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Case Report

Management of Necrotizing Epiglottitis and Subsequent Cervical Necrotizing Fasciitis with Pharyngocutaneous Fistulae: A Pediatric Case Report and Literature Review

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ABSTRACT

Necrotizing epiglottitis (NE) is a rare and serious variant of acute epiglottitis. It is an airway emergency and a potentially fatal infection of the supraglottis and surrounding structures. Most documented cases are reported in immunocompromised adults. A retrospective chart review of the case in review was performed. A review of the literature using Pubmed and Embase databases was performed to examine the documented cases, epidemiology, microbiology, treatment, and prognosis of NE. Only six published cases of pediatric of NE were identified on review of the current literature, only one of whom was immunocompetent. Furthermore, there are no published reports of any aged patient with NE complicated by cervical necrotizing fasciitis and pharyngocutaneous fistulae. This is the first published case of a pediatric patient with NE that progressed to cervical necrotizing fasciitis with bilateral pharyngocutaneous fistulae. Previously published clinical experience mandates the rapid recognition and surgical management of necrotizing epiglottitis with cervical necrotizing fasciitis. In this case, a combination of repeat debridement and negative pressure wound therapy successfully treated the infection and maintained the integrity of the remaining laryngeal and cervical neck tissues.

Keywords: Necrotizing Epiglottitis, Cervical Necrotizing Fasciitis, Pharyngocutaneous Fistula, Negative Pressure Wound Therapy, Vacuum-assisted Wound Closure

1. Introduction

Acute epiglottitis (AE) is a life-threatening infection that causes profound swelling of the upper airways, which can lead to asphyxia and respiratory arrest[1]. This infectious entity has become rare with the advent of Haemophilus influenzae type b (Hib) vaccination. The epidemiology of the disease has shifted from one plaguing mainly children to one more prevalent among

adults and featuring varied pathogens. A more uncommon and serious variant of AE is necrotizing epiglottitis (NE), a rapidly progressive soft tissue infection more often described in immunocompromised adults. NE is exceedingly rare in children with only six cases reported in the literature[1-5]. There are no reported cases of NE progressing to pharyngocutaneous fistulae.

Here, we present a unique clinical case of a 15-year-old male diagnosed with NE. This work emphasizes the importance of rapid recognition of infection, airway management, surgical debridement and subsequent negative pressure wound therapy.

2. Methods

The Children's Mercy Institutional Review Board provided exemption status for this case presentation. Informed consent was obtained from both parents before undertaking the recounting of this case. We performed a review of the electronic medical record. A literature review was performed through December 2023 using Pubmed and Embase databases to understand the epidemiology, microbiology, treatment, and prognosis of NE. Keywords and Medical Subject Headings (MeSH) were used to identify relevant articles.

3. Case Presentation

An otherwise healthy 15-year-old male presented to our tertiary care center with fever, sore throat, and odynophagia for 2 days, though no airway compromise. He was triaged by the Emergency Department (ED), noted to be rapid streptococcus and influenza A&B negative and discharged with symptomatic management. He returned to the ED the next day with worsening symptoms. A contrasted neck computed tomography (CT) scan showed supraglottic edema concerning for epiglottitis without evidence of abscess or subcutaneous emphysema **(Figure 1A)**. Vital signs showed temperature of 39.4C, tachycardia, normotension. Shortly after returning from the scanner, he began to decompensate from a respiratory perspective. On ENT arrival to bedside, the patient was in acute respiratory distress. Exam was notable for diaphoresis, tripoding, persistent inspiratory stridor, drooling, and anterolateral cervical neck edema. Flexible fiberoptic laryngoscopy revealed significant supraglottic edema with devitalized arytenoid mucosa, more prominent on the right **(Figure1B)**. The vocal folds were mobile bilaterally. The patient was emergently taken to the operating room (OR). He was orally intubated with a 6.0 cuffed ETT via Seldinger technique over a rigid telescope.

Initial working diagnosis was acute epiglottitis with septic shock. The patient was subsequently admitted to the pediatric intensive care unit (PICU). Medical management included intravenous (IV) vancomycin, ceftriaxone, clindamycin, crystalloid fluids, and vasopressors. Blood cultures were obtained on day1 and showed Streptococcus dysgalactiae bacteremia. Initial laboratory findings demonstrated an absolute neutrophil count (ANC) of 0, which was attributed to severe disseminated bacterial infection. The patient was ultimately weaned from vasopressors and his ANC slowly rebounded, but his fever persisted and the appearance of his anterior neck edema worsened. A repeat CT on day4 and showed similar appearance of the epiglottis with complete airway effacement, ill-defined hypodense area in the right neck inferior to the parotid gland extending inferiorly along the sternocleidomastoid (SCM) muscle without notable fluid collection, extensive cervical cellulitis with maintenance of cervical fascial planes, and paranasal sinus disease **(Figure 1C-F)**.

Given persistently high fever in the setting of sinus disease and recent neutropenia, the patient was taken to the OR on day5 for middle turbinate biopsy to rule out invasive fungal sinusitis. ANC was then 2.95. The patient was started on IV amphotericin until pathology returned as inflammatory changes without angioinvasion or necrosis. Cultures from the middle meatus grew methicillin-resistant Staphylococcus aureus (MRSA), alpha streptococcus species, Cutibacterium acnes, and Candida tropicalis.

Given the patient's continued fever and progressive anterior neck swelling, concern remained for emerging neck abscess. Daily neck ultrasounds (US) were negative for fluid collection, including one obtained the morning of the patient's third CT scan, which revealed several rim-enhancing fluid collections: one along the left anterior cervical triangle extending inferiorly along the left SCM and anterior to the left thyroid lobe measuring 8.7x5.3x2.0cm, another within the left retropharyngeal soft tissues measuring 3.3x1.2x0.8cm **(Figure 1G,H)**. Several other areas of low attenuation concerning for fluid accumulation were noted, particularly in the right neck. Additionally, foci of air in the anterior neck were noted in associated with the fluid collections bilaterally. The patient was taken for incision and drainage of these abscesses. Upon entering the neck, grey foul-smelling fluid and frankly necrotic tissue were encountered. Concern became high for necrotizing fasciitis. Careful debridement was performed and the neck was thoroughly irrigated. A right pharyngotomy at the thyrohyoid membrane was discovered and confirmed on direct laryngoscopy. This finding was attributed to necrotizing infection eroding through his neck into his pharynx, as there was obvious necrosis of the lateral hypopharyngeal tissue, epiglottic tip, and right aryepiglottic fold and arytenoid. Bilateral neck incisions were packed with betadine-soaked kerlix and kept open.

Wet-to-dry packing changes were performed twice daily for 4 days. On day13, because of poor clinical progress, vacuum-assisted closure (VAC) device was placed on the right neck wound. The patient was taken to the operating room for debridement, washout, and wound VAC application. The right pharyngotomy was slightly increased in size. For the first time, a left-sided pharyngotomy was noted in a similar location at the thyrohyoid membrane. Additionally, the infection had partially eroded through the left carotid sheath, leading to focal exposure of the carotid artery and jugular vein. The left neck was repacked and the patient returned to the ICU with carotid artery precautions.

On day16, again because of poor clinical progress, a wound VAC was placed in the left neck taking care to protect the carotid artery. On day17, the patient returned to the OR for open tracheotomy. A 6.0 cuffed Shiley was placed, avoiding communication between the tracheostomy site and bilateral neck wounds. The wound VAC sponges were routinely changed every other day at bedside under moderate sedation. By day36, the right neck defect was noted to be shallow with a bed of healthy granulation and it was closed primarily. The patient was transferred out of the ICU on day37. ENT continued to perform bedside wound vac changes of the left neck until it was felt the defect was suitable for closure. On day47, the left neck was closed primarily.

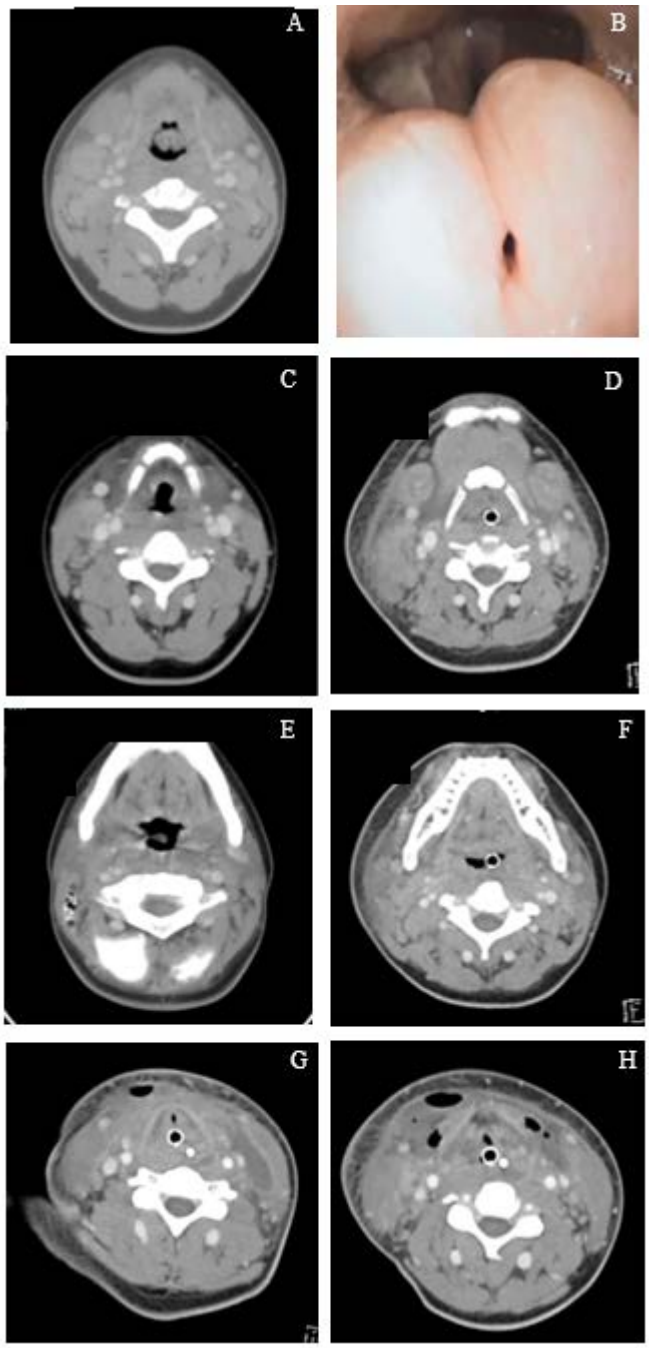

Figure 1: (A) CT scan, axial cut, demonstrating epiglottic swelling (Day 0). (B) Endoscopic appearance (videolaryngoscope) of the epiglottis immediately prior to initial endotracheal intubation (Day 0). CT scan: axial cuts at the level of the hyoid bone (C,D) and at the level of the mandibular symphysis (E,F) comparing days 1 to 4. Notably, at day 4, facial planes are maintained and there is still no drainable fluid collection. Increase in fat stranding, soft tissue edema, and myositis and cellulitis right>left. CT scan: axial cuts demonstrating two rim-enhancing fluid collections in the left neck (G) and numerous foci of air throughout the neck (H) on day 8.

The patient's severe dysphagia remained a hurdle. Flexible laryngoscopy on day36 demonstrated decreased mobility of the right TVC, mild tissues loss of the right arytenoid, and frank aspiration of secretions. The remaining epiglottis demonstrated moderate tissue loss at the tip and appeared partially scarred to the base of tongue **(Figure 2)**. A video swallow study performed on day42 showed frank aspiration of all consistencies of barium. Dysphagia was thought secondary to the combined effects of general deconditioning, laryngeal tissue loss, decreased laryngopharyngeal sensation, right vocal cord paresis, and/or relative immobility of the epiglottis secondary to scarring. On day56, the patient underwent right TVC injection with Prolaryn Gel™. Throughout this time, the patient remained strictly NPO and, though he no longer required his tracheostomy for airway protection, he was routinely utilizing it for pulmonary toilet. Several days after injection medialization, flexible laryngoscopy revealed improved glottic closure. The patient was permitted small sips of water during swallow therapy. A bedside swallow evaluation on day77 revealed significant improvement in his dysphagia relative to the swallow study performed on day58. The patient continued swallow therapy and ultimately, on day93, he was decannulated and his gastric tube removed prior to discharge. At his 3-month follow-up appointment, he showed no voice, breathing, or swallowing issues. Laryngoscopic findings seen in **(Figure 2)**.

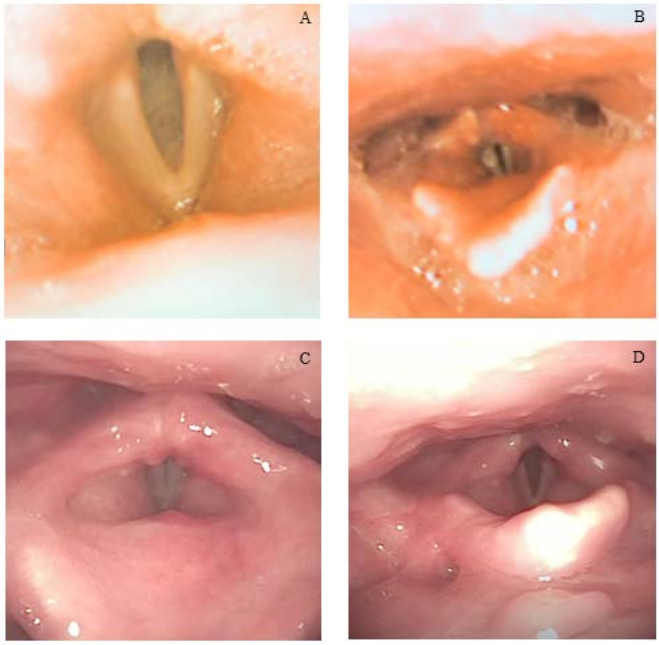

Figure 2: Flexible laryngoscopic examination on day 36 (A, B) versus 3 months after discharge (C, D). (A) demonstrated decreased mobility of the bilateral true vocal cords, vocal cord bowing likely secondary to deconditioning, and associated glottic insufficiency. (B) Presence of excess secretions in the vallecula as well as bilateral piriform sinuses, scarring of the epiglottis to the base of tongue, and significant epiglottic tissue loss, can also be appreciated. (C) Bilateral true vocal cord mobility and near complete closure. (D) Minimal scaring associated with the epiglottis though appreciable tissue loss, particularly on the right side.

4. Discussion

Though historically a pediatric disease, acute epiglottitis has become more prevalent in adults. Still, it remains a relatively rare infectious entity. Rarer still is NE. Most NE cases have been documented in immunocompromised patients[2-4,6-9] in association with active viral infections,9-11 or in the setting of comorbidities like uncontrolled diabetes[12,13]. Only a handful of NE cases have been documented in the pediatric population[1-5]. There is one case report of NE in an immunocompetent child[5]. There are no documented cases of pharyngocutaneous fistulae as an advanced complication of NE.

Our case provides the opportunity to examine a few key aspects of the workup, management, and treatment of NE. In acute bacterial epiglottitis, edema and erythema of the supraglottic structures, in particular the epiglottis, are classic laryngoscopic findings. In NE, pseudomembranes and areas of necrosis are present as well[14]. CT findings are typically similar for both[14]. Infections that progress to cervical necrotizing fasciitis often demonstrate obliteration of cervical fascial planes, which is all but pathognomonic for the diagnosis. As demonstrated in this case however, this imaging finding may take days to emerge. Other CT findings consistent with cervical necrotizing fasciitis include platysmal, SCM, and strap muscle enhancement, thickening and infiltration of superficial and deep cervical fascia, fluid collections in multiple neck compartments, and rarely, the presence of gas[15]. The possibility of delayed radiologic findings of necrotizing fasciitis underscores the point that this entity remains a clinical diagnosis and should be suspected in a patient with pain out of proportion to exam, loss of sensation, hemodynamic instability, or other signs of overwhelming infection.

Surgical debridement of cervical necrotizing fasciitis should be undertaken expeditiously and aggressively, but also with caution to preserve as much laryngeal tissue as possible with the goal of maximizing functional recovery. This patient suffered near complete necrosis of the superior half of the epiglottis and partial necrosis of the right arytenoid. Despite these findings, the patient's glottic function and swallowing were recovered once acute infection resolved and tissue healing occurred. Significant laryngeal necrosis with subsequent recovery has been described in other NE patients[5].

Management of the lateral neck infections proved very difficult in this case and did not respond well to repeated debridements with either penrose drain or wet-to-dry dressings. We found great success with employment of wound vacuum-assisted closure devices, with rapid improvement of the cervical infections, cessation of further necrosis, as well as shrinkage and ultimately closure of the pharyngotomies. The patient additionally suffered no ill effects of placement of the VAC on the exposed vessels of the carotid sheath in the left neck, a consideration which produced initial hesitancy in its deployment.

5. Conclusion

Pharyngocutaneous fistula as a complication of NE and associated cervical necrotizing fasciitis has not been described previously. Regarding imaging, CT may lag clinical progression in this specific disease process. US may not be a reliable screening technique for cervical neck abscesses or for cervical necrotizing fasciitis in particular. In a critically ill patient for which there is high clinical suspicion for developing abscesses, CT neck with IV contrast may be the better modality for surveillance. Negative pressure wound care systems may be useful in this setting to promote healing of tissues and closure of fistulae, reduce OR debridments, and create a healthy wound bed for subsequent closure.

6. Declarations

Ethical consideration: Our institution does not require ethical approval for reporting individual cases or case series.

Consent to participate: Written informed consent was obtained from a legally authorized representative for anonymized patient information to be published in this article.

Consent for publication: As above. Written informed consent was obtained from a legally authorized representative for anonymized patient information to be published in this article.

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