Abstract

Objectives: To describe a case report on an unusual presentation of acute angioedema response to Strep tonsillitis.

Study Design: Retrospective case review.

Methods: Retrospective review of the electronic medical record, CT imaging and photo documentation were obtained.

Results: A 10 year-old boy diagnosed with Strep A pharyngitis, developed tender swelling of bilateral parotid and submandibular glands, and diffuse facial soft tissue edema. The diagnosis of angioedema emanating from localized Strep A tonsillitis was proposed. Intravenous steroids and antihistamines were initiated and the angioedema resolved.

Conclusions: In this case, radiographic and physical findings of diffuse edema pointed to the diagnosis of angioedema.

Introduction

Angioedema is a disease process commonly encountered by the otolaryngologist. Our assistance is often requested in the evaluation of the upper airway to determine the need for airway protection when acute noninfectious swelling of the face, lips, tongue, palate, pharynx or larynx occurs. Typically, angioedema occurs as a result of exposure to allergic antigen or ACE inhibitors. Less commonly, although it has been reported in case series it can be a reaction to infectious agent: bacteria or toxin.

Case Report

A10 year-old previously healthy boy with no known medical allergies presented to an urgent care clinic with a two-day history of throat pain, and fever. He was diagnosed with Strep A pharyngitis and given oral amoxicillin, to which he had been exposed in the past without reaction.

One day later, he developed localized tender swelling of bilateral parotid and submandibular glands, and facial soft tissue edema. He then presented to our emergency room with rapidly increasing panfacial, submandibular and parotid swelling. Broad-spectrum antibiotics (Unasyn and Vancomycin) were given. However in the next eight hours his facial swelling continued to worsen, and the patient developed a hot potato voice. Although the salivary glands were all equally and diffusely swollen, they showed no clinical signs of infection; no redness of the skin or oral mucosa or purulence from either Stensen’s or Wharton’s ducts. The laryngeal complex was free of edema and the upper airway remained patent on fiber optic exam. The swelling appeared in a location consistent with angioedema extending from the tonsillar fossae. The possibility of mumps and HIV were entertained but ruled out by serology. Given the history, physical examination, and radiologic findings the diagnosis of angioedema in reaction to strep A infection was proposed.

Intravenous steroids and antihistamines were initiated. The patient’s physical exam showed reduction of swelling 12 hours later, and nearly complete resolution of hot potato voice and facial swelling 72 hours later.

The patient was seen in follow-up in otolaryngology clinic several weeks later after complete resolution of tonsillitis and angioedema. He subsequently underwent elective tonsillectomy and received post op amoxicillin without incident. This supports the conclusion that the previous angioedema incident was in reaction to the infectious agent and not medication related. Informed consent for release of medical information and photographs for academic medical use was obtained from the patient’s parent.

Discussion

Angioedema is a non-pitting edema that affects skin, subcutaneous tissue and mucosal membranes. Commonly affected sites are the head and neck/upper airway and intestinal mucosa. The common result of the various pathways is immunologically induced vasodilation and vascular permeability resulting in extravasation of fluid into the extra vascular space and soft tissue edema. The transudative nature of edema as a result of vasoactive mediators is reflected in CT findings of infiltrative transspatialeedema.

There are two main pathophysiological pathways by which angioedema is believed to propagate: the mast cell degranulation and bradykinin perpetuation pathways. In the mast cell pathway, IgE activates mast cells release of histamine, prostaglandin, and leukotrienes. These mediators increase vascular permeability resulting in cutaneous soft tissue and mucosal edema. Triggers for this pathway are commonly foods (milk, eggs, peanuts, shellfish) and drugs. Angioedema related to acute viral and bacterial infection implicates immune complexes in IgE cross linking and subsequent mast cell degranulation. The bradykinin pathway is associated with both hereditary and Angiotensin converting enzyme (ACE) inhibitor induced angioedema. In ACE inhibitor associated angioedema it is postulated that when ACE, being one of the main enzymes that degrades bradykinin, is inhibited, a build up of active bradykinin results leading to vasodilation, cGMP and nitric oxide release.

Treatment must include thorough airway evaluation and stabilization. The mainstay of treatment includes identifying and removing the offending trigger. In cases where mast cell degranulation is suspected, intravenous corticosteroids and antihistamines should be instituted. Several authors have noted the more rapid recovery from angioedema attacks in pediatric patients than adults. This may be reflective of underlying causes. In children, allergic mast cell mediated pathway is more common, which responds well to corticosteroids and antihistamines, whereas in adults ACE inhibitor mediated bradykinin pathway is more common, and less responsive to corticosteroids and requires return of ACE activity in order to be reversed.

Conclusions

In our patient example, CT findings of diffuse edema rather than suppurative parotitis pointed to the diagnosis of angioedema. After ruling out infectious parotitis and given the lack of an appropriate drug or food allergen culprit, infectious immune complex mediated mast cell degranulation was suspected. Although Penicillins are a common allergen in pediatric patients, the patient had previous and subsequent exposures to penicillin without reaction. Continued antibiotics may have even facilitated recovery by reducing bacteria and toxin. Furthermore, resolution of angioedema occurred despite continued penicillin based antibiotic therapy suggested against this as the trigger. Additional workup with allergy testing may help to confirm our diagnosis. Definitive treatment for this child was achieved by tonsillectomy thereby significantly reducing the chance of Strep A infection and subsequent immune complex formation triggering mast cell related angioedema.

References


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