Bowel-associated dermatosis-arthritis syndrome (BADAS) in a pediatric patient

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INTRODUCTION

Bowel-associated dermatosis-arthritis syndrome (BADAS) is a recurrent and episodic neutrophilic dermatosis characterized by erythematous macules, which quickly evolve into papules and vesiculopustules on the trunk and upper extremities.[1,2] Originally only associated with bariatric surgery,[3] BADAS is now reported secondary to gastrointestinal disease.[4] Although typically seen in adults, we report a case of a 4-year-old patient with numerous scattered hemorrhagic vesicles and flu-like symptoms whose biopsy specimen revealed a neutrophilic infiltrate consistent with BADAS. To our knowledge, this is the youngest reported case of BADAS in English-language literature.

CASE REPORT

A 4-year-old African-American girl presented to the emergency department after a several-day history of cough, congestion, intermittent fever, severe intermittent abdominal pain, and bloody stools. She had a medical history significant for sickle cell disease, ulcerative colitis, and autoimmune hepatitis being treated with hydroxyurea, mesalamine, and azathioprine. On examination, she was noted to have clusters of 2- to 7-mm hemorrhagic vesicles on her palms, knees, and soles and a tender, swollen right knee (Figs 1 to 3). Her laboratory workup revealed an elevated C-reactive protein (31.1 mg/dL) and white blood cell count ($25.5 \times 10^3/\mu L$), anemia (hemoglobin 6.6 g/dL and hematocrit 18.7%), and negative bacterial and viral cultures of vesicle fluid. She was treated with a blood transfusion and empiric antibiotics were started.

Her mother reported she had visited her pediatrician and the emergency department for a similar eruption diagnosed as Coxsackie virus and varicella on 2 separate occasions. Four months prior, she was admitted for a clinically identical episode that was attributed to a sickle cell crisis versus a flare of ulcerative colitis. At that time, dermatology was consulted and a biopsy specimen of a pustule on the patient's left arm revealed a neutrophilic panniculitis (Fig 4).

A repeated biopsy specimen obtained during her most recent hospital admission showed subcorneal and subepidermal edema and a prominent acute inflammatory cell infiltrate composed entirely of neutrophils. The inflammatory cell infiltrate extended into the adipose tissue and into the septa. A direct immunofluorescence study was also performed, which was negative.

Because of recurrent episodes of fever (maximum of 40.2°C), malaise, abdominal pain, and arthritis with the characteristic cutaneous lesions and histologic findings in the setting of an ulcerative colitis flare, the diagnosis of BADAS was made. The patient's acute flare was treated with an 11-day course of metronidazole and prednisone. Infliximab infusions were then started every 8 weeks in addition to her azathioprine and mesalamine in an attempt to control her colitis. Her skin lesions have not recurred since her last admission. To our knowledge, this is the youngest reported case of BADAS in English-language literature.

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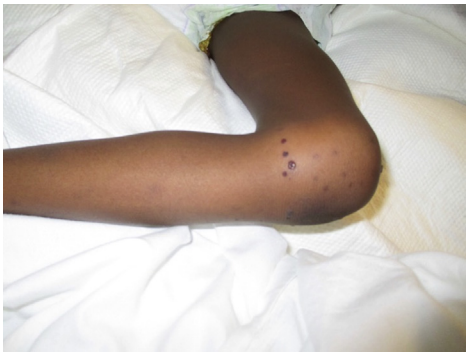

Fig 1. Bowel-associated dermatosis-arthritis syndrome. Scattered hemorrhagic vesicles overlying a joint effusion.

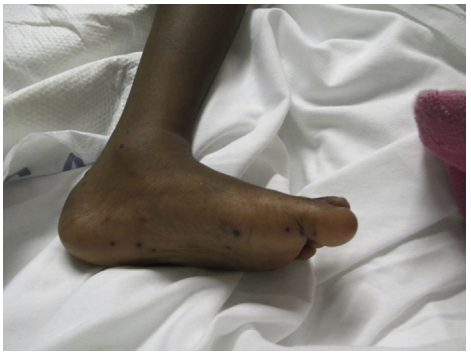

Fig 2. Bowel-associated dermatosis-arthritis syndrome. Scattered hemorrhagic vesicles.

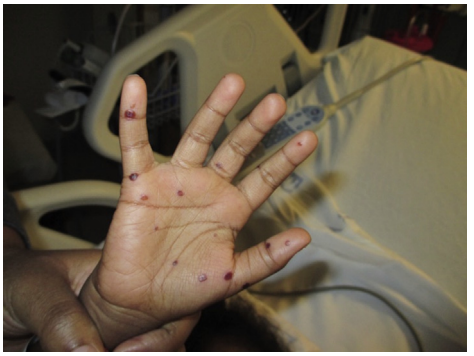

Fig 3. Bowel-associated dermatosis-arthritis syndrome. Scattered hemorrhagic vesicles.

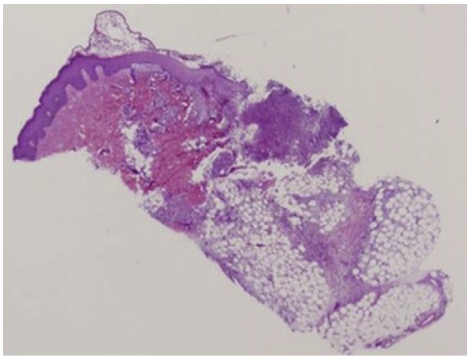

Fig 4. Bowel-associated dermatosis-arthritis syndrome. Neutrophilic panniculitis. (Hematoxylin-eosin stain; low-power magnification.)

DISCUSSION

BADAS is a recurrent and episodic neutrophilic dermatosis characterized by erythematous macules, which quickly evolve into papules and vesiculopustules on the trunk and upper extremities. In most cases after persisting for approximately 1 week, each flare spontaneously resolves without scarring and can be associated with relapses every 4 to 6 weeks.[1,2] Eruptions of BADAS can be accompanied by nonerosive tenosynovitis, polyarthralgias, diarrhea, and flu-like symptoms such as myalgias and fever.[2]

Originally, the condition now known as BADAS was referred to as bowel bypass syndrome because of its association with bariatric surgery, in particular, jejunoileal bypass.[3] BADAS has been noted to occur anywhere from a few postoperative days to up to 18 years postbariatric surgery. Although the incidence of postoperative BADAS previously has approached 20%, newer surgical techniques have led to a decline in these numbers.[4,5] It was not until 1983 that 4 cases were published in which patients presented with BADAS without a history of bariatric surgery. These cases were instead associated with gastrointestinal diseases within the adult population, expanding the scope of the syndrome and giving BADAS its current moniker.[4] Since then BADAS has been reported in cases of inflammatory bowel disease, diverticulitis, gastric phytobezoar, and even a case of acute appendicitis.[2] BADAS has also been reported as the presenting prodrome of Crohn's disease in an adolescent, however to our knowledge, this case illustrates one of the youngest pediatric patients with this condition in the English-language literature.[6,7]

The origin of BADAS is postulated to be the result of bacterial overgrowth from either surgery, with the creation of a blind loop in bariatric procedures, or inflammation, as seen in intestinal diseases.[2] Bacterial peptidoglycans accumulate during overgrowth resulting in an immune response and antigen-complex deposition that is thought to elicit the cutaneous findings in BADAS. This theory has been tested in animal models using bacterial peptidoglycans of group A streptococci, which share similar antigenicity and structure with human bacterial intestinal flora, producing similar arthritis and cutaneous findings to BADAS.[8]

Histologically BADAS is characterized by a marked neutrophilic infiltrate and a lack of fibrinoid necrosis.[2] This appearance, however, is not pathognomonic of BADAS. Histologically, Sweet

syndrome is identical to BADAS and thus would also be in the differential diagnosis, but clinical history can often help distinguish the 2 syndromes. Other diagnoses to consider could include pyoderma gangrenosum, leukocytoclastic vasculitis, rheumatoid neutrophilic dermatosis, dermatitis herpetiformis, and bacterial and viral infections such as subacute endocarditis and gonococcal sepsis.[1,3]

In cases where BADAS does not remit spontaneously, treatment is directed toward the underlying inflammatory disorder and bacterial overgrowth. As such, steroids and antibiotics have been the mainstay treatment for BADAS; antibiotic choices can include tetracycline, erythromycin, or metronidazole. In addition, improved management and control of any underlying intestinal disease can offer improvement in BADAS.[1-3] For BADAS associated with bariatric procedures, surgical intervention may be warranted to reduce the blind loops of bowel that may lead to the bacterial overgrowth.[2]

In this case, the patient's ulcerative colitis was treated with infliximab infusions without recurrence of BADAS. Given the sparse reports of BADAS in the pediatric patient population it is not surprising that the patient was initially given the diagnosis of a more common entity such as varicella or Coxsackie virus. However, this case illustrates that BADAS should be added to the differential diagnosis of vesicles even in very young patients with bowel disease.

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