



# Liver Function Abnormalities in Adult Onset Still's Disease : A Case Report

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## Introduction

- Adult Onset Still's disease (AOSD) has an estimated incidence of 0.16-0.4 per 100,000 people<sup>1</sup>.
- Hence, liver dysfunction due to AOSD is a rare entity with a wide spectrum from hepatomegaly and elevated transaminases to fulminant hepatic failure.
- Delayed diagnosis can lead to life-threatening liver failure. We report a case of liver dysfunction due to AOSD diagnosed after extensive evaluation.

## Case Description

- A 27-year-old African-American woman presented to the emergency department with 2 weeks of fever, chills, myalgia, non-bloody watery diarrhea, shortness of breath, and pleuritic chest pain, which started after a recent trip to Haiti.
- On exam, she was febrile (103 F), tachycardic, hypoxic, with tender bilateral axillary and inguinal lymphadenopathy.
- Treatment with antibiotics, methylprednisolone, and anti-tubercular therapy (RIPE) was initiated. She required intubation and vasopressor support due to worsened respiratory distress.
- Her liver enzymes were noted to be trending up after admission. RIPE therapy was modified but transaminases continued to rise. Extensive infectious and rheumatological workup was negative. Work-up for chronic liver disease was negative as well (table 1).
- She was immune to hepatitis A(IgM negative, IgG positive), immune to hepatitis B(HbsAg non-reactive, HbsAb reactive, HbcAb non-reactive) and hepatitis C antibody was negative.

**Table 1. Laboratory Investigations during Hospitalization**

TEST	ON ADMISSION	PEAK VALUE	ON DISCHARGE
WBC	10,870	33,710	8210
AST	91	625	40
ALT	19	294	82
ALP	48	1460	554
T. BIL	0.6	12.6	1.1
D. BIL	-	8.6	0.6
GGT	-	1136	-
CRP	120	168	<5
ESR	43	90	19
FERRITIN	>40,000	>40,000	518
RHEUMATOID FACTOR	12	-	-
ANA	NEGATIVE	-	-
HbeAb IgM	NON REACTIVE	-	-
HbeAb IgG	REACTIVE	-	-
HBV DNA	NOT DETECTED	-	-
ALPHA-1 ANTITRYPSIN	NORMAL	-	-
CERULOPLASMIN	NORMAL	-	-
ANTI-MITOCHONDRIAL	NEGATIVE	-	-
ANTI-LKM	NEGATIVE	-	-
ANTI-SMA	NEGATIVE	-	-

## Case Description

- A sonogram with doppler and MRCP showed normal hepatobiliary anatomy. Inguinal lymph node biopsy showed reactive hyperplastic patterns. A liver biopsy showed few nonspecific small lobular foci of spotty inflammation, necrosis and ballooning degeneration of hepatocytes.
- The patient was diagnosed with AOSD based on Yamaguchi criteria since she met 3 major and 3 minor criteria.
- Given lack of improvement with methylprednisolone, interleukin-1 $\beta$  inhibitor canakinumab was started. Inflammatory markers and liver enzymes improved significantly and she was discharged.

## Discussion

- AOSD is a rare disease with nonspecific signs and symptoms. Liver function abnormalities are the most common manifestation in AOSD.
- Liver involvement can occur in the absence of other features of the disease and can delay diagnosis. Diagnosis is based on clinical criteria.
- Treatment is largely based on expert opinion since controlled trials are lacking. The most effective treatment is corticosteroids and biologic therapy.
- This case highlights the need for a high index of suspicion for AOSD when abnormal liver enzymes are seen in the context of nonspecific signs and symptoms.

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## References

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