Prevalence of Periodic Fever, Apthous Stomatitis, Pharyngitis and Cervical Adenitis Syndrome (PFAPA): Eastern Wisconsin

Mona Ranade[1], Dennis J. Baumgardner[1,2,3], Indrani Banerjee[2], Meghana Doniparthi[4]

PFAPA Syndrome

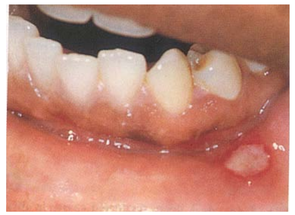
- Periodic Fever
- Apthous Stomatitis
- Pharyngitis
- Cervical Adenitis

Illustrative Case

Case Description

- 12 y/o male with multiple handicaps (probably incidental/immunocompetent)
- Began having episodes of apthous stomatitis with fever and pharyngitis &/or cervical adenopathy & non-specific symptoms
- Every 3 to 5 weeks
- At first were diagnosed as apthous stomatitis PLUS pharyngitis or otitis media or sinusitis. Antibiotics given.
- After about 6 bouts, Dx: PFAPA Syndrome
- Rx: oral cimetidine, po Lysine, Vits B/C
- No SDS/sodium lauryl sulfate in toothpaste
- So far: 34 total bouts, but symptoms less

Aphthous Stomatitis (AS)



- AKA – Canker Sores
- Etiology: Unknown, alterations in local cell mediated immunity may be important
- Diagnosis: Painful oral shallow round/oval ulcers with grayish base; vestibular/buccal mucosa, tongue, soft palate, fauces, floor of mouth
- AS is recurrent approx 50% of time
- Recurrent apthous stomatitis (RAS) is the most common cause of mouth ulcers in North America
- Mostly children & young adults
- Predisposing factors for RAS: stress, hormones, infection, food hypersensitivity, immune irregularity (HIV, etc.), familial. SDS in toothpaste may increase severity(?)
- RAS can be due to: vitamin & mineral deficiencies, esp B, Fe, folic acid, Zn; sprue, gluten sensitivity; inflamm bowel disease; methotrexate; neutropenia

PFAPA Syndrome

Etiology: Unknown
Epidemiology: Onset 2 – 5 y/o, Male>Female
No ethnic/racial predilection
Fever: 38.9 – 41.1 deg C X 3-4 days
Repeats q 26 – 30 days (on average)
Apthous Ulcers
Pharyngitis & Adenopathy
Sometimes other non-specific symptoms

- Diagnosis: Clinical. Three or more episodes; otherwise normal before onset & in between
- Treatment: Try single dose of prednisone (2mg/kg) at onset of fever. May give 4 day taper if recurrence of sxs that episode.
- Cimetidine may possibly work to prevent recurrences in some children
- The disorder may be more heterogeneous than previously recognized (eg. older aged children, range of symptoms)
- Thought to be rare, the true prevalence rate is unknown

Hypotheses

- PFAPA Syndrome is more common in a community population than reported
- The signs/symptoms are not recognized as a single syndrome
- Felt to be apthae PLUS pharyngitis, etc.
- Unnecessary antibiotic prescriptions result

Objective

Determine the prevalence, clinical features and diagnosis of PFAPA in a community population

Methods

- Retrospective electronic chart review of children with surnames A-J seen in a large eastern Wisconsin integrated health system, 2002-2007, with diagnosis of stomatitis (ICD-9 codes 528.0, 528.2)
- AS syndromes were enumerated and demographic, clinical and treatment histories were compared utilizing chi-squared test for categorical variables and Mann-Whitney test for non-normal continuous variables

Results

- Review of 949 records revealed 353 cases of apparent AS
- 47 (13%) were documented to be recurrent (at least two episodes in one year), significantly lower than 50% as reported in current literature
- There was a trend toward recurrent cases (RAS) being more commonly male (60% [versus 40% non-recurrent], p=0.08)
- Otherwise RAS cases did not differ from non-recurrent cases by age, race or specialty of diagnosing clinician; or presence of fever, cervical adenopathy or pharyngitis at the time of visit.
- Only 2 cases (0.6%) were PFAPA
- A 16 y/o female with typical symptoms for at least 2 years, sometimes also headache/nausea
- A 7 y/o male with onset age 5 – Probable Case Had at least 4 episodes: once 4/4 symptoms, once 3 of 4; concurrent otitis media diagnoses
- Neither case had been diagnosed with PFAPA

Other observations

- Antibiotics were prescribed in 4% of all AS cases without other diagnosis.
- Anecdotally, primary care clinicians exhibited significant confusion in their documentation of mouth sores: AS vs. Herpes vs. Herpangina.
  – e.g., wrong terms or two different terms used to describe the lesions in the same chart note.

Conclusions

- PFAPA Syndrome indeed appears to be rare in a community population.
- The recurrent nature of AS appears to be poorly documented, but does not contribute significantly to inappropriate antibiotic prescriptions.

Affiliations

1. University of Wisconsin School of Medicine and Public Health
2. Center for Urban Population Health
3. Aurora UW Medical Group
4. Arrowhead High School, Hartland, WI

Center for Urban Population Health
Working together to improve the health of communities

University of Wisconsin School of Medicine and Public Health | UWMilwaukee | Aurora Health Care