



# Index of Suspicion

## 3 Sore Throat and Fever in a 4-year-old Boy

Katsuaki Kojima, MD,\* Melissa Rosenberg, MD,<sup>†</sup> B. Keith English, MD\*

\*Departments of Pediatrics and Human Development, College of Human Medicine;

<sup>†</sup>Department of Pediatrics, College of Osteopathic Medicine, Michigan State University, East Lansing, MI.

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### PRESENTATION

A 4-year-old previously healthy and fully immunized boy presents with a 2-day history of sore throat and a 1-day history of fever and drooling. He awakened today with worsening sore throat, drooling, and subjective fever. He complains of pain during swallowing and refuses oral intake. He has no cough, congestion, sick contacts, or recent travel. His prenatal, natal, and postnatal histories contain no significant information. His immunizations are up to date.

At initial presentation, his temperature is 39.6°C (103.4°F), heart rate is 160 beats per minute, respiratory rate is 22 breaths per minute, and oxygen saturation is 95% in room air. Physical examination reveals an alert and interactive boy who is drooling and appears uncomfortable. He is in the tripod position and has a muffled voice but no stridor, nasal flaring, or labored breathing. His chest is clear to auscultation bilaterally. His neck is supple and has no swelling, erythema, or tenderness. His throat is not examined. Findings on the remainder of the physical examination are unremarkable.

Results of laboratory evaluation include:

- White blood cell count 30,900/ $\mu$ L ( $30.9 \times 10^9/\mu$ L), with 66% segmented neutrophils, 22% banded neutrophils, 5% lymphocytes, and 7% monocytes
- Hemoglobin 13.9 g/dL (139 g/L)
- Hematocrit 39.1% (0.39)
- Platelet count  $473 \times 10^3/\mu$ L ( $473 \times 10^9/L$ )
- Erythrocyte sedimentation rate, 8 mm/h
- C-reactive protein, 3.2 mg/L (30.5 nmol/L)

Blood is drawn for culture. Neck soft-tissue radiography leads to the diagnosis.

### DISCUSSION

The soft-tissue neck radiograph reveals a “thumb sign.” After clinicians administer ceftriaxone and dexamethasone, the patient is flown to a university hospital. He is transported to the operating room for endotracheal intubation under general anesthesia. His epiglottis is visualized and is erythematous and swollen. Blood culture becomes positive for *Haemophilus influenzae* type f after 40 hours. Repeat blood culture is negative. He receives 14 days of antibiotic treatment with ceftriaxone. Immunodeficiency evaluation is not performed.

## Condition

*H influenzae* type b (Hib) was historically the most common cause of acute epiglottitis. Immunization against Hib was introduced in 1985, although initially it was not a conjugate vaccine and was not recommended for infants. The first Hib conjugate vaccine was licensed in December 1987 and the incidence of Hib epiglottitis declined by 80% to 90% after the introduction. The incidence of immunoglobulin A deficiency and immunoglobulin G subclass deficiency are reported to be higher in those who had Hib vaccine failure. Epiglottitis caused by other *H influenzae* types (a, f, and nontypeable), *Streptococcus pyogenes*, *S pneumoniae*, and *Staphylococcus aureus* (including methicillin-resistant strains [MRSA]) now comprise a larger proportion of cases compared to the prevaccine era. The median peak age of children with epiglottitis has increased from age 3 years to ages 6 to 12 years due to the change in etiology of the disease. Among the pediatric population, the incidence of acute epiglottitis was 0.5 cases per 100,000 children in the United States in 2006.

Acute epiglottitis is defined as an acute inflammation of the epiglottis and adjacent structures. The clinical presentation is characterized by rapidly progressing dysphagia, drooling, dysphonia, and distress (“the four Ds”). Other characteristic findings include high temperature, toxic appearance, and muffled speech, which is sometimes described as a “hot potato” voice. The typical patient presents in the tripod position of sitting upright and leaning forward with a hyperextended neck and chin thrust, which maximizes airway patency. Of note, children with epiglottitis lack a cough or hoarse voice, which are characteristic of croup.

## Diagnosis

The diagnosis is based upon visualization of a large, cherry red, swollen epiglottis or demonstration of epiglottal swelling (“thumb sign”) by lateral neck radiography. Blood cultures and cultures obtained from the surface of the epiglottis help identify the causative organism. Epiglottal cultures are positive in 30% to 70% of patients.

When a clinician has a high index of suspicion for epiglottitis based on clinical presentation, direct visualization of the epiglottis with laryngoscopy should be performed by a physician skilled in intubation in a controlled setting, such as an operating room or intensive care unit, with preparation for subsequent artificial airway placement.

Laboratory testing and radiography should not be performed on patients in whom epiglottitis is strongly suspected until the airway is secured. In this case, soft-tissue neck radiography was performed due to the unclear cause of his respiratory distress because he had been fully immunized. Agitation and anxiety may increase respiratory effort

and worsen respiratory distress. An experienced physician with skills in airway management and intubation should be present at all times.

Occasionally, other infections related to Hib, including pneumonia, cervical adenopathy, or otitis media, may occur concurrently with epiglottitis, but this is a rare finding.

## Treatment

Acute epiglottitis is a medical emergency that warrants immediate intervention to secure the airway regardless of the degree of apparent respiratory distress. Without an artificial airway, 6% of children die compared to less than 1% of those with an artificial airway. The airway should be secured in a controlled setting, either an operating room or intensive care unit, via orotracheal intubation, nasotracheal intubation, or less frequently, tracheostomy.

The choice of antibiotics depends on local resistance patterns. In general, a combination of a third-generation cephalosporin and an antistaphylococcal agent against MRSA is recommended. Once an organism is identified by blood culture or culture of the epiglottis, antibiotics specifically directed to the organism’s susceptibility should be prescribed. The optimal duration of therapy is unknown, although most physicians treat for 7 to 10 days. A longer course may be required in cases of bacteremia.

Evidence to support routine use of glucocorticoids in patients with epiglottitis is insufficient. Retrospective studies do not show an association between glucocorticoid use and reduced length of intensive care unit or hospital stay or duration of intubation. Such use may be beneficial in select patients with severe disease or in patients who are difficult to extubate.

## Prognosis

Epiglottitis generally resolves after a few days of antibiotics, and the patient may be extubated once the inflammation is under control. Most patients recover without residual airway problems if the airway is secured promptly and they are treated with appropriate antibiotics.

## Lessons for the Clinician

- Acute epiglottitis is a medical emergency that warrants immediate intervention to secure the airway.
- A combination of a third-generation cephalosporin and an antistaphylococcal agent against methicillin-resistant *Staphylococcus aureus* is recommended.
- The incidence of *Haemophilus influenzae* type b epiglottitis declined significantly after the introduction of vaccination.

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