A bullous enigma in the form of Henoch-Schönlein purpura with diagnostic importance of direct immunofluorescence

Tejas Vishwanath[a], Vishalakshi Viswanath[a], Pradnya Joshi[a] (Poster ID- 8)

[a]- Department of Dermatology, Rajiv Gandhi Medical College and Chhatrapati Shivaji Maharaj Hospital, Kalwa, Thane-400605, India.

Background:

- Henoch-Schönlein purpura (HSP) is the commonest systemic vasculitis of childhood[1].

- Cutaneous involvement is in the form of palpable purpura on the lower limbs and buttocks.

- Rare cutaneous manifestations are hemorrhagic bullae and ulcers[2].

- Internal organs commonly involved include the kidneys, the gastrointestinal tract and the joints.

-Diagnosis is confirmed on histopathology and direct immunofluorescence showing perivascular IgA deposits.

Case-

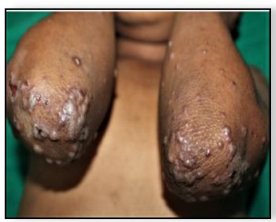
A five year old boy presented with

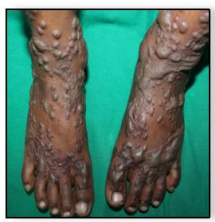
- Purpuric lesions on elbows, legs, genitals and tender erythematous-purpuric papules on buttocks evolving to form bullae since 4-5 days.

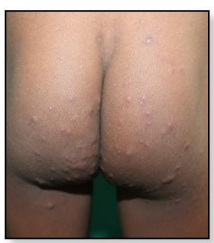
- Associated abdominal pain, black tarry stools since 4-5 days.

- Coryza 3 weeks back.

- No fever/ joint pain /urinary complaints/ recent immunization.







Differential diagnoses:

1.Small vessel vasculitis with internal organ involvement (Henoch-Schönlein purpura)

2. Atypical hand foot mouth disease

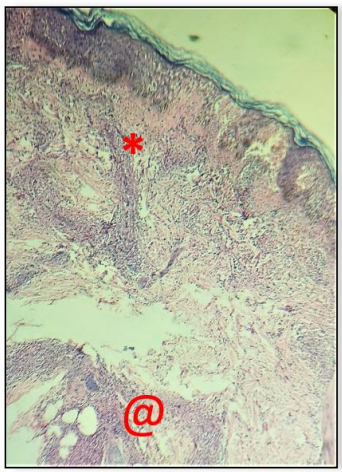
3. Erythema multiforme minor

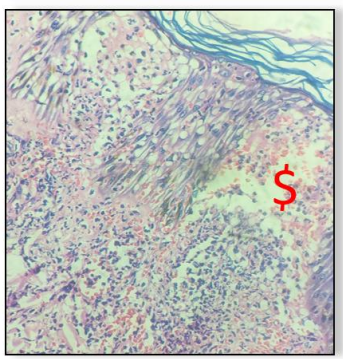
4. Childhood bullous pemphigoid

Investigations:

Hemoglobin	**9.9 g/dl**
Leukocyte count	18000/microlitre
Stool (Guiac test)	Occult blood present
Anto streptolysin O	300 (raised)
USG abdomen	Inflammed bowel loops

Histopathology (H and E)





Findings on histopathology-

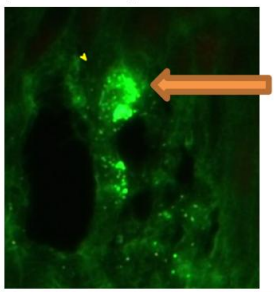
1.**Neutrophilic vasculitis (*)**

2.**Epidermal vacuolization($) with RBCs, polymorph**

3. **Neutrophilic lobular panniculitis(@)-rare feature**

Direct immunofluorescence (DIF)

(confirmatory of HSP in this case)



Perivascular **IgA, fibrin** deposits-clinched the diagnosis as Henoch-Schönlein purpura

Final diagnosis- **Henoch-Schönlein purpura (bullous variant)**

Highlights:

- Rare **bullous variant** of HSP

- **Lobular panniculitis** (rare feature)

- **Confirmatory role of direct immunofluorescenc**

References:

1. Nothhaft M, Klepper J, Kneitz H, Meyer T, Hamm H, Morbach H. Hemorrhagic Bullous Henoch-Schönlein Purpura: Case Report and Review of the Literature. Frontiers in Pediatrics. 2019;6.

2. Mukherjee D, Majumdar I, Pal P, Dhar S, Kundu R. Bullous Henoch–Schonlein purpura with involvement of face. Indian J Paediatr Dermatol 2017;18:338-40.