CASE OF THE MONTH

Sarah Smitherman MD

Initial Presentation:

A 25 year old previously healthy female presented with a sore throat of three days duration. Associated symptoms included dysphagia and odynophagia but no drooling was reported. Her pain progressed to the point that she refused all solid foods and liquids. She also complained of neck pain and swelling that limited the range of motion in her neck. Fever had ranged from 102-104 degrees F. She had a "raspy" voice but did not complain of respiratory difficulty, stridor or wheezing.

Initial exam revealed an erythematous posterior pharynx and a midline, non-edematous uvula. The epiglottis was not visible due to a significantly swollen and tender posterior third of her tongue. She had bilaterally enlarged cervical lymphadenopathy (> 1cm) of both the anterior and posterior chains. Her neck range of motion was limited by edema and tenderness. Respiratory exam was negative for stridor, wheeze, crackles or rhonchi.

Differential Diagnosis:

Bacterial pharyngitis can be from streptococci, staphylococci, diphtheria, meningococci or gonococci. Viral pharyngitis can be secondary to EBV, CMV, adenovirus, parainfluenza and primary HIV infection. Other causes of severe dysphagia and odynophagia include epiglottitis or abscesses (tonsillar, peritonsillar or retro-pharyngeal). Congenital defects of the thyroglossal duct and pharyngeal cleft cysts must also be considered in a patient with significant neck swelling and tenderness.

Evaluation and Management:

The patient's lab data revealed a WBC of 22,500 with 80.4% granulocytes. Streptococcal pharyngitis was ruled out with a normal rapid strep test and a negative throat culture. Blood cultures were negative. A monospot was performed and was positive.

A CT of the neck was obtained to look for a possible abscess; it demonstrated a 2.1 x 2.7 cm soft tissue enhancement at the base of the tongue. The epiglottis was not clearly distinct from this swelling and there was concern for epiglottitis; the CT also revealed significant airway compression, with the narrowest point measuring only 6 mm. Emergent ENT consultation was requested and a transnasal fiberoptic flexible laryngoscopy was performed at the bedside. Engorgement of the lymphoid tissue at the base of the tongue was encroaching on the epiglottis but the epiglottis itself was not inflamed.

While cultures were pending, the patient received empiric coverage with vancomycin and ceftriaxone and completed three doses of IV dexamethasone due to concerns for airway obstruction; a difficult intubation kit was kept at her bedside. The patient was closely monitored in the Stepdown Unit with the head of her bed elevated and continuous pulse oximetry in place. By the third hospital day, the patient had improved significantly; her lymphadenopathy had decreased, normal range of motion in her neck was restored and she was tolerating an oral diet.

Learning Points:

Epiglottitis in adults is a rare occurrence but should be considered when faced with a clinical presentation such as this; our patient had the typical warning signs of epiglottitis, including odynophagia, dysphagia and change in voice quality. Epiglottitis is a medical emergency because of its rapid onset and potential for compromise (cont)

(continued) of the airway. Lateral neck radiographs are not as reliable for diagnosing epiglottitis in adults as they are in children. The evaluation of possible epiglottitis in adults is best performed by the use of transnasal fiberoptic flexible laryngoscopy, as was done in our case. The agent of epiglottitis was formerly Haemophilus influenza B but, in the post HIB vaccination era, it is primarily caused by Staphylococcus aureus and Streptococcus pyogenes. Treatment of epiglottitis should include broad spectrum antibiotics and corticosteroids; if treated appropriately, most adult patients recover without the need for intubation.

While this patient did not have epiglottitis, she had significant EBV associated lymphadenopathy. She was diagnosed with EBV via a positive Monospot; in the context of mononucleosis-like symptoms, the Monospot sensitivity can reach 85%, with a specificity of 94%. However, 25% of adults may have a negative Monospot during the first week of infection. The Monospot sensitivity is further reduced in children under 12; in this group, only 25-50% of the Monospots are positive. Had the Monospot been negative in our patient, further testing would include screening for either EBV specific antibodies (IgG and IgM) or EBV nuclear antigen proteins.

Airway obstruction in EBV infection is rare (<5%), with young children at higher risk than adolescents or adults. When there is concern for airway compromise, a CT of the head and neck should be performed to look for concomitant bacterial abscess formation. Until imaging is obtained, empiric antibiotics should be initiated but steroids should be withheld until abscess formation has been ruled out; upper airway obstruction is one of the few indications for steroid therapy in EBV management. When indicated, a three day course of dexamethasone or methylprednisolone is recommended; subjective improvement often begins within 24 hours. Some advise caution regarding the use of steroids in patients with EBV infection because of the potential for contributing to subsequent EBV associated malignancies.

Summary:

EBV virus can be a significant cause of head and neck lymphadenopathy which may lead to airway compromise in severe cases. Our patient improved significantly after receiving empiric antibiotics and corticosteroid therapy; the steroid therapy was brief and the antibiotics were discontinued once epiglottitis and abscess were ruled out. The patient was discharged to home on the third hospital day and continued to improve during the course of her outpatient care.

References:

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