A Case of Behcet's in a 5-year-old Girl

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CHIEF COMPLAINT

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- A 5-year-old girl complains about painful genital ulcer for the last 2 days
- Pain: Described as "fire and needles" to the vulva. Mother initially thought that the patient had a scratch
- Significant history: intermittent painful ulcerations in the mouth for the last two years causing similar pain
- Most recent mouth ulcer occurred 2 weeks prior to presentation
- Pertinent negatives: denies vaginal discharge or itching; no history of sexual abuse, no fever or chills

PATIENT HISTORY

- Treated for Hand, Foot and Mouth Disease in the past without resolution
- Previously given a diagnosis of "vesicular stomatitis vs. aphthous ulcer"
- Gastroenterology Consult:
  - Ulcer biopsy was inconclusive
  - Enzymatic tests showed possible Sucrase-Isomaltase deficiency
  - Started on enzyme replacement therapy
- Family history:
  - No history of autoimmune diseases
  - No known hx of Behcet's disease
  - All of her siblings are healthy.

PHYSICAL EXAMINATION

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VITAL SIGNS: Stable, afebrile

HEENT: Mucous membranes clear without ulceration at this time

SKIN: Her skin turgor revealed normal hydration with no rashes, lesions or discolorations to the trunk or extremities

LYMPHATICS: No lymphadenopathy of the neck, axilla or groin

GENITALIA: 3 mm circular erythematous lesion over the left inner labia, beefy red with well-demarcated borders; urethra was normal, without erythema

No masses or tenderness, no discharge and no evidence of necrosis

No physical evidence to raise suspicion of sexual abuse

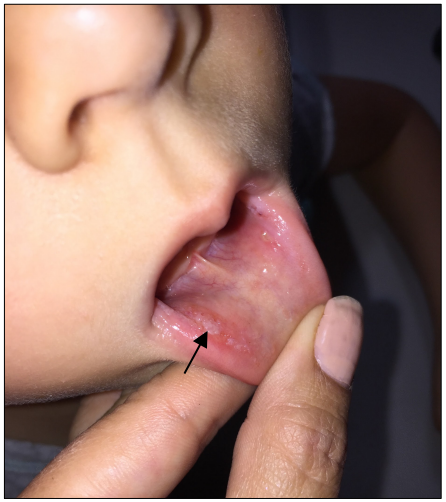

Figure 1: Ulceration of the oral mucosa present two weeks prior to the onset of the vulvar ulcer

DIFFERENTIAL DIAGNOSIS

A. Behcet's disease

B. Vulvovaginitis

C. Pyoderma gangrenosum

D. Herpes Simplex Virus -1

Commentary:
The positive HLA-B51 testing with history of oral and genital ulcerations led to the diagnosis of Behcet's disease. The lack of vaginal itching or discharge ruled out vulvovaginitis. There was no evidence of necrosis.

Outcome:
Decubitus cream was applied to the affected region, however, she complained of a burning sensation with application that was intolerable. Instead, 1% hydrochlorothiazide cream was given with a 5 day course of prednisone. The ulceration had resolved by follow up one week later.

DISCUSSION

- The etiology of this disease is unknown but is thought to be autoimmune.
- It is diagnosed based on a triad of oral ulceration, genital ulceration and ocular involvement.
- It has been associated with HLA-B51
- It can spread to every organ system, most commonly the GI tract but can also progress to neurological manifestations.
- Early recognition and suppression of inflammation will help prevent further organ system involvement and better overall prognosis.[1]
- This case illustrates the difficulty of diagnosis and progression to other regions without proper treatment.
- BD is most common in young adults, but children can also be affected by this disease.
- BD should be a differential diagnosis in any case of oral or genital ulceration.[2]

REFERENCES

1. Warren S, Alden K, and Lomax N. Behçet's disease – part of the differential diagnosis of ulcerative lesions. *Int J STD AIDS.* 2014;25:1044-1046.
2. Ozuguz P, Kacar S, Manav V, et al. Genital Ulcerative Pyoderma Gangrenosum in Behçet's Disease: A Case Report and Review of the Literature. *Ind J Derm.* 2015;60:105.

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ABSTRACT

A 5-year-old Caucasian female presented to a pediatric clinic suffering from an ulceration to her vulva for 3 days. Her mother noticed she began crying with urination, the patient described the pain as "fire and needles" in the vaginal area. The patient's mother examined her and noted a small lesion her vulva. The site progressively increased in size and the pain worsened.

The patient previously suffered from painful ulcerations to her mouth intermittently. She was initially treated for Hand, Foot and Mouth disease. She was seen by a gastroenterologist who diagnosed her with a sucrase-isomaltase deficiency and was started on an enzyme replacement medication. Genetic studies were performed and a positive human leukocyte antigen-B51 gene (HLA-B51) was identified.

Although vulvovaginitis, pyoderma gangrenosum and Herpes Simplex Virus-1 were considered in the differential, the positive HLA-B51 testing with history of oral and genital ulcerations led to the diagnosis of Behcet's disease (BD). She was started on a five day course of prednisone. On follow up one week later, the ulcer had resolved and a referral to rheumatology was provided.

The etiology of BD is unknown, it is a multisystemic disease characterized by recurrent oral and genital ulcerations with ocular disease. It is often considered an immune-mediated small vessel vasculitis. It can cause arthritis and vascular disease and can involve gastrointestinal, musculoskeletal, respiratory and central nervous system manifestations. BD has highest prevalence in adults ages 20-40 of Middle Eastern and Asian descent.[2]

The characteristic ulcer is small and round with well-demarcated borders, an erythematous halo and a yellow-gray base. These lesions can easily be confused with aphthous stomatitis, especially in the pediatric group which is less commonly affected by this condition.[2]

Unfortunately, there is no cure for BD. The aim of treatment is symptom relief and prevention of further disease progression. It is imperative that BD is recognized early to prevent further organ system involvement.[1] This case illustrates how easily these ulcerations can be misdiagnosed. BD should be considered in any case of oral or genital ulceration.

REFERENCES

1. Warren S, Alden K, and Lomax N. Behçet's disease – part of the differential diagnosis of ulcerative lesions. *Int J STD AIDS*. 2014;25:1044-1046.
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