HEMATOLOGICAL MANIFESTATIONS IN PATIENTS WITH ANTIPHOSPHOLIPID ANTIBODIES

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BACKGROUND

Selected hematological manifestations, expressed as “extracriteria”, are related to persistent antiphospholipid antibodies (aPL), irrespective of the presence of criteria manifestations of antiphospholipid syndrome (APS), and may lead to increased morbidity and worse prognosis. The aim of this study was to assess the prevalence of hematological manifestations in primary aPL-positive patients.

METHODS

In this cross-sectional study, we included patients persistently positive to aPL, without other autoimmune diseases (especially systemic lupus erythematosus). We assessed the following hematological manifestations: (1) autoimmune hemolytic anemia (AIHA), (2) leukopenia/lymphopenia ($< 4,000/\text{mm}^3$ and $< 1,000/\text{mm}^3$, respectively) and (3) thrombocytopenia ($< 100,000/\text{mm}^3$), confirmed 12 weeks apart in the absence of other causes. Findings were correlated with clinical manifestations and aPL profile. The adjusted global antiphospholipid syndrome score (aGAPSS) was calculated as published elsewhere. High aGAPSS was defined as score above the median value of our cohort. Statistical analysis was performed using Pearson chi-squared test, two-tailed Fisher’s exact test and Mann–Whitney U-test, as appropriate. The significance threshold was set at 5%.

RESULTS

We enrolled 148 patients, of which 143 fulfilled PAPS classification criteria; the other 5 presented with aPL-related thrombocytopenia. The majority was female (85.1%), the median age at the study was 46 years (39–56) and the median time of disease was 14 years (9–20). Approximately 90% and 40% of PAPS patients had thrombotic and obstetric events, respectively. One patient had AIHA (with negative ANA), 5 had leukopenia/lymphopenia and 14 had thrombocytopenia. Correlations between thrombocytopenia and aPL profile/aGAPSS are presented in Table 1.

CONCLUSION

The presence of autoimmune hemolytic anemia and leukopenia/lymphopenia associated with aPL were rare. On the other hand, thrombocytopenia was present on approximately 10% of patients and was associated with a higher risk profile (triple positivity and higher aGAPSS).

KEYWORDS

Thrombocytopenia, Anemia, Antiphospholipid, Hematological.

Table 1. Differences between thrombocytopenia and non-thrombocytopenia in aPL- related patients.

	Nonthrombocytopenic n = 134	Thrombocytopenic n = 14	p-value
Isolated LA, n (%)	61 (45.5)	3 (21.4)	0.083
Isolated aCL, n (%)	2 (1.5)	0 (0)	1.000
Double positivity, n (%)	42 (32.3)	4 (28.6)	1.000
Triple positivity, n (%)	27 (20.1)	6 (42.9)	< 0.001
aGAPSS $\geq$ 9, n (%)	58 (43.3)	11 (78.6)	0.012
aGAPSS, n (IQR 25–75)	8 (5–12)	12 (8.5–13)	0.050

aCL: anticardiolipin antibody; aGAPSS: adjusted global anti-phospholipid syndrome score; LA: lupus anticoagulant; IQ: interquartile range. Isolated anti- beta 2 glycoprotein-1 antibody was not present.