HENRY FORD HEALTH

# Acute Liver Injury as initial presentation of Adult-Onset Still's Disease

Yara Dababneh, MD <sup>1</sup>Susheela Sreedhar <sup>2</sup>, Ankita Nagirimadugu, MD <sup>3</sup>, Vivek Mendiratta, MD <sup>4</sup>, Dilip Moonka, MD <sup>3</sup>, Donard Haggins, MD <sup>5</sup> 1-Department of Internal Medicine.2-Wayne state University. 3-Division of Gastroenterology and Hepatology. 4-Department of

gastroenterology and Hepatology/OSU. 5-Department of Rheumatology

Henry Ford Health, Detroit, Michigan

#### Introduction

 We present a case of acute Liver injury as a first presentation of Adult-Onset Still's Disease (AOSD).

## **Case presentation**

- A 20-year-old man presented with fever and nausea.
- Liver function tests (LFTs) were notable for ALT 81 IU/L, AST 42 IU/L, ALP 58 IU/L and total bilirubin 6.7 mg/dL.
- Infectious work up was notable for positive EBV IgM that was treated appropriately.
- He presented again with worsening headache, fever, dark urine and petechial rash on both lower extremities.
- LFTs were still elevated. Patient was symptomatically managed.
- He presented again, nine days after initial symptoms onset with worsening symptoms, jaundice and new left hip pain with lower extremity weakness. Patient had taken one pill of Metronidazole from India but denied any other drugs.
- LFTs were ALT 214 IU/L, AST 215 IU/L, ALP 215 IU/L and total bilirubin 6.6 mg/dL.
- Patient's liver enzymes continued to rise, with ALT 186 IU/L, AST 146 IU/L, ALP 219 IU/L, total bilirubin 10.7 mg/dL and direct bilirubin 6.4 mg/dL. Therefore, a liver biopsy was ultimately performed.

## Case presentation, Cont.

- Histology showed a portal inflammatory process, with acute eosinophil spill over to the liver, without hepatic necrosis.
- Rheumatology team was consulted due to arthralgia.
- The patient met the Yamaguchi criteria for AOSD.
- He was started on prednisone 40 mg daily, with subjective and objective improvement with down trending LFTs.
- Outpatient follow-up with both rheumatology and hepatology continued to show significant improvement and down-trending LFTs.
- He continues to be on a prolonged steroid taper over 12 weeks...

### Discussion

- Contemporary opinion considers SOJIA and AOSD as a disease continuum with different ages of onset (before or after 16 years of age), but similar characteristics in regard to gene-expression, clinical course, prognosis, and responsiveness to therapy
- In our case the patient met the Yamaguchi criteria which involves major and minor criteria with transaminitis included.
- Major criteria includes fevers lasting more than 1 week, polyarthralgia, skin rash (nonpruritic salmon-colored maculopapular rash), and neutrophilic leukocytosis greater than 10,000.
- Minor criteria includes sore throat, splenomegaly, transaminitis, negative rheumatoid factor, and negative ANA.
- In this case, the main challenge was to find a suitable diagnosis that fits all the symptoms.
- A previous case report suggested that Drug Induced Liver Injury (DILI) might be the inciting factor in AOSD activation, which might be proposed in our case as well.
- It was reported that liver involvement in AOSD can be attributed to its association with Macrophage Activation Syndrome (MAS).
- Treatment mainly focuses on treating AOSD itself with systemic steroid therapy.
- In conclusion, acute liver injury is being increasingly reported in literature and this case supports acute liver injury in association with AOSD which can be aggravated by DILI or MAS.