Atypical adult-onset Still's disease with an initial and sole manifestation of liver injury: A case report and review of literature

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Abstract

BACKGROUND
Adult-onset Still's disease (AOSD) typically presents with a high spiking fever, polyarthritis, transient maculopapular rash, neutrophilic leukocytosis, and hepatosplenomegaly. It has a wide spectrum of clinical symptoms ranging from mild to severe, with extensive involvement of almost every organ. Although liver involvement in the form of increased hepatic enzymes and bilirubin is common, no AOSD case with liver involvement as the initial manifestation of AOSD has been reported.

CASE SUMMARY
A 35-year-old woman presented to the hepatology department with progressively worsening jaundice for one week. Liver chemistry tests revealed a significantly increased liver enzymes and bilirubin level. Given that the clinical examination was unremarkable, liver biopsy was considered because the patient had a history of AOSD 6 years ago. Liver histopathology revealed that most hepatic lobules were still recognizable. Fusional necrosis was observed around most central veins. A few bridging necrotic zones were also found. Infiltration of multiple plasma cells were observed in the necrotic zone, and the reticular scaffold was still expanded. Additionally, no obvious fibrosis was observed in the portal area. Mild mixed inflammatory cell infiltration was noted in the interstitium of the portal area. Further examination was unremarkable except for a remarkably high level of ferritin. Collectively, a presumptive diagnosis of liver injury secondary to AOSD was made. The hepatic involvement responded well to glucocorticoid treatment.

CONCLUSION
This case highlights that hepatic involvement as an initial and sole manifestation could be a pattern of relapsed AOSD. The diagnosis of AOSD should be
considered in the case of nonresolving liver injury after the exclusion of common etiologies for liver diseases. A liver biopsy can be useful for the differential diagnosis of liver injury associated with AOSD.

**Key Words:** Adult-onset Still’s disease; Liver injury; Liver biopsy; Histopathology; Glucocorticoid treatment; Case report

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**Core Tip:** Liver involvement in the form of increased hepatic enzymes and bilirubin is common in adult-onset Still’s disease (AOSD). Herein, we presented a patient with relapsed AOSD with hepatic involvement as an initial and sole manifestation responding well to glucocorticoid treatment. This case highlights that hepatic involvement as an initial and sole manifestation could be a pattern of relapsed AOSD. The diagnosis of AOSD should be considered in the case of nonresolving liver injury after the exclusion of common etiologies for liver diseases. A liver biopsy can be useful for the differential diagnosis of liver injury associated with AOSD.

**INTRODUCTION**

Adult-onset Still’s disease (AOSD) is a rare condition characterized by a high spiking fever, polyarthritis, transient maculopapular rash, and neutrophilic leukocytosis\(^4\)\(^5\). It has a wide spectrum of clinical symptoms ranging from mild to severe, with extensive involvement of almost every organ. Moreover, life-threatening complications, such as myocarditis and disseminated intravascular coagulopathy (DIC), appear in approximately 15%-20% of patients with AOSD\(^5\). The early diagnosis and intervention of AOSD are vital to achieving sustained clinical remission and reducing the mortality rate\(^5\). However, early identification of AOSD in some cases remains a challenge because of highly varied symptoms, the involvement of multiple organs, and the lack of diagnostic tests and serologic markers\(^6\). Although hepatic involvement is commonly observed in AOSD with a prevalence of 50%-75%\(^6\), liver injury is not a prerequisite to diagnose AOSD according to the Yamaguchi’s criteria\(^9\), partly because many diseases can cause liver injury. Thus, it remains a challenge to decide whether the involvement of the liver as the initial organ is caused by AOSD or other etiologies. To date, no AOSD case with liver involvement as the initial manifestation of AOSD has been reported. We here present a patient with relapsed AOSD with hepatic involvement as an initial and sole manifestation.

**CASE PRESENTATION**

**Chief complaints**

A 35-year-old woman presented to the hepatology department with progressively worsening jaundice.

**History of present illness**

Patient’s symptoms started a week ago with progressively worsening jaundice. The patient denied fever, anorexia, fatigue, and abdominal distention.

**History of past illness**

The patient had a history of AOSD 6 years previously and responded well to standardized glucocorticoid therapy in the first episode, but experienced a relapse 5

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**Yu F et al. Liver injury secondary to AOSD**

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mo before the self-discontinuation of methylprednisolone. She had no risk factors for viral hepatitis or toxin. She does not smoke and drink. Her family history was not notable for autoimmune diseases or hepatic virus diseases. She had no risk factors for viral hepatitis.

**Personal and family history**

No abnormalities.

**Physical examination**

The patient’s temperature was 36.7 °C, heart rate was 84 bpm, respiratory rate was 18 breaths per min, blood pressure was 116/75 mmHg and oxygen saturation in room air was 100%. The clinical examination revealed that the skin and sclera were jaundiced.

**Laboratory examinations**

Liver chemistry tests revealed a significantly increased alanine transaminase (ALT) level (1009.71 IU/L; normal: 5-40 IU/L), aspartate transaminase level (455.41 IU/L; normal: 5-40 IU/L), gamma-glutamyl transferase level (721.85 IU/L; normal: 8-57 IU/L) and bilirubin level (137.59 µmol/L; normal: 0-21 µmol/L). Peripheral blood revealed a mildly elevated white blood cell (WBC) count (11.6 × 10⁹/L; normal: 4-10 × 10⁹/L) with 76.5% neutrophils (normal: 50.0%-70.0%) and a C-reactive protein (CRP) level of 31.22 mg/dL (normal: 0-8 mg/L). Results of other liver markers are demonstrated in Table 1. Full-blood screening analyses to determine the etiology of liver involvement were all negative or within normal limits.

**Imaging examinations**

Ultrasound showed a smoothly contoured liver and healthy thin-walled gallbladder. Thorax and abdominal computed tomography (CT) scans demonstrated generalized lymph nodes with the largest one measuring 1.1 cm × 1.2 cm; no solid masses were identified. Echocardiography and serum protein electrophoresis were normal.

**Histopathology examination**

Liver histopathology revealed that most hepatic lobules were still recognizable. Fusional necrosis was observed around most central veins. A few bridging necrotic zones were also present. Infiltration of multiple plasma cells was observed in the necrotic zone, and the reticular scaffold was still expanded, without obvious collagen deposition (Figure 1A). Additionally, no obvious fibrosis was observed in the portal area. Mild mixed inflammatory cell infiltration was noted in the interstitium of the portal area. Interfacial inflammation was not significant (Figure 1B).

**MULTIDISCIPLINARY EXPERT CONSULTATION**

Given that the clinical examination was unremarkable, liver biopsy was considered by the hepatology team at this point. However, it was decided to await rheumatology review because the patient had a history of AOSD 6 years ago, with symptoms including unexplained fever, arthritis, rash, and neutrophilic leukocytosis. Additionally, laboratory tests showed a remarkably high level of CRP and ferritin, and elevated liver enzymes. The patient responded well to standardized glucocorticoid therapy in the first episode but experienced a relapse 5 mo before the self-discontinuation of methylprednisolone. At the rheumatology review, the patient did not describe any systemic symptoms but felt constitutionally unwell with significant fatigue. There was no history of Raynaud’s phenomenon or any features suggestive of other autoimmune connective tissue diseases or vasculitis. Systemic examination was unremarkable except for a remarkably high level of ferritin (> 1650.0 ng/mL). After a discussion with the rheumatology team concerning diagnosis options, liver histological examination was still needed to determine whether the liver injury was caused by AOSD or liver disease per se (Figure 2).

**FINAL DIAGNOSIS**

There were two reasons why we considered liver injury as a result of relapsed AOSD rather than liver diseases. First, liver involvement is a well-characterized feature of AOSD, with highly varied manifestations ranging from minimally elevated hepatic
Table 1 Results of serologic tests

| Serologic tests                                           | Negative/positive |
|-----------------------------------------------------------|-------------------|
| Anti-HAV                                                  | Negative          |
| Anti-HAV                                                  | Negative          |
| HBsAg                                                    | Negative          |
| Anti-HBs                                                  | Positive          |
| Anti-HBc IgM                                              | Negative          |
| Anti-HCV                                                  | Negative          |
| Anti-HEV                                                  | Negative          |
| Anti-EBV-VCA IgM                                          | Negative          |
| Anti-CMV IgM                                              | Negative          |
| Anti-HIV                                                  | Negative          |
| HCV-RNA                                                   | Negative          |
| Anti-nuclear antibody                                     | Negative          |
| Anti-HIV                                                  | Negative          |
| Anti-nuclear antibody                                     | Negative          |
| Anti-smooth muscle antibody                               | Negative          |
| Anti-soluble liver antibody                               | Negative          |
| Antineutrophil cytoplasmic antibody                       | Negative          |
| Antimitochondrial antibody                                | Negative          |
| Anti-liver-kidney microsomal type-1 antibody              | Negative          |

HAV: Hepatitis A virus; HBsAg: Hepatitis B surface antigen; Anti-HBc: Hepatitis B core antibody; IgM: Immunoglobulin M; HCV: Hepatitis C virus; HEV: Hepatitis E virus; EBV: Epstein Barr virus; VCA: Viral capsid antigen; CMV: Cytomegalovirus; HIV: Human immunodeficiency virus.

Figure 1 Liver histopathology. A: Infiltration of multiple plasma cells was observed in the necrotic zone, and the reticular scaffold was still expanded, without obvious collagen deposition; B: Mild mixed inflammatory cell infiltration was noted in the interstitium of the portal area. Interfacial inflammation was not significant.

enzymes to hyperbilirubinemia and even fulminant hepatic failure. However, the liver injury typically appears as one of the disease activity signs rather than as the only manifestation in patients with AOSD\(^2\), further highlighting the importance of histological examination. Second, both fevers with unknown causes and elevated ferritin levels are features of active AOSD. Fever with unknown causes in Europe accounted for 3% - 20% of AOSD\(^1\),\(^2\). A retrospective study in China involving 517 individuals revealed that fever occurred in 472 (91.3%) patients with AOSD who satisfied the Yamaguchi criteria\(^3\). Additionally, hyperferritinemia still served as a marker of disease activity in AOSD\(^4\), although its specificity remains to be confirmed\(^5\). Collectively, given the patient’s presenting features, especially the liver
histological findings, in combination with the history of AOSD, a presumptive diagnosis of liver injury secondary to AOSD was made.

**TREATMENT**

After early treatment with intravenous methylprednisolone (80mg/d) for three days, the patient was switched to oral steroids (starting at prednisolone 40 mg daily), followed by a tapered dose. Additionally, she was referred for intensive physiotherapy, which is an essential part of overall management.

**OUTCOME AND FOLLOW-UP**

In response to treatment, the liver enzymes were normalized within four weeks and remained normal with symptom improvement (Figure 3). The patient also reported considerable improvements in anorexia and fatigue. Six months after discharge, the patient’s subsequent clinical course was stable, while the dose of oral methylprednisolone continued at 4 mg/d.

**DISCUSSION**

Although hepatic involvement is a well-characterized feature in patients with AOSD, it is uncommon for the prevalence of liver injury as the initial manifestation of AOSD. For the first time, we present an AOSD case with elevated hepatic enzymes and bilirubin as an initial and sole manifestation that was successfully treated with methylprednisolone.

It remains a challenge to make a differential diagnosis for hepatic enzyme elevations because many causes could induce liver injury. Seven AOSD cases with increased liver enzymes have been previously reported. The etiologies of liver injury in these cases included NSAID-induced liver injury (n = 2), hepatic injury associated with AOSD itself (n = 2), and AOSD with concurrent autoimmune hepatitis (n = 3).
The largest challenge to identify the etiologies of liver injury in patients with AOSD is the lack of specific biomarkers and pathological findings. Highly variable hepatic pathological features in AOSD have been previously reported, ranging from mild portal inflammatory cell infiltration and Kupffer cell hyperplasia\cite{20,21,26-28} to portal fibrosis and massive or submassive hepatic necrosis\cite{29-33}. Taken together, no histological finding has been identified to be specific for liver injury secondary to AOSD. Although the value of liver biopsy in diagnosing AOSD remains debatable\cite{34}, the liver histological findings may provide valuable information to rule out liver disorders induced by viruses, autoimmune factors, and drugs, facilitating the early detection of AOSD and subsequent prompt intervention with methylprednisolone\cite{29}.

Several potential mechanisms responsible for liver injury in AOSD have been proposed\cite{33}. Notably, the serum interleukin-18 (IL-18) concentration is markedly increased in patients with AOSD and active hepatitis and correlates with serum aminotransferase levels\cite{35,36}. Moreover, previous studies have found a marked increase in IL-18 expression by activated macrophages and Kupffer cells within the liver parenchyma of a patient with AOSD\cite{37}. However, it remains unknown whether serum IL-18 serves as an early predictor of liver injury in patients with AOSD\cite{38}.

Collectively, liver involvement in the form of elevated bilirubin and liver enzymes can be an initial and sole presentation of relapsed AOSD which responds well to glucocorticoid treatment. Therefore, the diagnosis of AOSD should be considered in the case of nonresolving liver injury after the exclusion of common etiologies for liver diseases.

**CONCLUSION**

Although liver involvement in the form of increased hepatic enzymes and bilirubin is common in AOSD patients, no AOSD case with liver involvement as the initial manifestation of AOSD has been reported. Liver involvement in the form of elevated bilirubin and liver enzymes can be an initial and sole presentation of relapsed AOSD which responds well to glucocorticoid treatment. Therefore, our study suggests that the diagnosis of AOSD should be considered in the case of nonresolving liver injury after the exclusion of common etiologies for liver diseases.
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