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Brief report

A case of hepatosplenic T-cell lymphoma confirmed by autopsy

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Background : Hepatosplenic T-cell lymphoma was one of the most troublesome lymphomas because of the difficulty of accurate diagnosis, the worst prognosis of less than 2 years, and the rarest lymphomas, less than 1 % in whole lymphomas in frequency and. therefore, they were found in the end stage. We experienced one case diagnosed after autopsy and reported in this paper.

Case : The case was 93 y/o male patient, registered as SN 06-013 in our laboratory, suffering from (1) carcinoma of stomach treated with gastrectomy on 19 years ago and (2) polycythemia vera treated with melphalan for 13 years (Fig. 1). Splenomegaly of mild degree was confirmed on computed tomography scan and was due to polycythemia. After 6 years of hemodialysis for chronic renal failure he was suddenly died in less than one month with thrombocytopenia , jaundice , and marked splenomegaly. There was no evidence of myelodysplastic syndrome, leukemia, and lymphoma. On autopsy we revealed the followings macroscopically : prominent splenomegaly (730 g) without any macroscopic mass, cavity effusions, and old myocardial infarct. Liver remained in normal size, 840 g. There was no nodal swelling and no peripheral blood abnormality. Microscopically both the red pulps of spleen and the sinusoids of liver were occupied with medium-sized lymphocytes, which was negative for both T-cell γ rearrangement and bcr-abl in in situ hybridization. He was diagnosed as hepatosplenic T-cell lymphoma by extranodal immunostaining with T-cell intracellular antigen-1 (TIA-1, anti-cytotoxic granule associated protein) antibody (Fig.2,3). Bone marrow was normocellular or mildly cellular marrow with few atypical lymphocytes. Intrahepatic cholestasis was also found.

Conclusion : Thrombocytopenia was more consistent with lymphoma rather than polycythemia vera. On diagnosing cases with splenomegaly it was very important to keep hepatosplenic T-cell lymphoma in mind.

Key words : hepatosplenic T-cell lymphoma, T-cell intracellular antigen-1, TIA-1, immunostain, thrombocytopenia, polycythemia vera

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和　文　抄　録

短報

剖検後に診断された肝脾T細胞リンパ腫の1症例

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背景：肝脾T細胞リンパ腫は、発生頻度がリンパ腫の1％以下と極めて稀で、臨床検査上において異常細胞を同定することが困難とされ、発症後2年以内に死亡する予後不良なことより、病期終末に診断される。今回、肝脾T細胞リンパ腫の1症例を経験したので報告した。

症例内容：症例は93才男性で、13年来真性多血症を先行し、メルファラン治療を受けていた。6年間の腎不全・血液透析後、血小板減少症と黄疸を発症し1ケ月の急性経過で死亡した。脾腫が著明で、免疫組織染色より、肝脾T細胞リンパ腫と診断された。

結論：脾腫症例においては、肝脾T細胞リンパ腫の可能性を念頭に置くことが重要と思われる。

キーワード：　肝脾T細胞リンパ腫、TIA－1陽性、血小板減少症、真性多血症

Fig. 1 Clinical laboratory results

		1987/3	2000/3	2004/3	2006/6/6	2006/7/8	2006/8/1	2006/8/10
total bilirubin	＜1.24				1.03	2.2	3.23	11.9
direct bilirubin								8.94
aspartate aminotransferase, AST	13－33				48	63	79	82
alanine aminotransferase, ALT	8 －42				36	38	55	44
alkaline phosphatase, ALP	104－338				354	398	493	365
lactate dehydrogenase, LDH	124－226	498			318	301	327	508
choline esterase, ChE	97－249					62		
blood urea nitrogen, BUN	7.5－20.4		23.3	56.2	42.2	33.3	76.1	106.6
creatinin	0.75－1.34		1.08	3.82	4.17	6.68	6.43	6.3
total protein	6.5－8.2				5.8	5.6	5.6	5.2
albumin %	61.4－72.8				60.8	55.8	55	56.4
α1 %	1.3－2.9				2.9	3.2	2.8	2.5
α2 %	5.4－10				6.3	6.3	5.8	5.5
β%	6.5－10.8				8	7.2	6.3	8
γ%	9.7－20.2				22	27.5	30.1	27.6
IgG	870－1700						1513	
IgA	110－410						285	
IgM	34－220						191	
serum iron	80－200				146		178	
unsaturated iron binding capacity, UIBC	111－255				86		7	
alminium	＜16.1						10	
blood sugar	70－139				109	128	50	
C-reactive protein, CRP	0 －0.27				0.11	0.52	0.42	0.38
transferrin	190－320				179		128	
ferritin	26－240				61.3		155.7	
β2 －microglobulin	1 －1.9				30.6		61.94	
red cell count, RBC	404－559	872			447	515	436	330
hematocrit, Hct	13.5－17.6	68			13.8	16.5	13.7	10.6
hemoblobin, Hb	40.5－51.8	21.5			45.8	52.1	42.7	31.6

mean corpuscular volume, MCV	86.3－104.2				102.5	101.2	97.9	95.8
mean cell hemoglobin, MCH	29－34.9				30.9	32	31.4	32.1
mean cell concentration, MCC	32.2－35.6				30.1	31.7	32.1	33.5
reticulocyte	0.2－2.7				1.37	0.99	1.14	1.29
platelet, Plt	15.1－32.5	40			18.4	7.3	1.6	1.1
white blood cell, WBC	38.5－98.7	16310			159.3	115.5	66.8	61.4
neutrophil	40.7－71.6				93.4	90.7	85.2	88
metamyelocyte %	0							7
stab cell %	3 － 6				23	24	19	20
segmented cell %	45－55				72	70	69	67
lymphocyte	21.1－50.9				3.7	4.9	9.1	7.8
atypical lymphocyte	0				0	0	0	
neutrophil alkaline phosphatase score	150－350	324						
abnormal RBC							＋	＋
parathyroid hormone, PTH	10－65				164			
erythropoietin in serum	8 －36	8.6						
erythropoietin in urine	0.7－3.48	＜5						
vitamin B12	＜800	880						
proteinuria			＋＋					
anti-adult T-cell leukemia （ATL） antibody								－
IL－6	＜8pg/ml							297
anti-platelete antibody								－
hepatitis B virus related							－	
hepatitis C virus related							－	
Treponema pallidum hemagglutination test, TPHA							－	
prothrombin time, sec	10.5－13.5							25.2
activated partial thrombplastin time, sec	23－37							70.1
thrombo test	70－130							35.1
fibrinogen	180－370							65
fibrin/fibrinogen degradation products, FDP	＜4.9							7.4
antithrombin III, ATIII	75－125							102

Fig. 2　Immunostaining results

reagent / cells from	liver , spleen, bone marrow
CD45	＋
terminal deoxynucleotidyl transferase, TdT	－
CD79a	－
CD20	－
CD3	＋
CD45RO	＋／－
CD43	－
CD8	＋／－
CD4	－
CD56	－
glycopholin A	－
neutrophil elastase	－
peroxidase	－
CD13	－
T-cell intracellular antigen－1, TIA－1	＋
granzyme B	－

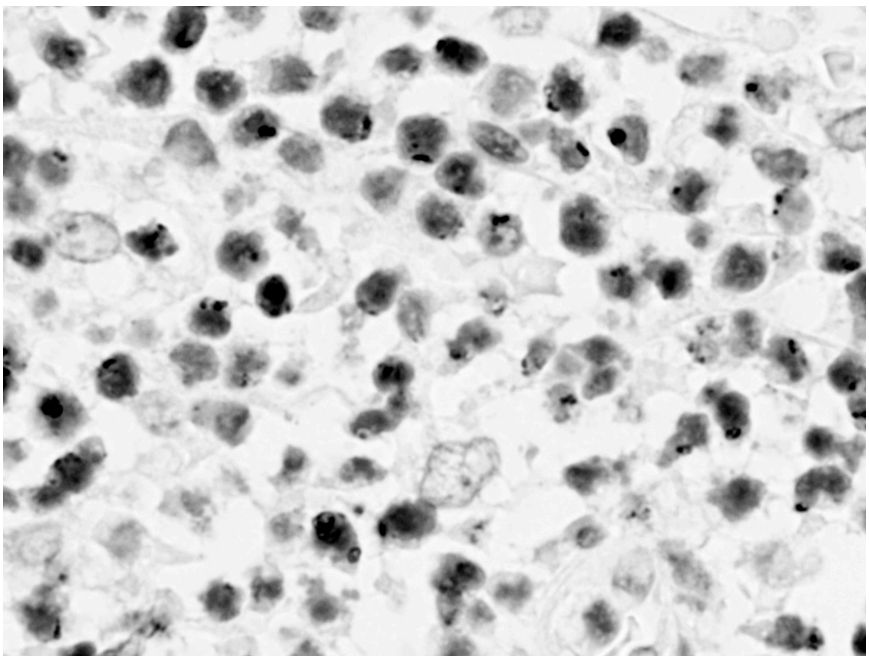

Fig. 3　Immunostain of lymphocytes in spleen with anti-TIA-1 antiboty, dotted positive staining