Reviewer 05394499：

Dear Authors, this is an interesting case of a rare combined presentation of two genetic/metabolic diseases in the same patient. The description of the clinical presentation and the paraclinical findings was adequate. Please elucidate the following minor points of interest:

1) please provide the normal range for kappa and lamda light chain in the text,

Answer: Thank you for your comments. Reference ranges of serum, urine kappa and lamda light chains have been given in Table1.

Analyte / Date	2018.11	2020.07	2021.08	2021.12	2022.01	2022.03	2022.04	2022.05	Reference range
WBC(*10⁹/L)	1.14	14.94	10.1	9.6	13.9	9.6	7.8	8.5	3.5-9.5
Hb(g/L)	83	97	96	86	97	91	88	95	115-150
PLT(*10⁹L)	33	359	253	316	267	377	361	345	125-350
ALT(U/L)	58.2	33.6	56.1	81.9	66.2	716	42.8	31.6	7-40
AST(U/L)	136.4	108.6	175.8	236.5	315.2	298.3	211.9	125.8	13-35
TBIL(umol/L)	66.1	179	14.2	46.9	67.2	36.9	33.2	23.04	<23
DBIL(umol/L)	47.8	128.6	9	31.85	39.2	26.7	18.39	13.81	≤8
ALB(g/L)	35.3	24.6	16.9	25.86	22.71	26.39	25.33	27.58	40-55
ALP(U/L)	501	1174	426	904	887	1160	875	978	35-100
GGT(U/L)	268.26	1367	439.51	387.3	971.19	554	382	542.22	7-45
ChE(U/L)	2796	2301	3958	4623	3112	4728	2738	3288	5000-12000
CREA(umol/L)	48	49.83	56.87	/	96.6	66	68	46.9	41-73
NT-proBNP(pg/ml)	/	176	238.4	/	3020	475	/	110.9	41.4-153
cTnT(ng/ml)	0.003	0.009	/	/	0.645	0.465	/	0.001	0-0.014
LDH(U/L)	136	244	279	426	295	481	364	290	120-250
uPRO	/	Positive (2+)	Postive (3+)	Postive (3+)	Positive(3+)	Positive (3+)	Positive(2+)	Positive(1+)	Negative(-)
uALB(mg/L)	/	964	/	/	744.98	/	/	352.17	<140
Serum κ light chain(mg/dl)	/	/	/	/	1050	1710	689	649	629-1350
Serum λ light	/	/	/	/	537	916	330	249	313-723

chain(mg/dl)									
Urinary κ light chain(mg/dl)	/	/	/	/	52	72.4	/	4.08	0-1.85
Urinary λ light chain(mg/dl)	/	/	/	/	37.9	62.5	/	<5	0-5
Drug	UDCA					UDCA+Daratumumab			

2) please refer that you did not conduct a heart muscle biopsy but based on the context you highly suspected a cardiac amyloidosis,

Answer: Thank you for your comments. Although heart involvement was suspected, the patient and his family refused the myocardial biopsy considering the danger, but the diagnosis basis of primary light chain (κ) amyloidosis was very sufficient.

3) please explain the reason for the initial splenectomy when only a cholecystectomy was indicated.

Best Regards

Answer: Thank you for your comments. Preoperative blood routine showed significant decrease of platelet (PLT33*10^9/L). Considering that the giant spleen led to hypersplenism.

Reviewer 04213276:

An interesting case of liver involvement of systemic amyloidosis and concomitant cholestasis potentially due to ABCB4 heterozygous gene mutations. Comments:

1. What was the liver histology pattern? was it consistent with changes in patients with ABCB4 gene mutations?

Answer: Thank you for your comments. Liver histology pattern:Hepatic lobule structure is disordered and pseudolobule is formed; Regional balloon-like degeneration of hepatocytes, scattered focal necrosis and fusion necrosis can be seen; Portal area is enlarged, fibrous tissue proliferates, fibrous septa are formed, bridging fibrosis and pseudolobular formation can be seen, inflammatory cell

infiltration dominated by mononuclear cells, interlobular bile duct hyperplasia and moderate interfacial inflammation can be seen.

The mutation of the ABCB4 gene has the potential to result in cholestasis and injury to the bile duct. The observable symptoms include the presence of stones in the gallbladder or intrahepatic bile duct, as well as the recurrence of jaundice. It is a recessive genetic disease that affects the chromosomes and is characterized by progressive familial intrahepatic cholestasis,Known as progressive familial intrahepatic cholestasis (PFIC-3), a small number of patients may progress to portal hypertension, cirrhosis and even liver failure.

The liver tissue's pathological biopsyin in the patient exhibited bile duct injury, cholestasis, and cirrhosis, which were in line with cirrhosis caused by PFIC-3, in conjunction with elevated GGT and ALP levels.

2. The discussion section is rather small. More regarding the characteristics of both these types of lver injury and how they can be identified and treated should be added.

Answer: Thank you for your comments. We have revised the discussion section

3. what was the type of amyloidosis?

Answer: Thank you for your comments. The type of amyloidosis is primary light chain (κ) amyloidosis.

4. There are significant problems with the syntax of the manuscript with quick references of results without connections between phrases or paragraphs. The authors should explain more of their train of thought that led to the exainations that provided the final diagnosis, rather than list a number of tests without commentary.

Answer: Thank you for your comments. We have conducted a comprehensive re polishing of the article and have it reviewed by relevant experts in English native language. We have reorganized the entire text and focused on diagnosis and treatment ideas and marked it in green.