# A Rare Case of Isolated ALECT-2 Hepatic Amyloidosis

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## Background

- Amyloidosis is a rare disease defined by deposition of extracellular fibrils from immunoglobulin light chains which results in organ dysfunction[1]
- Many patients with primary systemic amyloid have hepatic involvement, but isolated hepatic amyloid deposition is very rare[2,3]
- Hepatic amyloid deposition is characterized by mild hepatomegaly, occasionally elevated liver enzymes, increased echogenicity on ultrasound or decreased attenuation on CT[4,5]
- Biopsy is the gold standard for diagnosis, seen as extracellular amorphous material, and Congo red staining will reveal apple-green birefringence under polarized light
- Leukocyte cell-derived chemotaxin 2 amyloidosis (ALECT-2) is a novel amyloid subtype, previously thought to be found in renal amyloid with a predominance in Hispanic patients but has recently been reported in the liver[6]

## Patient Presentation

**53-year-old Hispanic female with history of diabetes mellitus, hyperlipidemia, and nonalcoholic fatty liver disease presented to hepatology clinic for elevated liver enzymes.**

**Initial Workup:**

- AST was 36U/L, ALT 67 U/L, ALK P 67 U/L
- Positive ASMA (22U), IgG 1049 mg/dL, normal ANA screen, negative AMA, creatinine 0.71mg/dL
- CT with post-cholecystectomy changes and calcified granuloma but otherwise normal
- Underwent liver biopsy, which demonstrated focal amyloid deposition with positive findings on Congo red stain consistent with amyloidosis as well as mild steatosis, mild lobular activity, and rare ballooning hepatocytes without fibrosis
- Positron emission tomography-CT, bone marrow biopsy, urine and serum protein electrophoresis were unremarkable
- Further amyloid testing showed leukocyte chemotactic factor-2 (ALECT-2) amyloidosis

**Subsequent Management and Follow-up:**

- A referral for second opinion and experimental treatment options was recommended, however the patient declined. The patient has had stable lab values over the last several years with observation

## Images and Histology

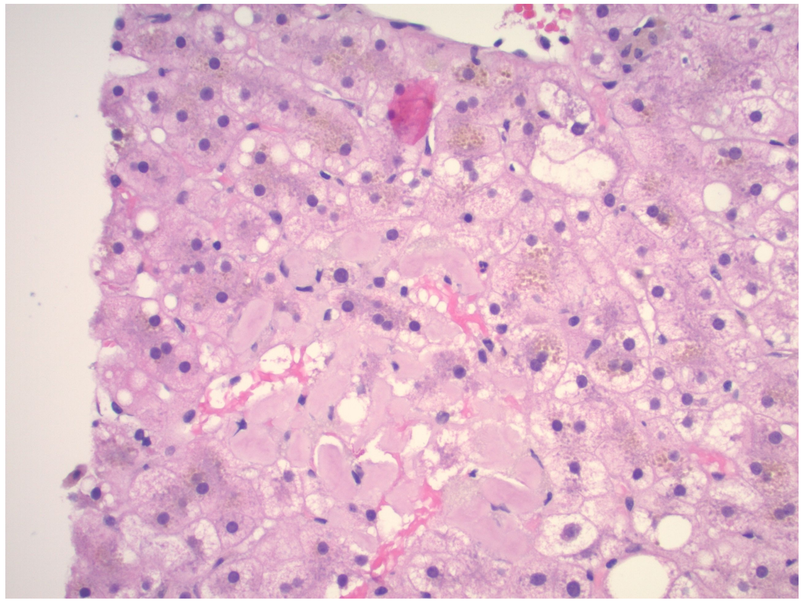

**Figure 1**: Hematoxylin and eosin stain at high magnification demonstrating hepatic amyloidosis. The pink, amorphous, acellular material seen is amyloid

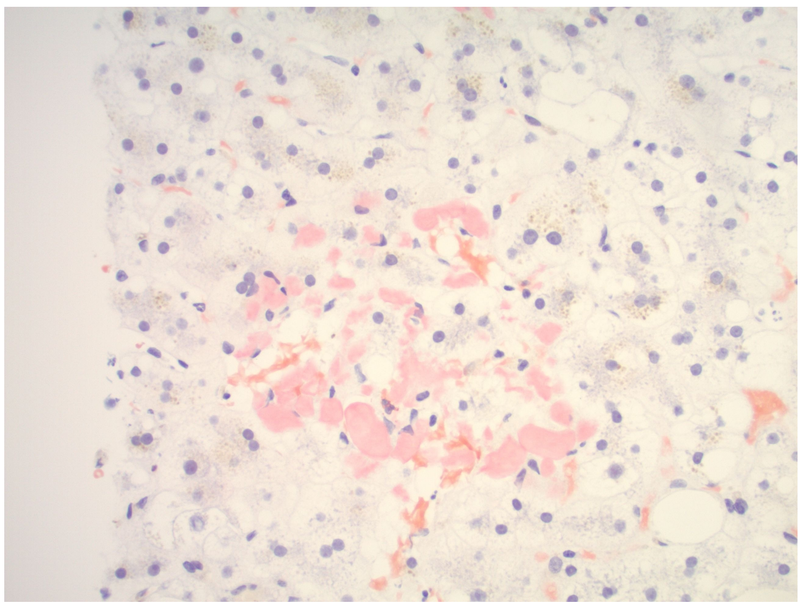

**Figure 2**: Hepatic sample with Congo Red staining. The salmon-pink material is amyloid in a globular configuration, which is typical for ALECT-2

## Lab Values and Disease Course

| Time | Initial | Year 1 | Year 2 | Year 3 |
|---|---|---|---|---|
| Total protein (g/dL) | 7.3 | 7.2 | 7.7 | 7.1 |
| Albumin (g/dL) | 4.6 | 4.4 | 4.6 | 4.4 |
| Total bilirubin (mg/dL) | 2.7 | 2.7 | 3.2 | 2.4 |
| Alkaline phosphatase (IU/L) | 67 | 66 | 61 | 63 |
| AST (IU/L) | 36 | 57 | 34 | 20 |
| ALT (IU/L) | 67 | 96 | 54 | 33 |

## Discussion

- This is a rare case of isolated hepatic amyloidosis, highlighting the need for maintaining a broad differential diagnosis in a patient presenting with elevated liver enzymes
- Given the rare nature of this condition, it is important to highlight this patient's presentation and three-year outcomes with observation given lack of available treatment regimens
- ALECT-2 may have a more indolent clinical course compared to other amyloidosis types[8], and differentiating it from other causes of amyloidosis avoids other harmful treatments

## References

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