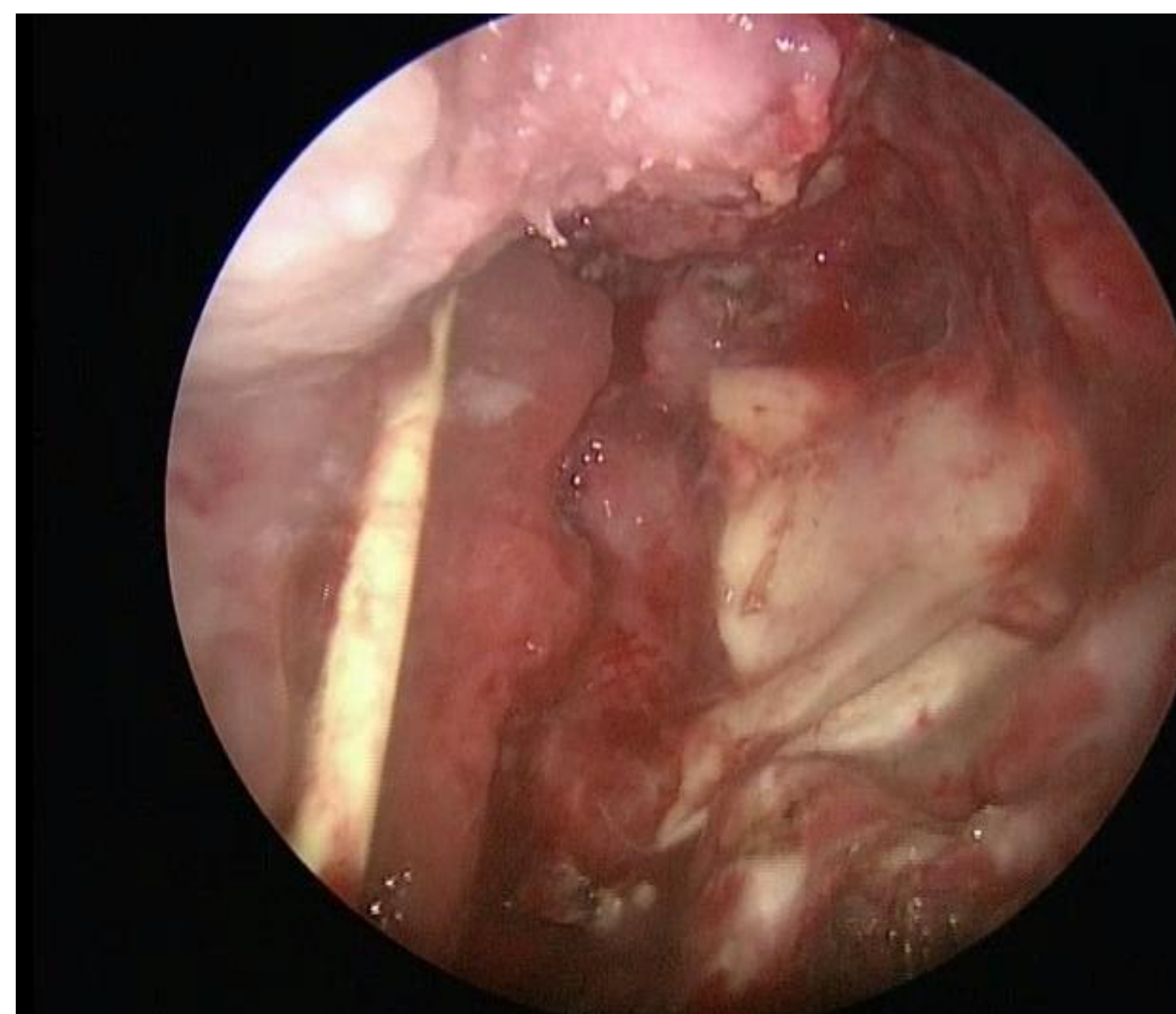


Figure 4: AlloDerm covering carotid artery

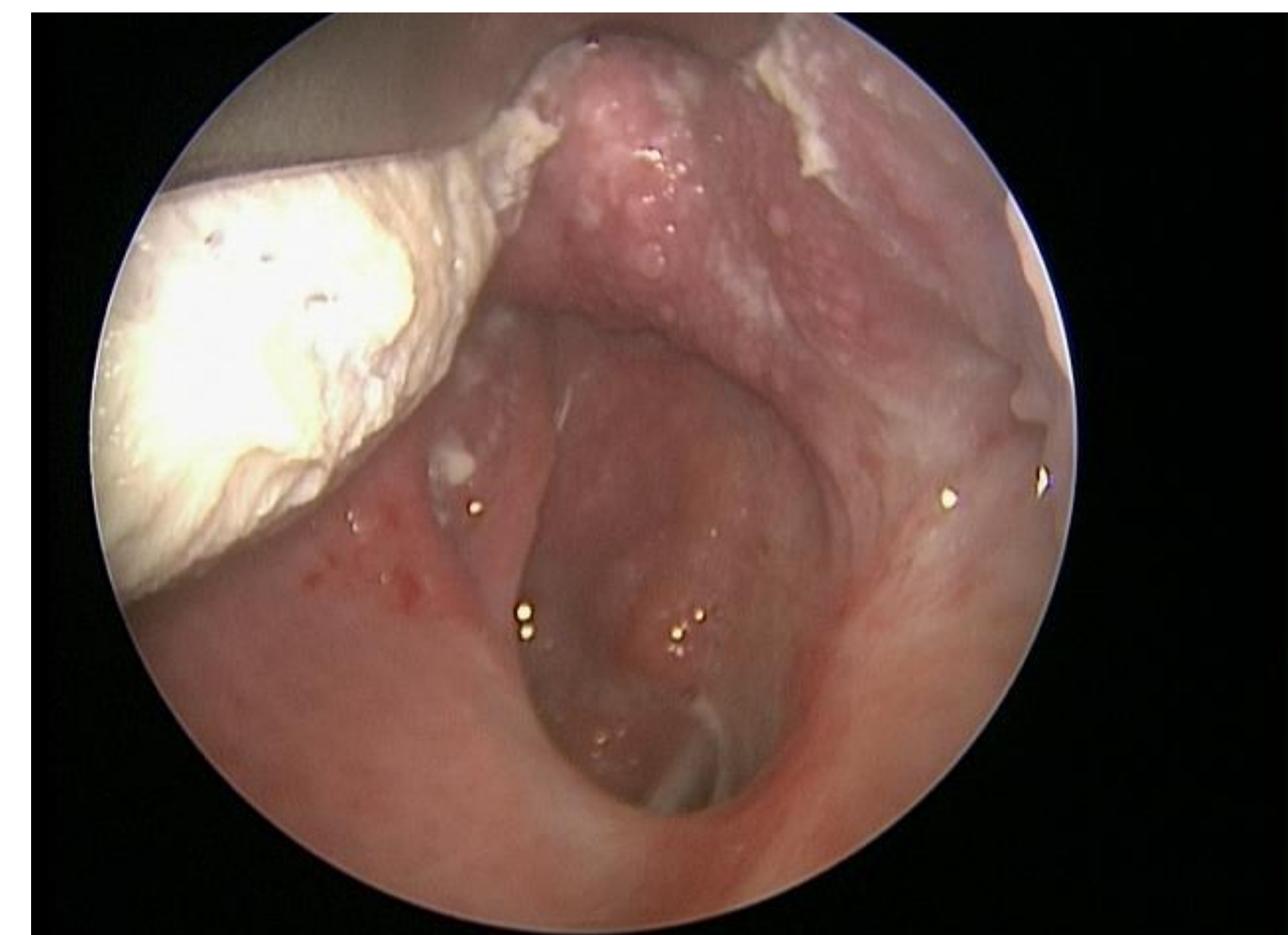


Figure 5: Healed surgical site with pharyngeal stenosis

Case Report

A 10-year-old female with aplastic anemia s/p bone marrow transplantation developed fever associated with otalgia and throat pain. A CT scan was obtained, showing a fluid collection in the right parapharyngeal space (Figure 1). Due to persistent symptoms after needle aspiration and IV antibiotics, patient underwent operative exploration which showed devascularized tissue with no fluid collection. Fungal cultures returned with *Rhizopus arrhizus* and there was evidence of fungal invasion on pathology.

Serial resections and tracheostomy were performed, including the right soft palate, right tonsil and pharyngeal musculature to the carotid artery, and posterior pharyngeal wall to the prevertebral fascia. The right tongue base was resected serially, but this area and tissue overlying the carotid artery were never able to be cleared of fungal disease surgically (Figures 2,3). There was no evidence of sinus or nasal involvement. AlloDerm was placed over the carotid artery, but no further primary reconstruction was completed (Figure 4).

Her immunosuppression regimen was adjusted and she was maintained on antifungal therapy with amphotericin B liposome and posaconazole, with discontinuation of amphotericin after 6 weeks due to kidney injury. With positive margins at the tongue base, she continued on antifungal therapy for a total of 18 months.

She required prolonged ventilatory support but was achieved engraftment and was discharged with a tracheostomy in place after 180 days.

She has a mild pharyngeal stenosis, however tolerates an oral diet and uses an obturator for velopharyngeal insufficiency (Figure 5). She has a grade IV laryngeal exposure due to scarring, but obstructive sleep apnea has resolved after contralateral tonsillectomy and adenoidectomy. She is capping her tracheostomy, with plans for decannulation.

Conclusion

- Early diagnosis and prompt surgical debridement remains the best treatment for invasive fungal disease, including extra-nasal sites of infection.

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