Adult Presents with Evolving Rash

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Case Introduction
A 30-year-old adult from the US Pacific Northwest presents to his primary care office two weeks after an acute onset of rash. Initially noted on his ankles, the rash subsequently spread to the upper thighs and torso. The rash was noted after travel to Colorado. He shares that while he enjoyed hiking on vacation, outings were mostly on paved paths. Though he denied seeing any ticks, he initially thought that the “dots” on his legs were bug bites. Additionally, while in Colorado, he noted that he felt sort of “off,” with muscle soreness and a mild fever. He associated these symptoms with altitude.

Last week he was seen at an urgent care for the rash and was treated with fluocinonide cream. He presents to his primary care clinic as over the past week the rash has changed from itchy to sharply painful and from small 2-4mm “dots” to larger and darker nodules (Image 1). He is started on a course of oral steroids as laboratory evaluation is conducted. Complete metabolic panel reveals no abnormal values, A complete blood count reveals a mild elevation in white blood cells, values otherwise are in normal range. Blood cultures and lesion cultures are collected. Two days later, he continues to have muscle ache and nausea and is seen in the Emergency Department. Cultures show no growth to date. He is started on a course of doxycycline for presumed underlying etiology of symptoms. The next day he is admitted to the hospital as neither rash nor symptoms have improved, and he develops acute abdominal pain and vomiting. A lesion biopsy is performed with dermatology consultation.

Discussion
Biopsy Provides the Diagnosis
Differential diagnosis providing a scaffold for further evaluation and inpatient treatment includes IgA Vasculitis, MPOX, Pityriasis rosea, Rocky Mountain Spotted Fever, and Stevens Johnson Syndrome.

Diagnosis: IgA Vasculitis
Biopsy determines diagnosis of IgA Vasculitis, formerly known as Henoch-Schönlein purpura, a leukocytoclastic, immune complex-mediated, small vessel vasculitis. Though the specific etiology of the disease is unknown, predominant IgA deposits associated with this vasculitis are thought to be related to specific antigen exposure in those with genetic predisposition. IgA vasculitis can be associated with infections, malignancies, and autoimmune disease. Adults’ associations with infection are lower than in children but association with malignancies, specifically solid tumors, is higher.

While IgA Vasculitis is the most common vasculitis in childhood with mean age of 6 and incidence of 3-26/100,000 annually, adults may also have this disease though at lower incidence of 0.1-0.8/100,000 annually and at median age of 50. The disease is more common in those assigned male at birth [1].

A Classic Tetrad
Clinical findings of IgA Vasculitis include a classic tetrad: clustered and palpable purpura or petechial lesions of the lower limb (Image 1), abdominal pain, arthralgia, and renal involvement. Notably, thrombocytopения и coagulopathy are absent. Adults and children present similarly; however adults are at greater risk of significant renal damage including end-stage kidney disease and have less incidence of intussusception. Symptoms may develop over days to weeks. Diagnosis is based on clinical presentation and histopathological findings. Biopsy confirmation of diagnosis is more important in adults due to lower incidence rates.

Differential Diagnosis Evaluation
Ultimately biopsy provided the diagnosis of IgA Vasculitis in our patient. However, the distinctive characteristics of each considered etiology, MPOX, Pityriasis rosea, Rocky Mountain Spotted Fever,
and Stevens Johnson Syndrome, assisted in determining the diagnosis of IGA Vasculitis (Table 1). MPOX, a zoonotic infection with unknown reservoir, is an orthopoxvirus known for a 2022 global epidemic. The recent epidemic variant is associated with one or multiple lesions, which may be at different stages of evolution, typically clustered around the site of inoculation. Previous variant rashes evolved through a consistent stage of macule to papule to vesicle to pustule to scab and finally desquamation. The current epidemic variant may not appear in each distinct stage. It may or may not be associated with a prodrome of fever, fatigue, headache, and painful lymphadenopathy. Transmission is mainly skin to skin during sexual contact [2].

Table 1: Comparison of Distinct Characteristics of the Differential Diagnosis.

<table>
<thead>
<tr>
<th>Condition</th>
<th>Characteristics</th>
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<tbody>
<tr>
<td>IgA Vasculitis</td>
<td><strong>Classic tetrad:</strong> clustered, palpable purpura or petechial lesions of the lower limb, abdominal pain, arthralgia, and renal involvement</td>
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<tr>
<td>MPOX</td>
<td>Up to ten lesions present at different stages of evolution, clustered around inoculation site</td>
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<td>Pityriasis rosea</td>
<td>Herald patch usually on the trunk; generalized rash of scaly papules and plaques along Langer lines</td>
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<tr>
<td>Rocky Mountain Spotted Fever</td>
<td>Maculopapular or petechial lesions; may be present on palms and soles</td>
</tr>
<tr>
<td>Stevens Johnson Syndrome</td>
<td>Erythematous or violaceous patches, atypical targetoid lesions, bullae, erosions, and ulcers</td>
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Pityriasis rosea is associated with a herald patch usually erythematous and greater than 3cm in diameter that appears on the trunk. This primary eruption is followed by a generalized rash of scaly papules and plaques, found commonly along the “Langer” lines. Fatigue, nausea, headaches, joint pain, lymphadenopathy, fever, and sore throat may present prior to the herald patch. An infectious etiology, particularly human herpes viruses 6 and 7 (HHV-6 and HHV-7) is suspected for this self-limiting condition [3].

Rocky Mountain Spotted Fever is a tickborne illness caused by *Rickettsia rickettsia*. More often the Eastern United States dog tick is the vector of this disease, however the wood tick present in the Rocky Mountain states may also transmit this bacterium. This potentially lethal disease progresses rapidly with common symptoms of fever, headache, myalgia, nausea, and rash of either maculopapular or petechial characteristics that may be present on palms and soles [4].

Stevens-Johnson Syndrome (SJS) presents as erythematous or violaceous patches, atypical targetoid lesions, bullae, erosions and ulcers and prodromal skin or eye pain, headaches, cough, sore throat and myalgias. The pathogenesis of the SJS is a complex immunological process most likened to a delayed type hypersensitivity reaction, often from a medication [5].

**Management**

**Symptoms Guide Treatment**

Whereas IgA Vasculitis in children is typically self-limiting, management in adults may be warranted based on symptom severity and risk of renal involvement. Glucocorticoids, Dapsone, and immunosuppressive agents such as Cyclophosphamide or Rituximab, may be indicated treatments for severe IgA Vasculitis disease. Clinical outcomes in adults include remission rates as low as 20% in a 15-year follow-up with the remainder going on to have chronic disease of varying degree, including a 20% rate of relapse [1].

Our patient was treated with a 3-month course of daily Dapsone 100mg and an extended glucocorticoid taper. Dermatological symptoms resolved during the treatment and renal function as well as blood counts and coagulations studies have remained without compromise.

**References**