

POSTER 2

PRIMARY AMYLOIDOSIS – A CASE REPORT AND FUTURE INVESTIGATIONS

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Primary amyloidosis is an uncommon and devastating disease characterized by extracellular tissue deposition of insoluble amyloid fibrillar monoclonal light chains. The disease primarily affects the heart and kidneys and rarely presents with primary hepatic involvement. We report a case of a previously healthy 54-year-old patient with primary systemic amyloidosis presenting as liver failure. The patient was transferred from an outside hospital with a 6-week history of anasarca, ascites, jaundice, and tender hepatomegaly. At the time of admission, renal function and echocardiogram were normal. Extensive laboratory and radiology studies ruled out viral, drug, and cancerous causes of liver failure. Liver and bone marrow biopsy stained with congo red demonstrated apple-green birefringence under polarized light, and additional immunological studies confirmed primary systemic amyloidosis involving the liver and bone marrow. Due to systemic involvement the patient was not a candidate for liver transplant and he elected to return home on palliative care. Currently, life-extending therapies for this disease are limited although successful treatment with liver transplantation has been documented in rare cases. Given the infrequent incidence, devastating impact, and variety of presentations, it is necessary to analyze each case in an effort to achieve better outcomes such as earlier diagnosis, shorter hospital stays, and improved pain control at end of life.