Acquired Platelet Dysfunction with Eosinophilia or Idiopathic Purpura with Grey Platelets?

Dear Editor,

An acquired, transient bleeding disorder characterised by a thrombocytopathic bleeding with a platelet storage pool disorder has been reported from the Southeast Asian region and elsewhere since the 1960s.[1,2] The condition has been coined acquired platelet dysfunction with eosinophilia (APDE) as cases reported earlier were also found to have eosinophilia. However, a large series of cases published in the 21st century shows that 14% of the affected children do not have eosinophilia.[3] The following cases also illustrate that significant eosinophilia is not a constant feature (Table 1). Idiopathic purpura with grey platelets is proposed as a better terminology to describe the condition, so long as the aetiology remains obscure.

Case 1 is a Eurasian 5-year-old male who presented with abrupt onset of generalised ecchymosis and scattered petechiae. Non-accidental injury was initially suspected and hence, he was brought to medical attention. Multiple bruises and petechiae were found in the extremities and a couple of ecchymosis were seen on the trunk. The child was otherwise well and no other abnormal signs were seen. The complete blood count was normal with mild eosinophilia (0.75×10^9/L). The peripheral blood film revealed plenty of grey platelets and the results of platelet aggregation were consistent with a platelet storage pool disorder. His bleeding symptoms resolved without treatment after 2 months.

Case 2 is a 4-year-old Chinese female who presented with recent onset of generalised bruises and petechiae. The complete blood count was marked by extreme eosinophilia (4.81×10^9/L). On the peripheral blood film, grey platelets with eosinophils were abundant (Fig. 1A). Platelet aggregation abnormalities were consistent with a storage pool defect. Her bleeding symptoms resolved a month later. A repeat test 8 months later while she was evaluated for skin allergy showed platelet and eosinophil counts of 377 and 1.09×10^9/L, respectively, with disappearance of the grey platelet on the blood film.

Case 3 is a 3-year-old female of Indian descent who presented with an abrupt onset of generalised bruises mainly on the lower limb and abdominal wall. The complete blood count was normal and no eosinophilia was found. However, grey platelets were seen under the microscope (Fig. 1B)

Table 1. Summary of Haematologic Findings in the 3 Cases

	Case 1	Case 2	Case 3	Normal Ranges
Sex/age, year	Male/5.3	Female/4.7	Female/3.7	-
Hb, g/dL	13.0	13.0	11.1	-
WBC, $\times 10^9$/L	6.80	13.35	9.74	-
Eosinophil, $\times 10^9$/L	0.75	4.81	0.29	-
Platelet, $\times 10^9$/L	153	269	173	-
Platelet aggregation tests with:				
ADP	44%	61%	50%	64% to 111%
Collagen	13%	9%	25%	68% to 117%
Epinephrine	12%	25%	23%	46% to 122%
Ristocetin	100%	82%	87%	80% to 115%
Arachidonic	82%	74%	78%	52% to 110%

ADP: Adenosine diphosphate; Hb: Haemoglobin; WBC: White blood cells

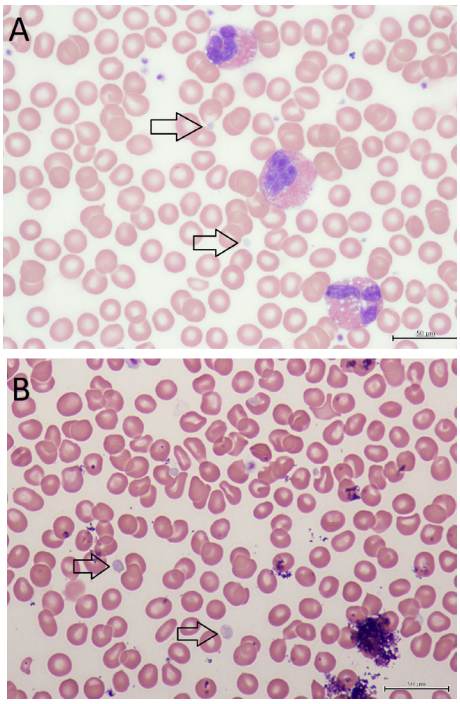

Fig. 1. Photomicrographs from Cases 2 and 3, A) and B) respectively, showing at least 50% of the platelets on the peripheral blood film are devoid of normal contents and thus appear as grey platelets. Eosinophilia is obvious in Case 2 but absent in Case 3.

and a platelet storage pool disorder was evident on platelet aggregation tests. She recovered without treatment a month later with the disappearance of the grey platelets on blood film.

Idiopathic thrombocytopaenic purpura (ITP) is the most common cause of acquired bleeding disorder in childhood. The name clearly defines the cardinal clinical and laboratory features and the diagnosis is usually straightforward. APDE, in contrast, does not fulfill the same purpose. Platelet dysfunction has to be defined by specific platelet aggregation tests, which is only available in specialised laboratories and the results may take a few days to turn around. While extreme eosinophilia was common in earlier reports like Case 2, a recent series of 168 children reported from Thailand shows a wide range of eosinophil counts and 14% do not have eosinophilia at all.[2] In this respect, eosinophilia may be an epiphenomenon seen in communities with endemic parasitic infections and allergic disorders. Thus, APDE is an inconvenient and inaccurate name to describe the condition and may distract the unfamiliar clinician from making the diagnosis. Indeed, as illustrated by Case 2, eosinophilia may persist even when the thrombocytopathic bleeding diathesis has resolved.

Hence, idiopathic purpura with grey platelets may be a better terminology. Cutaneous bruises with or without petechiae, clinically indistinguishable from ITP, are universally present. The condition can be rapidly recognised when grey platelets are found under the microscope in the absence of thrombocytopaenia. Thus, a presumed diagnosis can be made in any laboratory providing routine haematology service. Additional tests on platelet dysfunction to document a storage pool disorder will lend support to the diagnosis.

REFERENCES

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