
بسم الله الرحمن الرحیم

مناجات امام علی (ع)

خدایا این عزت مرا بس که من بنده تو باشم

واین افتخار مرا بس که

توپروردگارمن باشی

،تو آن چنانی که من دوست دارم ،

پس مرا آن چنان کن که تو دوست داری

Vasculitis

Dr. Mohsen Jari

Pediatric Rheumatologist

Isfahan University of Medical Sciences

Vasculitis

- **Vasculitis: Inflammation in a blood vessel wall.**
- **Perivasculitis : Inflammation around a blood vessel wall.**
- **Vasculopathy : An abnormality of blood vessel**

Classification

- ١- Large -Vessel Vasculitis(Giant Cell Arteritis, Takayasu A)
- ٢- Medium- Vessel Vasculitis
- (Polyarteritis Nodosa, Kawasaki disease)
- ٣- Small- Vessel Vasculitis (SVV):

A- ANCA- associated V (Granulomatosis with Poly Angeitis [GPA=Wegener,s], Eosinophilic GPA[EGPA= Churg-Strauss] ,MPA)

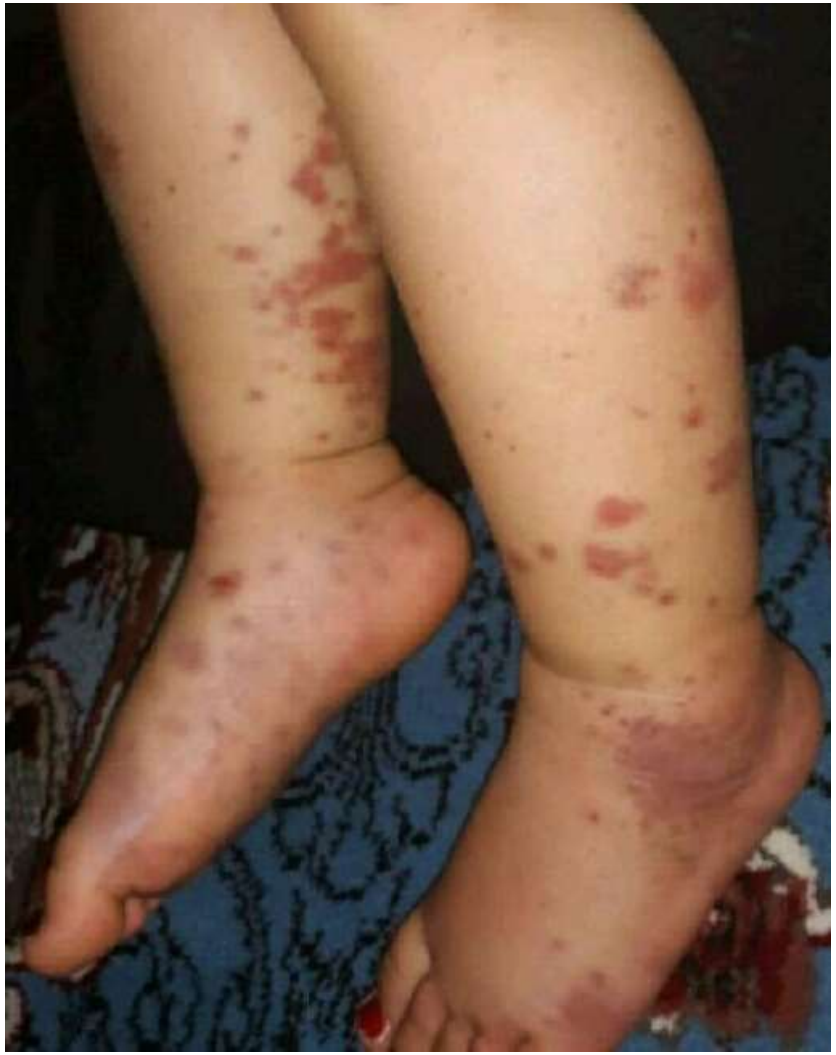
B- Immune complex SVV (HSP,Anti GBM)

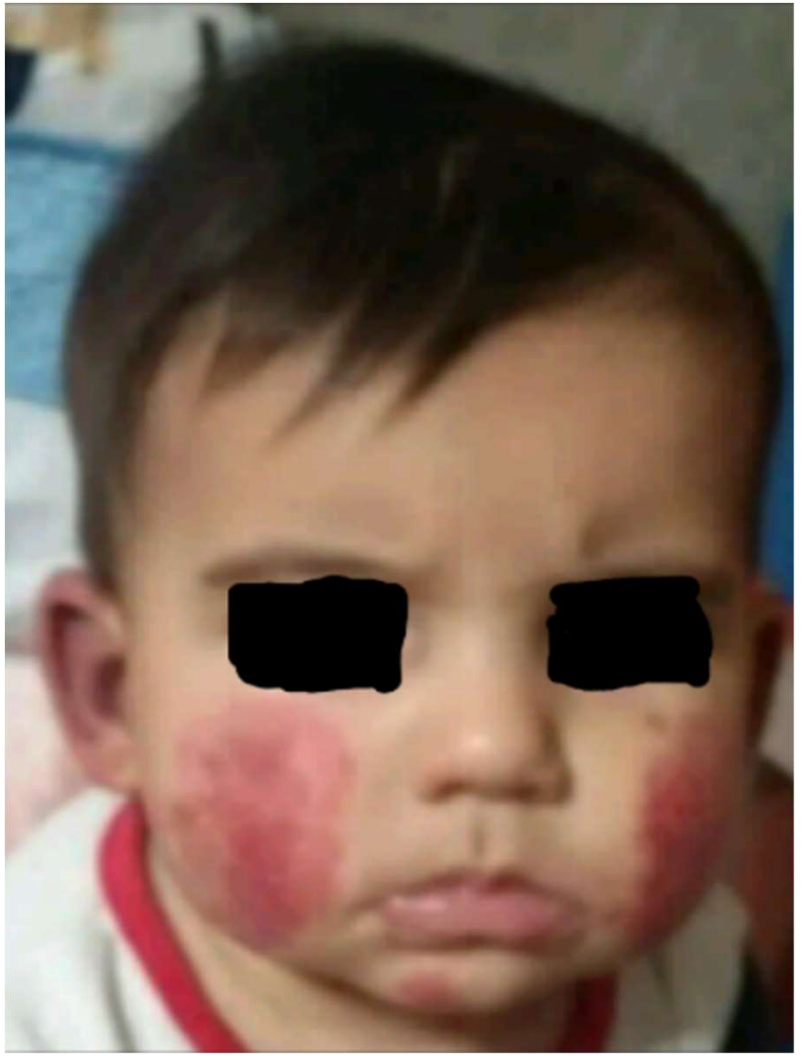
Classification(continue)

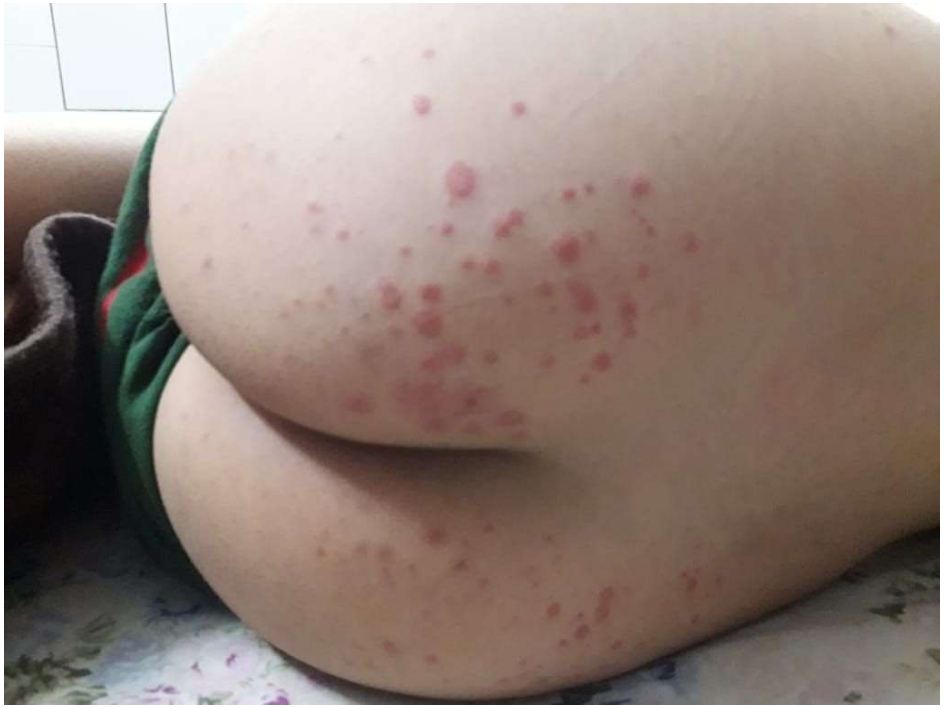
- ۴-Variable Vessel Vasculitis(Behcet disease, Cogan Syndrome)
- ۵- Vasculitis associated with systemic disease

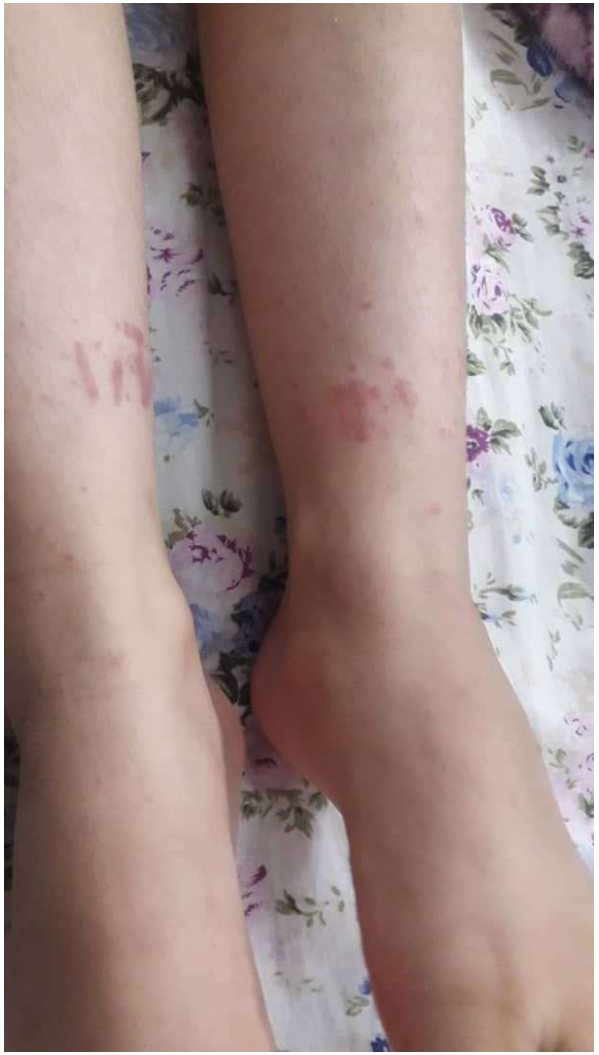
(Lupus V. Sarcoid V)

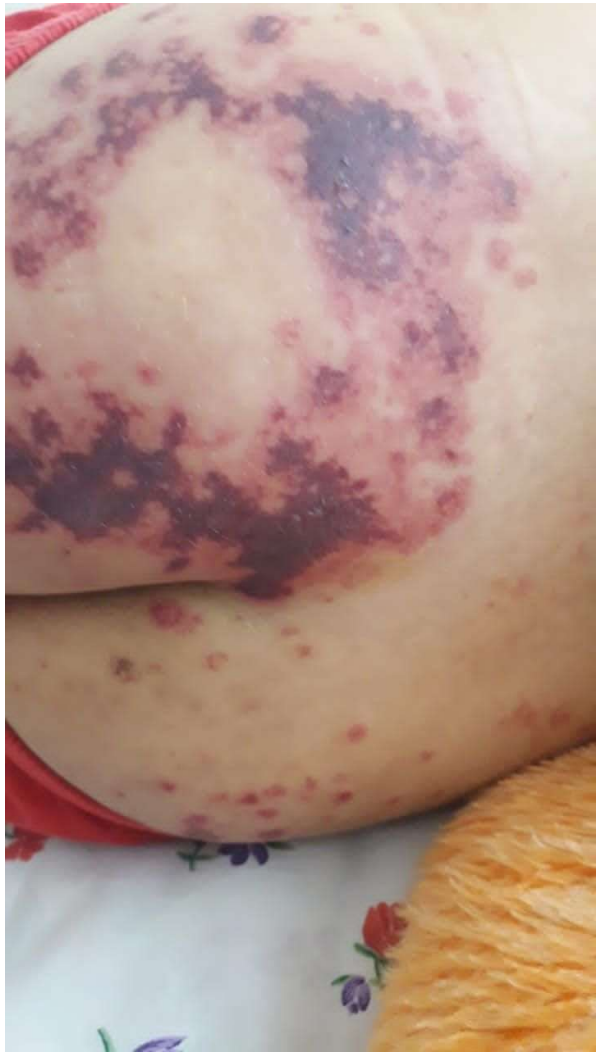
- ۶- Vasculitis associated with probable etiology (HCV,HBV, Syphilis , Drugs , Cancer)

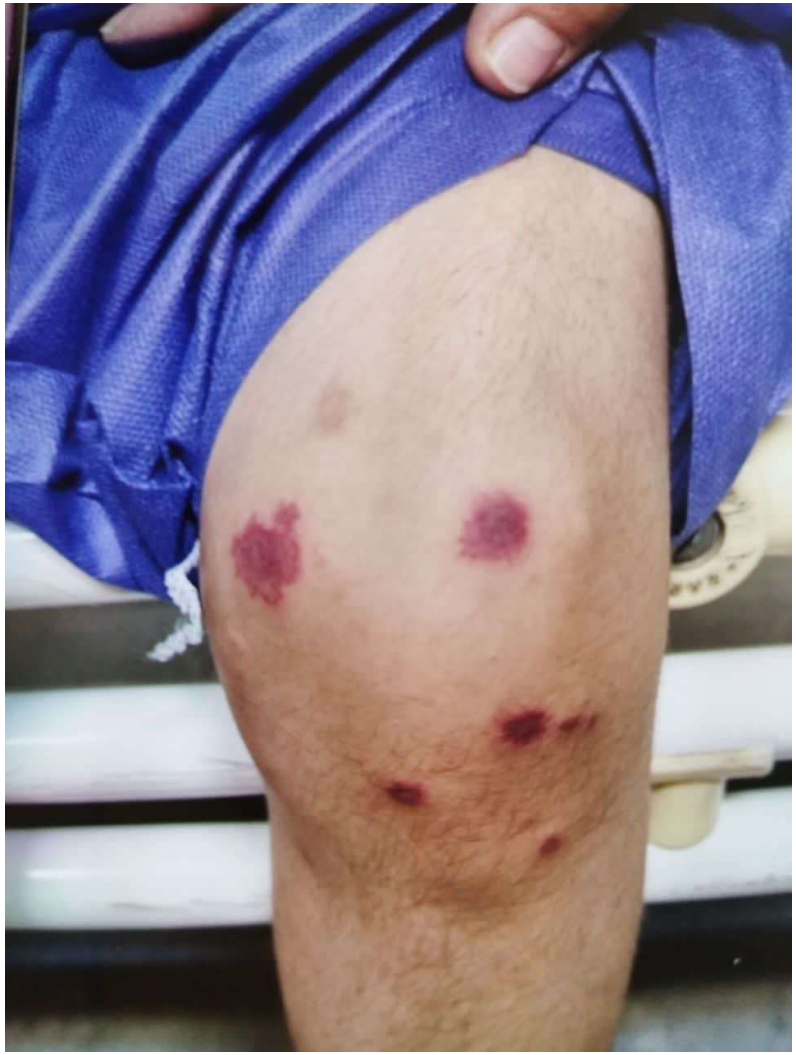


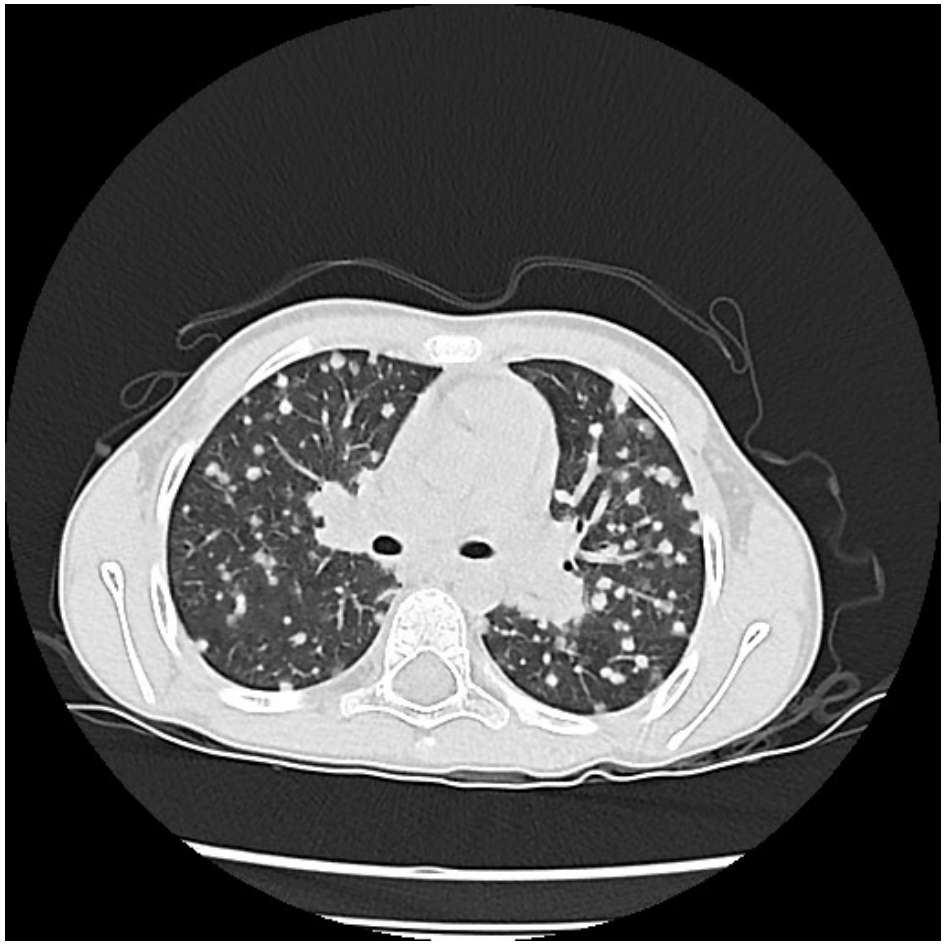


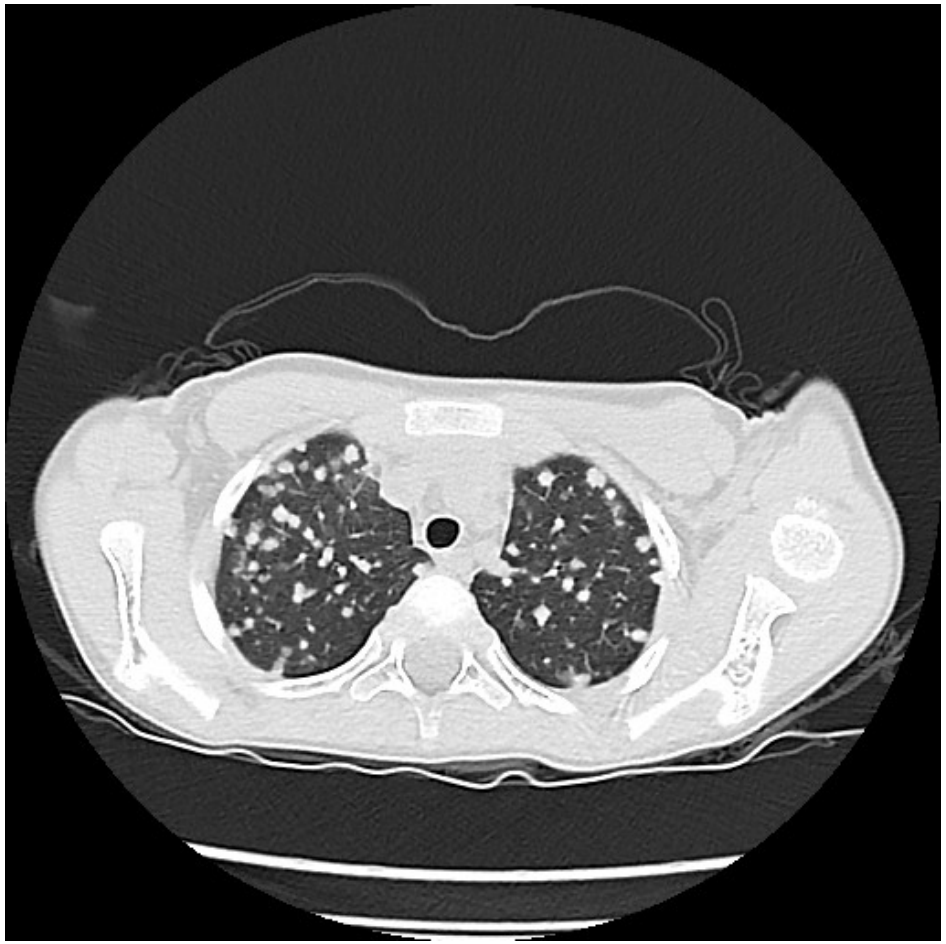


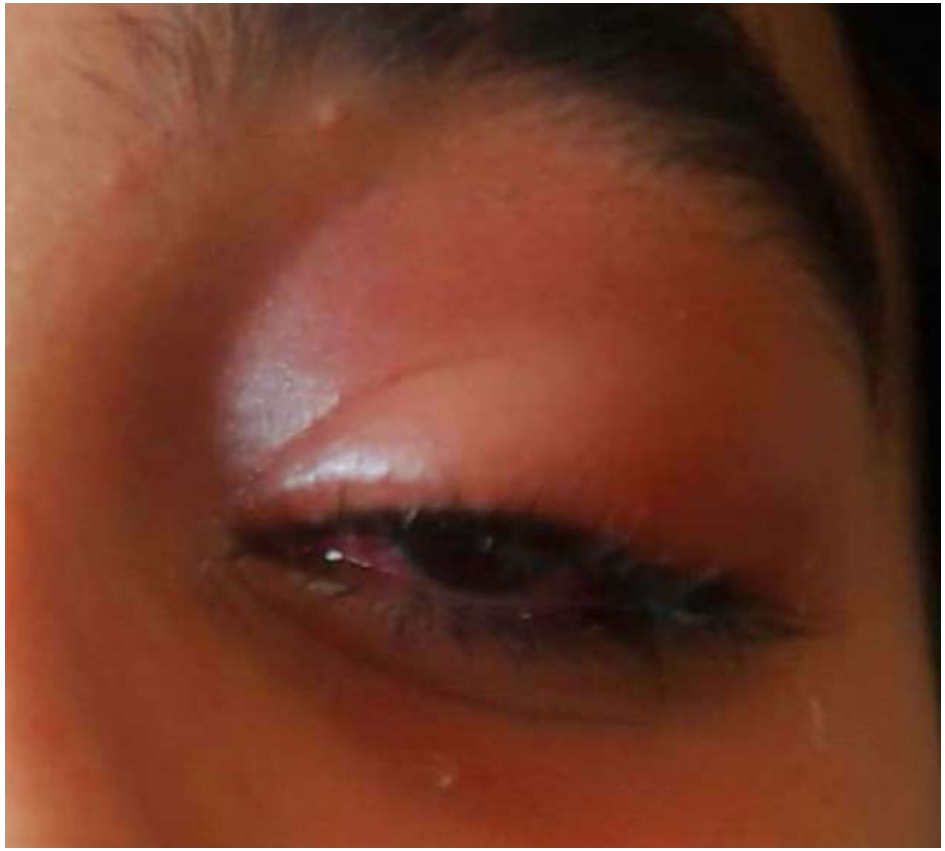


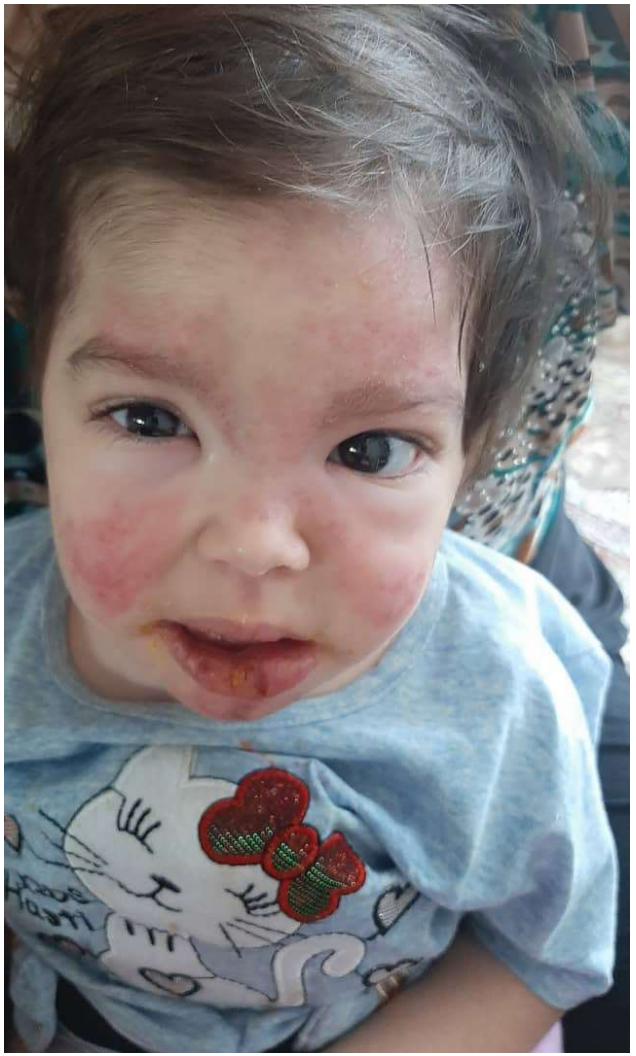


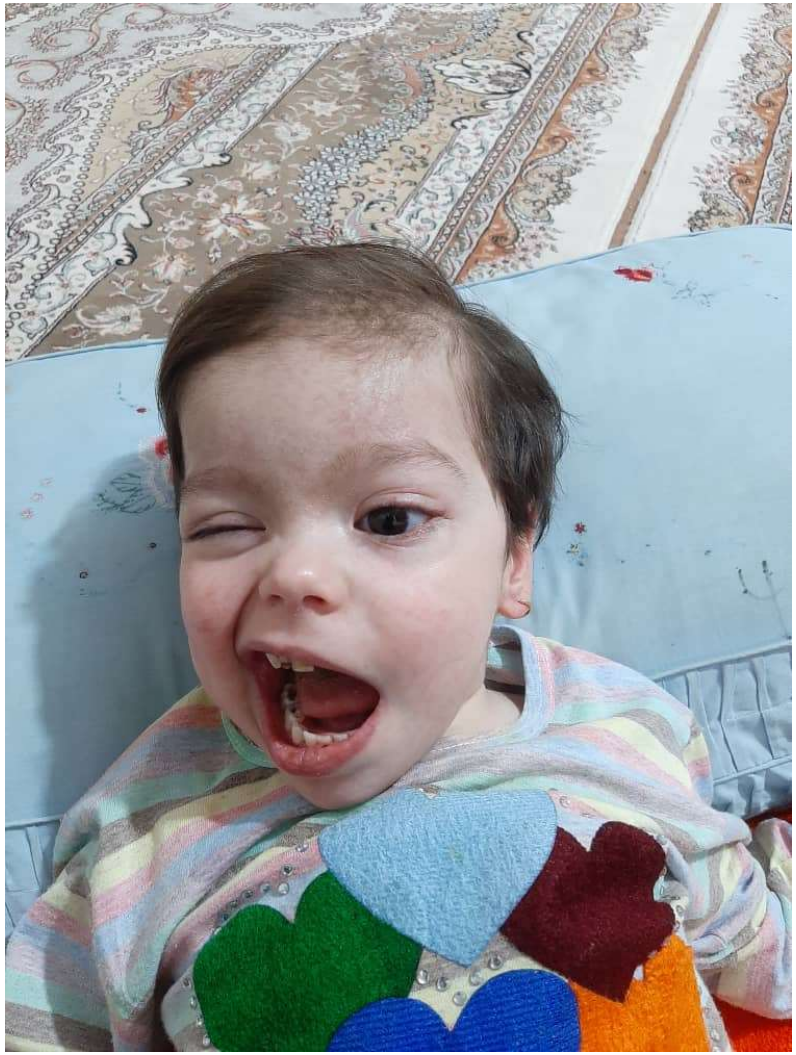












[Although childhood vasculitis is uncommon, it *is* ***an important component*** *of referrals to pediatric rheumatology clinics, and these children often require disproportionately large amounts of time and expertise. Diagnosis can be difficult, monitoring disease activity is problematic, and the outcome for some of the vasculitides may be serious or fatal due to cardiac disease (EGPA) or infections(GPA,SLE).*]

Ref :Text book of pediatric Rheumatology. Petty, laxer,...٨th edition,٢٠٢٠.

Clinical Features

- Constitutional symptoms (Fever, Weight loss, Fatigue,…)
- Skin lesions(palpable purpura , fixed urticaria ,livedo reticularis, nodules, ulcers)
- Neurological lesions
- Arthralgia/ Arthritis

Clinical Features

- Myalgia/ Myositis
- Hematuria, Renal failure
- Pulmonary hemorrhage
- Myocardial ischemia

Laboratory Features

- Increased ESR, CRP
- Leukocytosis , thrombocytosis, anemia
- C-ANCA(DX & Follow up of GPA), P-ANCA
- Elevated von Willebeand factor
- Cryoglobulnemia, Circulating immune complexes
- Hematuria

Secondary Vasculitis
Mimics of Vasculitis

- Drugs(Antithyroids, Anti-TNFs, Leflunomide)
- Toxic Exposure (cocaine, marijuana)
- Infections (subacute endocarditis, meningococcemia, HIV, CMV, EBV, HBV, Parvo B١٩,Herpes, TB, ….)

Secondary Vasculitis
Mimics of Vasculitis(continue)

- Malignancy
- Autoimmune inflammatory disorders (SLE, IBD,JDM, Sarcoidosis,….)
- Noninflammatory mimics (coarctation , Thrombocytopenia ,…)

[Although childhood vasculitis is uncommon, it *is* ***an important component*** *of referrals to pediatric rheumatology clinics, and these children often require disproportionately large amounts of time and expertise. Diagnosis can be difficult, monitoring disease activity is problematic, and the outcome for some of the vasculitides may be serious or fatal due to cardiac disease (EGPA) or infections(GPA).*]

Ref : Text book of pediatric Rheumatology. Petty, laxer,...Λ^{th} edition,٢٠٢٠.

Henoch-Schönlein Purpura

Clinical Manifestations:

- Nonthrombocytopenic purpura(۹۵-۱۰۰ %),
- gastrointestinal involvement(۶۰-۸۰%), arthritis(۵۰-۸۰%), nephritis(۳۰%).

Age: ۳-۱۵ years

- Boys/girls : ۱/۵/۱
- Etiology: infections, drugs, insect bites, dietary allergens

Renal disease &HSP

- Seldom precedes the purpura
- Develops within ۴-۶ weeks of the onset of the rash.
- Ranges from microscopic hematuria, and mild proteinuria to nephrotic syndrome.

Renal disease &HSP

- Increased risk of nephritis : Age>٧ years, persistent purpura, severe abdominal symptoms , decreased factor XIII activity .
- Follow up:
- All patients a minimum of ٩ months(U/A, Blood pressure)
- Patients with clinical nephritis for ٥ years.

Treatment (HSP)

Treatment is supportive with maintenance of good hydration, nutrition, and electrolyte balance; control of pain is accomplished with simple analgesics such as acetaminophen.

Short term glucocorticoid therapy is effective in relieving the pain of severe orchitis. Severe gastrointestinal disease or hemorrhage. The severity of disease may occasionally prompt the use of intravenous corticosteroids.

Pulmonary hemorrhage in HSP is an extremely rare and sometimes fatal complication, which requires aggressive immunosuppressive treatment, combining intravenous (IV) methylprednisolone with another immunosuppressive agent such as cyclophosphamide or cyclosporine, and supportive care.

Course of the Disease :

In two thirds of children, HSP runs its entire course within ۴ weeks of onset. Younger children have a shorter course and fewer recurrences than do older patients. *One third to half of the children have at least one recurrence that commonly consists of a rash and abdominal pain, with each episode usually being similar but briefer and milder than the preceding one.*

Course of the Disease(continue)

- Most exacerbations take place within the initial ۶-week period but may occur as late as ۲ years after onset. They may be spontaneous or coincide with repeated respiratory tract infections.
- The severity of the cutaneous leukocytoclastic vasculitis does not correlate with visceral involvement.

References

- Text book of pediatric Rheumatology. Petty, laxer,…٨th edition,٢٠٢٠.
- UPToDate
- Medscape Rheumatology

با تشکر از توجه شما