



DRUG OR DISEASE ?

Dr. Venkateswari. R.

Dr. Janani Sankar's unit

Kanchi Kamakoti CHILDS Trust Hospital



Acknowledgements:

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Dr. Ramkumar – Consultant Dermatologist

Dr. Ramprakash – Consultant Ophthalmologist

Dr. Prasad Manne – Consultant Cardiologist

Dr. V.K.Sairam – Consultant Nephrologist

Dr. BalaRamachandran – PICU team

- Southern Railway hospital

Dr. Kannan – Head of the Department and his team



HISTORY

- 16 year, adolescent girl
- Previously well

4 weeks back:

- Generalised maculopapular rash
- Swelling and pain involving multiple joints
- Fever



EVALUATED ELSEWHERE

High uric acid - ?Gout – started on allopurinol

- Arthritis settled: Rash persisted

Suspected connective tissue disorder


- Started on hydroxy chloroquine
- Received 2 doses of steroid in between



COURSE OF EVENTS

- 
- Sudden increase in severity of rash for 3 days

- 
- Associated involvement of eye and oral mucosa

- 
- ?Steven Johnson syndrome
 - Referred to us for further evaluation



ON EXAMINATION

- Febrile ,sick looking
- Facial rash
- Diffuse violaceous maculopapular rash involving palms and soles
- Severe mucositis (eye & oral mucosa involved)
- Joints – normal
- Vitals, BP – normal
- Systems – NAD

DIAGNOSIS

SLE

Disease flare

Drug rash

SJS

Drug

Allopurinol



INVESTIGATIONS

- Normal counts.
- Anemia (Hb-9.6 gm/dl)
- DCT 2+
- ESR – 90 mm/hr
- Urine microscopy – albumin 2+, 15-20 RBC's/HPF
- Serum creatinine – 1 mg/dl
- ECHO – mild pericardial effusion

- ANA, dsDNA, complement C3 – samples sent



CONSULT

Dermatologist

- Probable SLE / allopurinol induced SJS – suggested skin biopsy

Ophthalmologist

- Probable SJS



TREATMENT

- Supportive care
- IV Fluids
- Skin care
- Oral hygiene
- Eye care



COURSE IN THE HOSPITAL

- Developed large painful blisters over face
- Surgical consult
- Fluid aspirated
- Started on IV clindamycin



COURSE IN THE HOSPITAL

- ANA - 4+ speckled
 - anti ds – DNA – positive
 - C3 and C4 – low
- } confirmed SLE
- Started on pulse methylprednisolone
 - Nephrology consult – 24hrs urinary protein and proceed

SLE – 1997 REVISED CRITERIA

Malar rash

Discoid rash

Photosensitivity

Oral or nasal ulcers

Arthritis: Nonerosive, affecting 2 or more joints

Serositis: Pleuritis, pericarditis, peritonitis

Renal manifestations: Persistent proteinuria or cellular casts

Consistent renal biopsy

Seizure or psychosis

Hematologic manifestations: Hemolytic anemia

Leukopenia ($<4,000$ leukocytes/ mm^3)

Lymphopenia ($<1,500$ leukocytes/ mm^3)

Thrombocytopenia ($<100,000$ thrombocytes/ mm^3)

Immunologic abnormalities: Positive anti-double-stranded DNA or anti-Smith antibody test result

False-positive rapid plasma regain RPR test result, positive lupus anticoagulant test result, or elevated anticardiolipin immunoglobulin (Ig) G or IgM antibody

Positive antinuclear antibody test result



DAY 4

- Incidentally detected Bradycardia
- HR – 38/min
- ECG – AV dissociation

- Shifted to PICU



IN THE PICU

- Started on isoprenaline infusion
- Developed tachycardia with chest discomfort
- Isoprenaline infusion stopped

- Shifted to Southern Railway hospital for further care



FOLLOW UP:

- Responded to steroid – bradycardia improved
- Had a setback - seizures - controlled with fosphenytoin
- Discharged



DIAGNOSTIC POSSIBILITIES

Allopurinol induced SJS in Juvenile SLE

- Most likely

SLE presenting as SJS

- Less likely

References:

- Allopurinol is the most common cause of Stevens-Johnson syndrome and toxic epidermal necrolysis in Europe and Israel

Halevy S, Ghislain PD, Mockenhaupt M et al.

J Am Acad Dermatol 2008; 58:25–32.

- Steven johnson syndrome in a juvenile systemic lupus erythematosus patient.

Cavalcante EG, Guissa VR, Jesus AA, Campos LM, Sallum AM, Aikawa NE, Silva CA.

Lupus 2011 Nov;20(13):1439-41. Epub 2011 Jul 18.



References:

- Systemic lupus erythematosus presenting as Stevens-Johnson syndrome and toxic epidermal necrolysis: a report of three cases.

Lee HY, Tey HL, Pang SM, Thirumoorthy T

Lupus 2011 May;20(6):647-52. Epub 2010 Dec 10.



References:

- Complete atrioventricular block as initial manifestation of systemic lupus erythematosus
Clin Exp Rheumatol. 2009 Mar-Apr;27(2):344-6.
- Second-degree atrioventricular block as an early manifestation of adult systemic lupus erythematosus.
J Microbiol Immunol Infect 2005 Aug;38(4):296-9.
- High grade heart block in association with SLE
C S Edwards, R Mootoo, A Bhanji
Ann Rheum Dis 2004;63:606



THANK YOU