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A Case of Acquired Platelet Dysfunction with Eosinophilia in Our Hospital "Omer Nishani", Gjirokaster

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Abstract: Acquired platelet dysfunction with eosinophilia (APDE) first reported in 1975 is an acquired, transient bleeding disorder of unknown etiology characterized by spontaneous ecchymosis, eosinophilia and platelet dysfunction. This bleeding disorder, otherwise known as "non-thrombocytopenic purpura with eosinophilia", occurs commonly in indigenous Southeast Asian children. Cases of APDE occur rarely outside this endemic area. This syndrome runs a benign course and patients can recuperate spontaneously from the bleeding episodes within 6 months to a year. The aim of our case report is to highlight the importance of recognizing this little known bleeding disorder in our daily clinical practice. We reported the first documented case of a 7 year old girl managed in our hospital "Omer Nishani", with clinical features characteristic of APDE. The patient was in good health, presented with spontaneous bruising on the extremities with no organomegaly. Marked eosinophilia and prolonged bleeding time with normal platelet count were noted on laboratory work-up. The patient was started on antiparasitic therapy despite negative stool results, and subsequent follow-up after 3 weeks showed resolution of ecchymosis and eosinophilia. Awareness of this benign bleeding disorder, which clinically mimics Idiopathic Thrombocytopenic Purpura (ITP) would help to diagnose and treat early, and lessen parental anxiety.

Keywords: APDE, Eosinophilia, Ecchymosis

1. Introduction

Acquired platelet dysfunction with eosinophilia (APDE) is a transient state of bleeding diathesis characterized by spontaneous bruising in the presence of marked eosinophilia in a healthy person [1]. The diagnosis of APDE requires three components: (1) clinical presence of reversible spontaneous ecchymosis on the extremities or trunk, (2) CBC showing eosinophilia, and (3) evidence of platelet dysfunction [2].

The spontaneous ecchymosis seen clinically is indistinguishable from that of idiopathic thrombocytopenic purpura (ITP). Hypereosinophilia is often the first clue to diagnosis. This bleeding disorder, otherwise known as "non-thrombocytopenic purpura with eosinophilia", occurs commonly in children from South-East Asia or have a history of traveling to this region [2]. Cases of APDE are rare outside this endemic area.

In this paper, we report a case of APDE on our pediatric ward, with no evidence of parasitic infection, neither had ever traveled to an endemic area.

2. Case Report

A seven year-old girl was brought in with 2 weeks history of spontaneous bruising on her bilateral lower limbs. There was no history of trauma, other bleeding episodes, gastrointestinal manifestations, medication intake, nor recent viral infection. There was no remarkable past history of bleeding tendency. Family history of bleeding disorder or atopy was negative. Apart from that the child was healthy and continued to go to school as usual. Physical examination showed multiple, non-tender ecchymotic patches of varying sizes over her bilateral lower limbs (Figure 1). No hepatosplenomegaly or lymphadenopathy was noted. The rest of the systemic examination was normal.

Initial laboratory investigations with complete blood count, prothrombin time, activated partial thromboplastin time, and bleeding time was done (Table 1). Full blood count showed

the following values: haemoglobin (Hb) 11.9 g/dL, platelet count (PLT) 327 x 10^3/uL and white blood cell count (WBC) 9.4 x 10^3/uL with 27.3% eosinophil (absolute eosinophil count 2.6 x 10^3/uL, normal <0.3 x 10^3/uL). Peripheral blood smear examination showed eosinophilia and adequate qualitative platelet count. The clotting profile (PT and aPTT) was normal. Prolonged bleeding time of 08 min 30 s (by Ivy's method) was noted.

Based on the history, physical examination, and initial investigations, acquired platelet dysfunction with eosinophilia (APDE) was suspected. We did not perform any bone marrow examination. Unfortunately we did not assess the patients' platelet function using a platelet function analyzer, because those tests are offered only in specialized laboratories. Stool examination did not show any ova or parasite. Nonetheless, the patient was started on mebendazole 100 mg x 1 for 3 days and asked to come for review. Subsequent follow-up after 3 weeks showed resolution of ecchymosis and eosinophilia (AEC - 100/uL.), with no noted recurrence (Table 1).

Table 1. *Laboratory investigations done during initial consult and follow-up after 3 weeks.*

Parameter	Reference Range	Units	Initial Consult	Follow - up
Complete Blood Count				
RBC count	4.00 - 5.30	x10^6/uL	4.40	4.51
Hemoglobin	11.0 – 13.0	g/dL	11.9	12.4
Hematocrit	33 – 43	%	36.3	37.5
MCV	76 - 88	fL	82.5	83.1
MCH	25 - 31	pg	27.1	27.5
MCHC	32 – 36	g/dL	32.8	33.0
RDW	10.8 – 14.0	%	11.8	11.8
Platelet Count	150 – 400	K/uL	327	324
PDW	8.5 – 25	fL	10.5	9.6
MPV	7.2 - 11	fL	8.7	8.4
WBC Count	4 - 11	K/uL	9.4	13.1*
Neutrophil	32 – 59	%	28.9	76.7
Lymphocyte	27 – 57	%	36.4	16.1
Monocyte	3 – 9	%	6.5	6.10
Eosinophil	0 – 3	%	27.3	0.8
Basophil	0 – 1	%	0.9	0.3
Absolute Eosinoph. Count	0 – 0.3	K/uL	2.6	0.1
Coagulation Tests				
PT	70 – 110	%	73	not done
aPTT	< 43	sec	29.6	not done
Platelet Function Tests				
Bleeding Time	2 – 8	min	8.5	not done

*the patient had a viral infection during the follow-up

3. Discussion

APDE was first recognized by Suvatte et al. in 1974 and Mitrakul (1975) as "transient, spontaneous bruising with long bleeding times and normal platelet counts" [3]. It is a unique disease that has been mainly reported in the region of Malaysia, Thailand and Singapore [4]. This syndrome mainly affects the pediatric age group, but few adult cases have also been reported [5]. The mean age of onset was about 6.5 years [6]. The majority of cases have a parasitic infection [1]. However, the geographic occurrence is still a mystery [7]. In around 56% of patients with APDE, parasites can be detected through stool examinations [1, 8]. Empirical therapy with anthelmintics is still been used [9]. More than half of the patients with APDE experience prolonged bleeding times [1, 3]. Reduced or absent platelet aggregation to collagen, epinephrine and ADP are found in the majority of patients with APDE [10]. Wright stained blood smears may show gray, pale staining platelets with smooth, round cell membrane contour.

The physiopathology of APDE is not obviously understood. Previous studies have suggested that parasites induce eosinophils, leading to the high production of IgE antibodies from a type I hypersensitivity reaction and the formation of immune complexes, which can bind to platelets, resulting in abnormal secondary aggregation [1, 11]. Nevertheless, parasites are not the only etiological reason in APDE. It could also be found in patients with eosinophilic inflammation, such as asthma or hay fever [12] Therefore, both allergic disorders and asthma should be suspected in patients with APDE who do not live in or have no travel history to Southeast Asia.

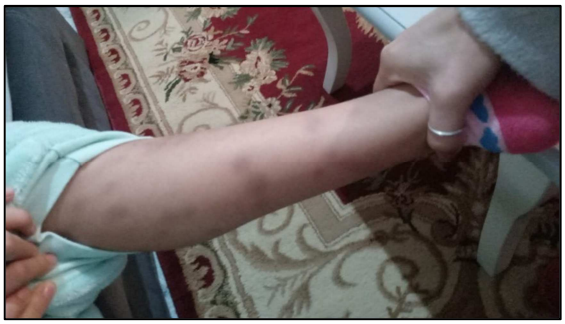

Figure 1. *Multiple spontaneous bluish colored patches on lower limbs since 2 weeks (photo taken at home).*

APDE is a disease without confirmative diagnostic signs or test. We need to exclude other conditions before making the diagnosis. The presence of thrombocytopenia does not rule out the diagnosis of APDE [13]. Esosinophilia can be found in disorders like parasitic infections, drug reactions, allergic disorders etc. This syndrome runs a benign course and patients can recuperate from the bleeding episodes within 6 months to 1 year [14]. Recurrence is apparently rare once resolved [15].

4. Conclusion

In conclusion, it is important to recognize this benign bleeding disorder (APDE) which clinically mimics Idiopathic Thrombocytopenic Purpura (ITP). All that is required is to reassure the parents and follow up closely.

Conflict of Interest

The authors have no conflict of interest and the patient's privacy is preserved.

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