



HEMATOLOGY, TRANSFUSION AND CELL THERAPY

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Letter to the Editor

Regarding “Challenges in diagnosing thrombotic thrombocytopenic purpura”

1 Dear Editor,

2 We appreciate the thoughtful critique of our recent case
3 report reported as Images in Clinical Hematology [1] and
4 would like to clarify points raised regarding the diagnostic cri-
5 teria for immune thrombotic thrombocytopenic purpura
6 (iTTP).

7 In their response, Jacobs et al. [2] express concern that our
8 case lacked sufficient information to confirm a diagnosis of
9 iTTP, specifically citing the absence of reported ADAMTS13
10 autoantibody data and genetic testing. We agree with the
11 importance of distinguishing immune-mediated from con-
12 genital TTP, as this has significant implications for manage-
13 ment and prognosis.

14 The critique reads as follows:

15 “As such, without the identification of an autoantibody, genetic
16 testing should be performed to exclude mutations in
17 the ADAMTS13 gene. Given that the authors did not report an anti-
18 body, nor did they assess for genetic mutations, this case cannot be
19 considered a ‘confirmed’ case of iTTP.”

20 We would like to clarify that an ADAMTS13 inhibitor
21 screen was indeed performed in our patient and yielded a
22 positive result. The inhibitor titer, measured using the
23 Bethesda assay, was 2.2 (reference <0.4), indicating the
24 presence of a circulating autoantibody against ADAMTS13.
25 This supports the diagnosis of acquired, immune-medi-
26 ated TTP. Due to word count limitations and the case
27 vignette format, this detail was not included in the origi-
28 nal publication.

29 We agree that in the absence of detectable autoantibodies,
30 the possibility of congenital TTP should be considered, and
31 genetic testing may be warranted. However, in our case, the
32 presence of a quantifiable inhibitor supports an immune-
33 mediated process, and the clinical picture (including age of
34 onset and concurrent autoimmune disease) makes congenital
35 TTP unlikely.

36 We appreciate the ongoing discussion regarding the
37 importance of diagnostic precision in TTP. We fully agree that
38 thorough documentation of relevant laboratory findings is

critical, not only for accurate diagnosis, but also for manage- 39
ment and epidemiological purposes. 40

Conflicts of interest

No conflicts of interest to declare.

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