Acquired Platelet Dysfunction with Eosinophilia

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Key words: thrombocytes, thrombocytopathy, cell morphology

ISSN 2507-8364 (Online)
Printed in the Philippines.

Received: 30 April 2020.
Accepted: 19 June 2020.
Published online first: 22 June 2020.
https://doi.org/10.21141/PJP.2020.07

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Table 1. Laboratory work-up done during initial consult and follow-up after two months

Parameter	Reference Range	Units	Initial Consult	Follow-up
Complete Blood Count				
RBC Count	4.00 - 5.20	$x10^6$/L	4.54	4.78
Hemoglobin	11.5 - 15.5	g/dL	12.0	12.4
Hematocrit	35 - 45	%	34.9	36.6
MCV	75 - 87	fL	76.9	76.6
MCH	25 - 33	pg	24.6	25.9
MCHC	32 - 36	%	34.4	33.9
RDW	11.5 - 15.0	%	13.8	12.8
Platelet Count	150 - 450	$x10^9$/L	179	338
PDW	9.5 - 16.0	%	13.3	9.0
MPV	6.5 - 10.0	fL	10.2	8.2
WBC Count	5.0 - 14.5	$x10^9$/L	8.46	8.46
Neutrophil	32 - 54	%	41.0	55.0
Lymphocyte	28 - 48	%	16.5	28.0
Monocyte	3 - 6	%	4.5	10.0
Eosinophil	0 - 3	%	36.0	6.0
Basophil	0 - 1	%	1.5	1.0
Absolute Eosinophil Count	0 - 0.6	$x10^9$/L	3.05	0.33
Coagulation Tests				
PT	8.7 - 11.5	sec	10.2	not done
APTT	31.8 - 43.7	sec	35.6	not done
Platelet Function Tests				
Bleeding Time (Ivy Method)	2 - 8	min	6.5	not done
Platelet Closure Time (PFA-100)				
Col/Epi	82 - 150	sec	>287	104
Col/ADP	62 - 100	sec	115	70

INTRODUCTION

A six-year-old male was brought in with 1-month history of recurrent spontaneous bruising which resolves without intervention. There was no history of trauma, other bleeding episodes, medication intake, nor recent viral infection. Birth, past medical, and family histories were unremarkable. Pertinent physical examination showed multiple, non-tender ecchymosis of varying chronicity and sizes on his upper and lower extremities and abdomen. The rest of the examination was essentially normal.

Initial laboratory work-up with complete blood count, prothrombin time, activated partial thromboplastin time, and bleeding time (Ivy Method) was done (Table 1). Peripheral blood smear examination showed eosinophilia and adequate qualitative platelet count. However, the platelet morphology shows numerous agranular and hypogranular grey platelets (Figure 1). Based on the history, physical examination, and initial work-up, acquired platelet dysfunction with eosinophilia (APDE) was suspected. Additional work-up with platelet closure time using Platelet Function Analyzer-100 (PFA-100) was done (Table 1) which provides evidence of platelet dysfunction. No intestinal parasites were seen in three consecutive fecalysis done. Nonetheless, the patient was started on mebendazole. Subsequent follow-up after two months showed resolution of ecchymosis, eosinophilia, and platelet dysfunction (Table 1), with no noted recurrence.

APDE is a syndrome with transient state of platelet dysfunction mostly reported from children in Southeast Asia.[1] APDE requires the following features for diagnosis: clinical manifestation of spontaneous ecchymosis on the trunk or extremities; hemogram showing eosinophilia; and evidence of platelet dysfunction.[2] The spontaneous ecchymosis seen clinically is indistinguishable from that of idiopathic thrombocytopenic purpura (ITP). APDE is differentiated from ITP by the absence of thrombocytopenia. The eosinophilia observed may be an epiphenomenon seen in areas with endemic parasitism, since intestinal parasitism is the evident cause of eosinophilia.[3,4] The platelet dysfunction is demonstrated by light transmission aggregometry, with results consistent with a platelet storage pool disorder.[3,5] Alternatively, PFA may be used especially in pediatric patients since it requires less blood.[2]

However, platelet aggregation studies are offered only in specialized laboratories and it may take a few days before results are released. A presumed diagnosis can be

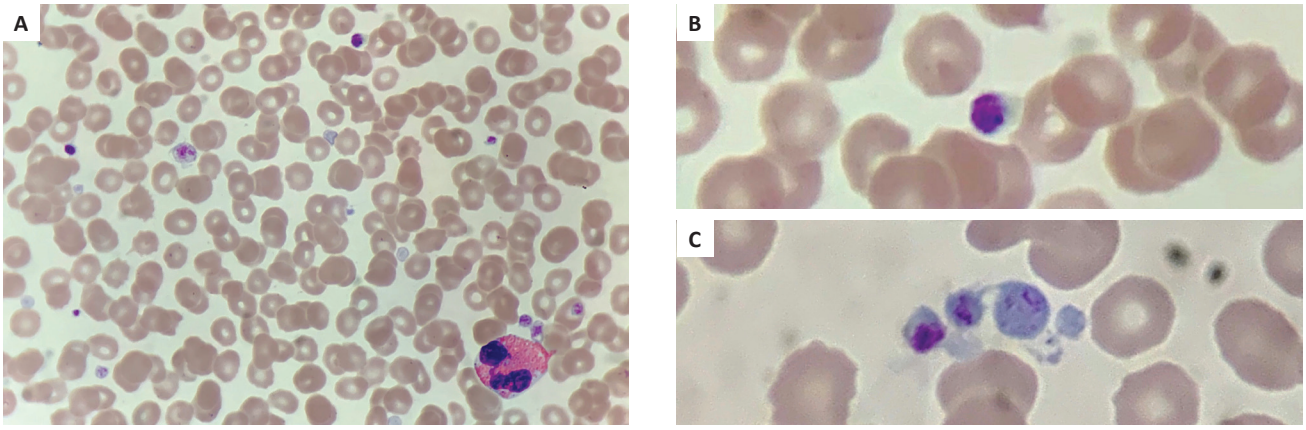

Figure 1. Peripheral blood smear photomicrograph showing an oil immersion field with an eosinophil, agranular grey platelets, hypogranular grey platelets, and a platelet with normal morphology **(A)** 1000X magnification; normal platelet **(B)** and three side-by-side platelets with decreasing granularity and a fourth agranular platelet **(C)**, 3000X magnification.

made rapidly in any clinical laboratory offering routine hematology services, by microscopic examination of the platelet morphology.[3] Wright stained blood smears show gray, pale staining platelets with smooth, round cell membrane contour. The cytoplasm may show fewer granules to none at all. Platelets with abnormal morphology may comprise about 30-80% of the platelets examined. This proportion correlates with the severity of the bleeding.[6]

The pathogenesis is still unknown. The platelet dysfunction has been thought to be due to the high production of IgE antibodies from a type I hypersensitivity reaction. The binding of immune complexes to platelets leads to in-vivo platelet activation. This activation promotes the release of adenine nucleotides, resulting in acquired storage pool deficiency.[5-7]

The prognosis is generally good, with a benign clinical course, and resolves spontaneously within 3 to 6 months to about a year.[1,5]

STATEMENT OF AUTHORSHIP

All authors certified fulfillment of ICMJE authorship criteria.

AUTHOR DISCLOSURE

The authors declared no conflict of interest.

FUNDING SOURCE

None.

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