

Acute Epiglottitis in an Adult Leukemia Patient An Unusual Presentation

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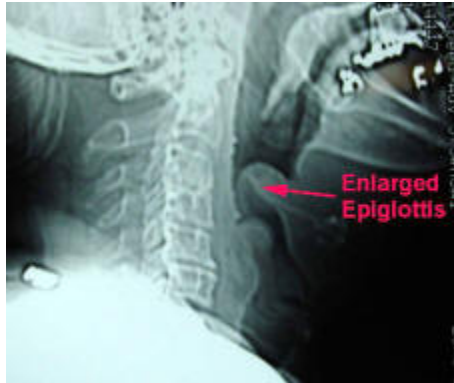
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Abstract: Epiglottitis is an acute and life threatening infection of the epiglottis and surrounding airway. The peak incidence of the disease has traditionally been identified as occurring in young children. We report a case of a 50 years old lady who presented to us with complaints of pain on swallowing and neutropenia. Further investigations lead to the diagnosis of epiglottitis in a case of promyelocytic leukemia. A review of literature lead to the identification of 18 cases of acute epiglottitis in cancer patients, 15 of them in adults, 16 patients had hematologic malignancies. Streptococcus pneumonia and Candida albicans were the most frequent organism identified. Early recognition and aggressive supportive care is required for successful management.

Introduction: Epiglottitis is defined as an acute infection of the supraglottic larynx which may require an emergency airway management. Haemophilus influenzae Type B (Hib) accounted for 75% to 90% of the cases in the prevaccination era.¹ In 1985, a polysaccharide vaccine was introduced against Hib, followed in 1987 by a conjugate vaccine which has drastically reduced the incidence of H. influenzae infections including epiglottitis.² Recent reports have suggested that the disease occurs in adults as well, with evidence of increasing incidence in adults.^{3,4} As the frequency of Hib disease has decreased, the epidemiology of epiglottitis has shifted to other bacterial, viral, or combined viral-bacterial infections, and noninfectious aetiologies. Organisms like Streptococcus pneumoniae, other Streptococcus species, Staph aureus, Moraxella catarrhalis, Pseudomonas species, Klebsiella pneumoniae, Pasturella multocida, and Nisseriae species are known to cause this disease.⁵ Other causes include viruses like herpes simplex, parainfluenza, varicella zoster, EB virus, fungus like Candida albicans, trauma and thermal injuries. Adults affected by epiglottitis are often immunocompromised, suffering from Malignancies, HIV infection, Diabetes mellitus or hematologic disorders. Steptococcus pneumoniae and Candida albicans are the most commonly isolated organisms in patients suffering from hematologic malignancies.⁶ Eighteen cases of acute epiglottitis in malignancies has been reported in Literature. The majority of them were in adults (15 out of

18) and none were associated with H influenzae. Early recognition and aggressive supportive care are required for successful management.



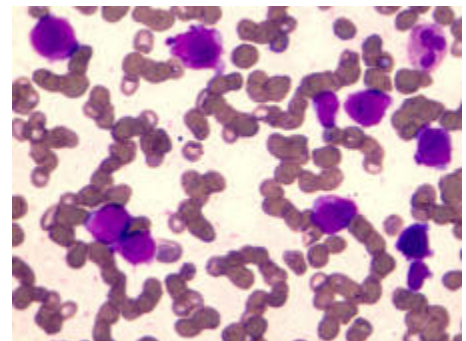
Emergency room diagnosis of a patient is often due to symptoms of airway obstruction. Fiberoptic laryngoscopy (see case report below) or soft tissue lateral x-ray neck (see picture on right) are the other preferred methods of diagnosis. Indirect laryngoscopy should be done with caution since patient gagging may worsen or participate catastrophic airway obstruction.

This case report describes a neutropenic patient with epiglottitis who was subsequently diagnosed as having acute promyelocytic leukemia. **Enlarged Pictures At The End of Manuscript**

Case Report: A 50 year old woman presented with a week long history of pain on swallowing and dysphagia. The odynophagia was gradually progressive and more for solids than liquids. Four months back, she had been diagnosed as having tuberculous lymphadenitis and started on anti koch's treatment (AKT). The AKT had to be stopped three days before she came to us because of persistent neutropenia. On examination, she was otherwise normal with no temperature or dyspnea. On palpation, she did have a slight tenderness of the laryngeal frame work. We were unable to find a definitive cause of pain in the throat and performed an indirect laryngoscopic examination which revealed a 'cherry-red' epiglottitis with slough at the base of the epiglottis. Fiberoptic laryngoscopy confirmed the diagnosis of epiglottitis (See Picture on Right).



She was admitted for observation and emergency airway management if such a situation arose. We sent a throat swab and routine haemogram(CBC) started her on Ceftriaxone. We did not give her steroids as she was a diabetic and was also immunosupressed. Her haemogram was repeated daily and it showed a persistent low white blood cell count (WBC) and platelet count. The throat swab revealed Beta hemolytic Streptococcus pyogenes sensitive to Cephalosporins. When her CBC did not improve even after three days of administering granulocyte-macrophage colony stimulating factor, we decided to do a bone marrow biopsy. This revealed that she was suffering from acute promyelocytic leukemia with thrombocytopenia (See Picture on Right -- Click to Enlarge).



She was started on induction chemotherapy, supported by multiple blood and blood factor transfusions. The disease went into remission on the 18th day. Her throat symptoms improved gradually on

antibiotics. Her AKT was restarted along with the chemotherapy. In the interim she also developed an opportunistic lung infection which had to be treated with Vancomycin. She was finally fit for discharge three weeks after hospitalization with normal blood counts. On follow up after one month, she had normal blood counts and was gradually recovering from the side effects of chemotherapy.

Discussion: Acute epiglottitis in adults is often referred to as supraglottitis since inflammation is generally not confined to the epiglottitis (as is the case with children) but also affects other structures such as the pharynx, uvula, base of the tongue, aryepiglottic folds, and false vocal cords. Diagnosis in adults may be delayed a sore throat and odynophagia are the only complaints and they do not have signs and symptoms of airway obstruction. Such patients may never seek medical attention or may be diagnosed as having pharyngitis and either recover without further intervention, have progressive symptoms or die at home.⁷ To reduce the possibility of delayed diagnoses, the clinician should suspect epiglottitis in patients with sore throat and odynophagia especially if the patient is immunosuppressed and the oropharyngeal examination is normal. All such adults should undergo an indirect laryngoscopic examination along with fiberoptic laryngoscopy to exclude the possibility of supraglottitis. Tenderness over the larynx should also raise the suspicion of this diagnoses. Lateral neck soft tissue roentgenogram can demonstrate swelling in the supraglottic region ‘Thumb Sign’ and is useful if laryngoscopy cannot be performed. A throat swab may or may not demonstrate the causative organism but is difficult to perform when the airway is compromised and should always be taken under controlled conditions for airway management. Blood cultures are often negative either due to previous antibiotic use or a viral cause.⁸ WBC counts are raised as a result of infection.

In our patient, her persistent neutropenia lead us to investigate her further for hematologic malignancy. Literature review showed 13% of adult patients with epiglottitis were immunocompromised, most of them had diabetes mellitus, HIV infection or full blown AIDS, chronic asthma. 18% of all such patients were due to hematologic malignancies.⁹ Chemotherapy and bone marrow transplant used in these cancers cause severe immunosuppression which also increase the risk of epiglottitis in these patients.

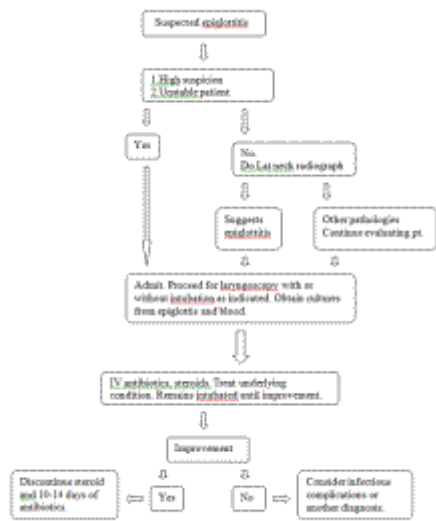


Fig 3: Suggested algorithm for the management of epiglottitis

The main consideration in management is airway maintenance. Patients without signs and symptoms of airway obstruction can be treated medically in a hospital unit with equipments and personnel available for airway management if required. A second or third generation cephalosporin is the most effective antibiotic against beta lactamase producing organisms and should be considered as initial antibiotic therapy. Simultaneous treatment of the underlying condition is mandatory. Corticosteroids have not proven in a prospective randomized trial to reduce the need for airway intervention or hasten recovery in adult acute epiglottitis. The algorithm shows the proposed decision tree for the management of epiglottitis. (See Figure on Right -- Click to Enlarge)

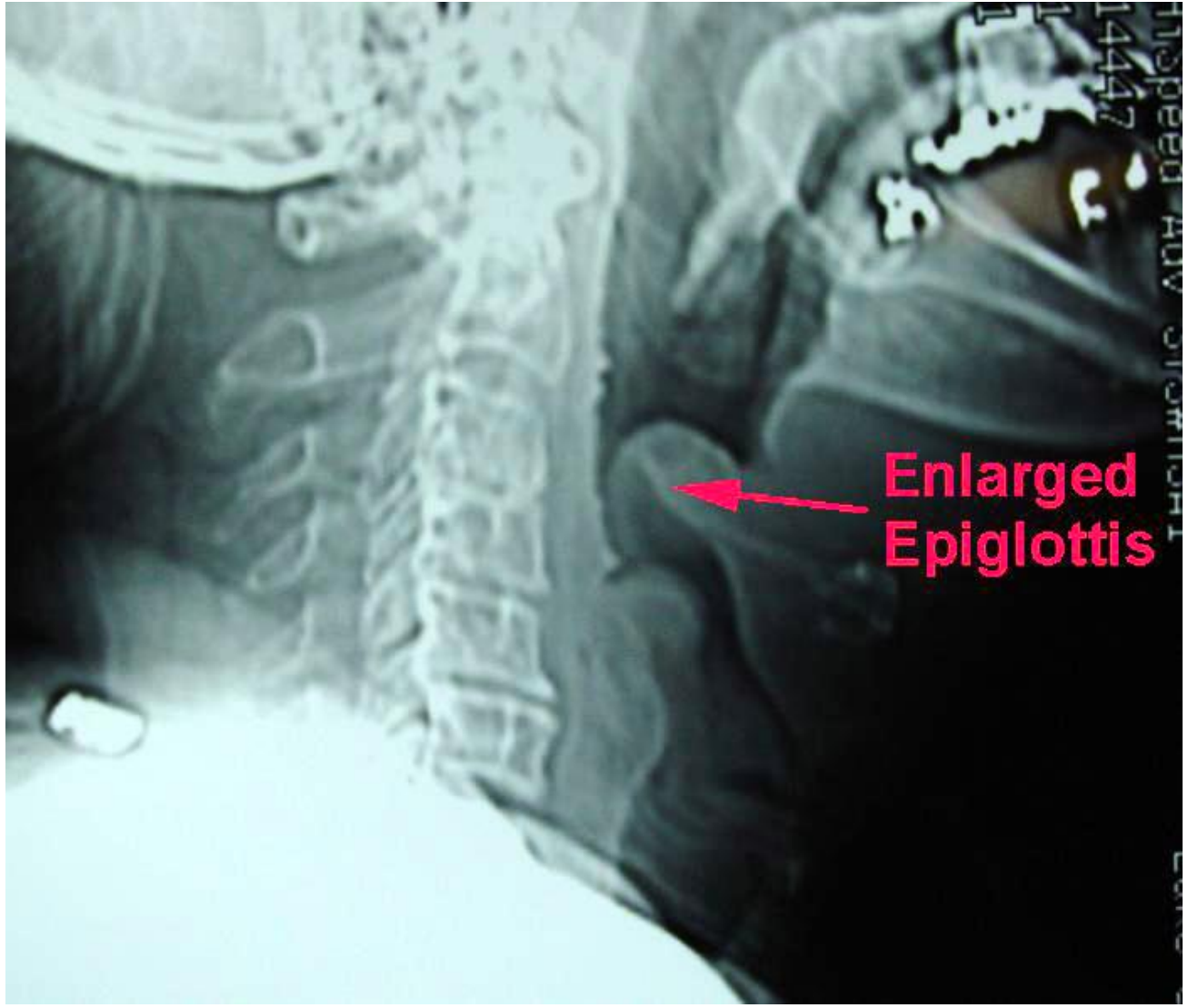
Conclusion: Adult epiglottitis may be more common than previously appreciated. We believe that an increased awareness of this condition with the widespread use of nasopharyngolaryngoscopy results in the identification of more

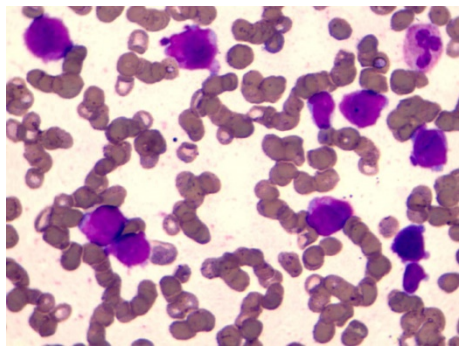
cases with a wide spectrum of disease severity. A strong suspicion of this condition in a symptomatic adult patient can help in early detection and thus avoid complications and fatalities.

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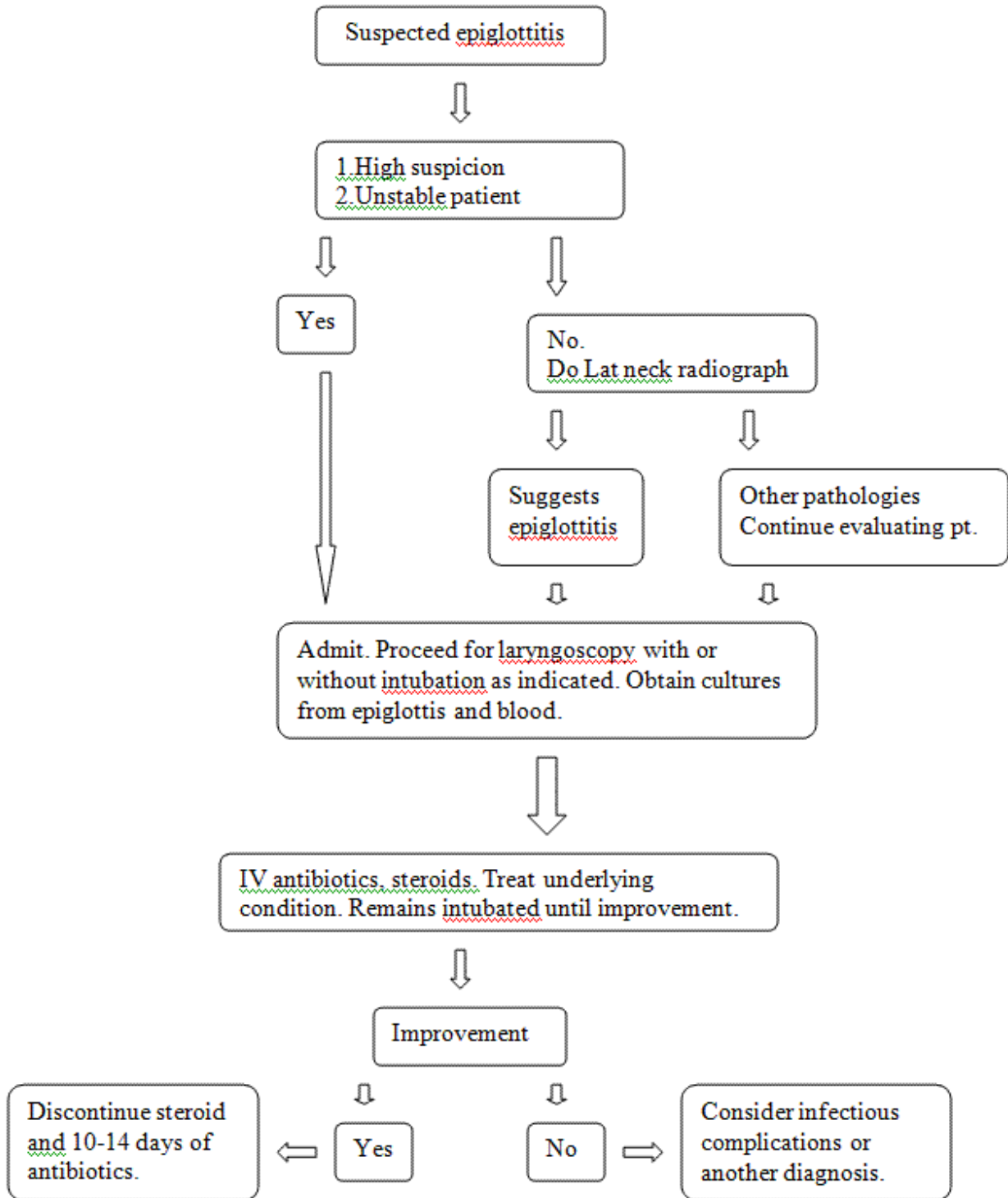


Fig 3: Suggested algorithm for the management of epiglottitis