Dear editors,

Thank you for the reviews of the manuscript. We have carefully read the editors' and reviewers' comments and revised our manuscript accordingly. Herein we list point-by-point our response to each of the issues that was raised in the peer review report and highlight in our manuscript.

Reviewer #1:
Scientific Quality: Grade B (Very good)
Language Quality: Grade B (Minor language polishing)
Conclusion: Minor revision
Specific Comments to Authors: 1. Kindly write according to journal criteria- abstract and case report subheadings should be revised 2. In abstract and case report- patient age mentioned 84 year and in table 2 -83 year- kindly check and correct 3. kindly quote references in table 2 regarding every case. 4. In discussion- AITL-associated HLH had a poor prognosis due to aggressive disease course, opportunistic infections compared with EBV-associated HLH[18].---- not in line with reference 18, kindly check.

Firstly, we thank the reviewer for the positive comments.

1. Kindly write according to journal criteria- abstract and case report subheadings should be revised

Response: We thank the reviewer for the kind suggestion. We have revised the running title and explained the abbreviation NLR in the abstract.

2. In abstract and case report- patient age mentioned 84 year and in table 2 -83 year- kindly check and correct

Response: We thank the reviewer for the advice. We have checked the patient age and revised in the manuscript.

3. kindly quote references in table 2 regarding every case.

Response: We thank the reviewer for the advice. We have quoted references in table 2 regarding every case. Including our case, only these three cases were AITL induced HLH confirmed at the time of AITL diagnosis, not during

chemotherapy or relapse. There was only one AITL-induced DIC case reported previously.

4. In discussion- AITL-associated HLH had a poor prognosis due to aggressive disease course, opportunistic infections compared with EBV-associated HLH[18].---- not in line with reference 18, kindly check.

Response: We thank the reviewer for the kind suggestion. We have checked the content was indeed not in line with reference 18. We have revised the reference in the manuscript.

Reviewer #2:
Scientific Quality: Grade C (Good)
Language Quality: Grade B (Minor language polishing)
Conclusion: Accept (General priority)
Specific Comments to Authors: 1 Title. Does the title reflect the main subject/hypothesis of the manuscript? Exactly 2 Abstract. Does the abstract summarize and reflect the work described in the manuscript? Yes 3 Key words. Do the key words reflect the focus of the manuscript? Yes 4 Background. Does the manuscript adequately describe the background, present status and significance of the study? Yes 5 Methods. Does the manuscript describe methods (e.g., experiments, data analysis, surveys, and clinical trials, etc.) in adequate detail? Yes 6 Results. Are the research objectives achieved by the experiments used in this study? What are the contributions that the study has made for research progress in this field? There has a specific demonstration for a very rare case as coexistence between Angioimmunoblastic T-Cell Lymphoma and hemophagocytic lymphohistiocytosis. It will have a value for literature. 7 Discussion. Does the manuscript interpret the findings adequately and appropriately, highlighting the key points concisely, clearly and logically? Are the findings and their applicability/relevance to the literature stated in a clear and definite manner? Is the discussion accurate and does it discuss the paper's scientific significance and/or relevance to clinical practice sufficiently? They made an adequately literature presentation and clearly discussion. 8 Illustrations and tables. Are the figures, diagrams and tables sufficient, good quality and appropriately illustrative of the paper contents? Do figures require labeling with arrows, asterisks etc., better legends? There has an enough presentation. 9 Biostatistics. Does the manuscript meet the requirements of biostatistics? Not need biostatistics. 10 Units. Does the manuscript meet the requirements of use of SI units? Yes 11 References. Does the manuscript cite appropriately

the latest, important and authoritative references in the introduction and discussion sections? Does the author self-cite, omit, incorrectly cite and/or over-cite references? İt is suitable and actual. 12 Quality of manuscript organization and presentation. Is the manuscript well, concisely and coherently organized and presented? Is the style, language and grammar accurate and appropriate? Yes 13 Research methods and reporting. Authors should have prepared their manuscripts according to manuscript type and the appropriate categories, as follows: (1) CARE Checklist (2013) - Case report; (2) CONSORT 2010 Statement - Clinical Trials study, Prospective study, Randomized Controlled trial, Randomized Clinical trial; (3) PRISMA 2009 Checklist - Evidence-Based Medicine, Systematic review, Meta-Analysis; (4) STROBE Statement - Case Control study, Observational study, Retrospective Cohort study; and (5) The ARRIVE Guidelines - Basic study. Did the author prepare the manuscript according to the appropriate research methods and reporting? 1) CARE Checklist (2013) - Case report 14 Ethics statements. For all manuscripts involving human studies and/or animal experiments, author(s) must submit the related formal ethics documents that were reviewed and approved by their local ethical review committee. Did the manuscript meet the requirements of ethics? Yes

Response: we thank the reviewer for the positive comments.

Reviewer #3:
Scientific Quality: Grade E (Do not publish)
Language Quality: Grade D (Rejection)
Conclusion: Rejection
Specific Comments to Authors: This paper is about a single case of AITl associated with DIC and HLH. Unfortunately, the documentation of this case is insuffficient as such. 1. Regarding AITL, the pathological description is not sufficiently detailed and there is no evidence of T cell clonality by TCR rearrangement. 2. The description of HLH in bone marrow is not provided. 3. No evidence that EBV was searched for in the biopsy and the blood. 4. DIC and HLH might be related to a concomitant infection

1. Regarding AITL, the pathological description is not sufficiently detailed and there is no evidence of T cell clonality by TCR rearrangement.

Response: We thank the reviewer for the kind suggestion. We have detected

TCR-Vb repertoire by flow cytometry and the result indicated neoplastic T-cell expansions. But unfortunately we lost the original data. In additon, CD10 has a high specificity for detecting neoplastic T-cell in AITL and pathological findings were supportive.

2. The description of HLH in bone marrow is not provided.

Response: We thank the reviewer for the kind suggestion. We have described that Bone marrow examination showed hemophagocytosis(Figure 1D).

3. No evidence that EBV was searched for in the biopsy and the blood.

Response: We thank the reviewer for the kind suggestion. EBV DNA was detectable in the blood, but the value of EBV DNA copies was at an extremely low level (131 IU/ml).We have described in table 2.

4. DIC and HLH might be related to a concomitant infection

Response: We thank the reviewer for the kind suggestion. Coagulopathy with hypofibrinogenemia could also occur with HLH. We could not exclude that DIC was part of the HLH process in our present case. We could also not exclude DIC and HLH might be related to a concomitant infection, however EBV infection may involved in the pathogenesis of AITL and AITL may aggravate the infection. Therefore, DIC and HLH might be related to a concomitant infection and AITL.

Round 2
Specific Comments To Authors: I agree with reviewer 3. This paper should not be published as such. Diagnosis of AITL requires demonstration of TCR clonality. I would suggest that the authors go back to biopsy material and perform additional molecular analyses (TCR gamma chain rearrangement, next generation sequencing).

Response: We thank the reviewer for the kind suggestion.
We tried to demonstrate TCR clonality. But unfortunately, tissue DNA from the thick needle puncture of lymph node was not enough for TCR gamma chain rearrangement detection when we went back to our biopsy material.
We performed literature of review and found that: [1] Polymerase chain reaction(PCR) techniques for the amplification of TCR revealed clonal T-cell receptor (TCR)gene

rearrangements in 70% of the AILD cases investigated. In the remaining 30% of cases, no dominant gene rearrangements were found. Minor clonal T-cell populations in those cases in which no clone was found by whole-tissue DNA analysis were not detectable even at single cell resolution. **(Reference 1-3)**. [2] In addition, CD10 is a phenotypic marker that specifically identifies the tumor cells in 90% of AITL, including the early cases. No CD10+T cells were present in the cases of peripheral T-cell lymphoma unspecified or reactive hyperplasia. **(Reference 4).** Furthermore, the diagnosis was confirmed by histopathology. So, we thought that there was a clear indication of AITL.

We deeply appreciate your consideration of our manuscript.

Reference

1. Lorenzen J, Li G, Zhao-Ho ̈hn M, et al. Angioimmunoblastic lymphadenopathy type of T-cell lymphoma and angioimmunoblastic lymphadenopathy: a clinicopathological and molecular biological study of 13 Chinese patients using polymerase chain reaction and paraffin-embedded tissues. Virchows Arch 1994,424:593–600;

2. Smith JL, Hodges E, Quin CT, et al. Frequent T and B cell oligoclones in histologically and immunophenotypically characterized angioimmunoblastic lymphadenopathy. Am J Pathol 2000, 156:661–669;

3. Klaus Willenbrock, Axel Roers, Christian Seidl, et al. Analysis of T-Cell Subpopulations in T-Cell Non-Hodgkin’s Lymphoma of Angioimmunoblastic Lymphadenopathy with Dysproteinemia Type by Single Target Gene Amplification of T Cell Receptor-βGene Rearrangements.American Journal of Pathology 2001，158：1851-1857.

4. Ayoma Attygalle, Rajai Al-Jehani, Tim C. Diss, et al. Neoplastic T cells in angioimmunoblastic T-cell lymphoma express CD10.BLOOD 2002,99:627-633