ACQUIRED PLATELET DYSFUNCTION WITH EOSINOPHILIA (APDE): AN UNDERRECOGNISED CONDITION

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SUMMARY

We present three cases of Acquired Platelet Dysfunction with Eosinophilia (APDE). The importance of recognising this benign condition, which usually does not require any specific therapy, is stressed to avoid the pitfalls of diagnosing more serious bleeding disorders in children presenting with ecchymosis.

INTRODUCTION

Acquired Platelet Dysfunction with Eosinophilia (APDE) is a syndrome where there is a transient state of platelet dysfunction in the presence of marked eosinophilia. This bleeding disorder, otherwise known as "non-thrombocytopenic purpura with eosinophilia", occurs commonly in children in South-East Asia.

We present three cases of this syndrome which came under our care recently.

CASE HISTORY

Case 1

An eight-year-old female was referred to the hospital by her general practitioner with the provisional diagnosis of Idiopathic Thrombocytopenic Purpura (ITP). She was well till about a week prior to admission when she developed spontaneous painless bruising all over the body. On the day of admission, she also complained of epistaxis. There was no history of ingestion of drugs, trauma, fever or respiratory symptoms. She did not complain of bleeding gums or change in the colour of urine or stool.

She was the youngest in a family of seven. Her siblings were well. There was no family history of bleeding disorders. On examination, she had multiple ecchymotic lesions of varying size and shape on her left forearm, gluteal regions, left leg and dorsum of both wrists. The rest of the physical examination was normal: in particular, she had no lymphadenopathy, hepatosplenomegaly or retinal haemorrhage.

A provisional diagnosis of ITP was made and the patient was then investigated.

The blood counts and coagulation studies done on this patient are presented in Tables I and II respectively.

The peripheral blood film showed platelets in abundance with normal morphology. There was also marked eosinophilia with a number of target cells on the film. The bone marrow was essentially normal, except for an increase in eosinophils. The megakaryocytes were normal.

The HbF was normal at 0.6% by alkaline denaturation method. Hb electrophoresis showed a normal level of HbA2 at 2.2% of the total. She was treated with a course of Mebendazole and was discharged home well. During follow-up, she was found to be doing well and the ecchymosis completely disappeared over a period of three months.
Cases 2 and 3

A ten-year-old Chinese male presented with bruises off and on for three months. The only medication taken during this period was paracetamol for fever and cold, and he was never on aspirin or related compounds. On examination, there was no hepatosplenomegaly. An enlarged submandibular lymph node was palpable. He had a few small bruises on the thigh and lower limbs.

The family history was interesting. His elder brother, 11 years-old, also had bruising for three months. He had not been on any medication. On examination, he too had a submandibular lymph gland with no hepatosplenomegaly. He had two bruises on his left forearm and one on his right upper arm.

Apparently, a month ago, both brothers were admitted to another hospital for two weeks, without any definitive diagnosis being made there. The hematological work-up carried out on the two brothers are shown in Tables I and II. The results of platelet aggregation studies are summarised in Table III.

Both brothers were found to have hookworm ova in their stools and were treated with antihelmintics and did well.

DISCUSSION

The syndrome of APDE was first recognised by Suvatte et. al., in 1974. The exact cause of this condition is not known. It is characterised by a transient defect in platelet function in the presence of marked eosinophilia.

Epidemiologically, this syndrome appears to be confined to children in South-East Asia. However, there has been a case report of a child who developed the condition two weeks after her return to the United Kingdom from Malaysia.

The hallmark of this disorder includes recurrent spontaneous bruising, normal hematocrit, normal number of platelets, normal coagulogram, eosinophilia, prolonged bleeding time, abnormal platelet adhesiveness, and associated parasitic infection.
### TABLE III
**SUMMARY OF PLATELET AGGREGATION STUDIES**

<table>
<thead>
<tr>
<th>Platelet Aggregation</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>A.D.P.</td>
<td>Fine aggregation after 4 seconds</td>
<td>no aggregation</td>
<td>abnormal</td>
</tr>
<tr>
<td>Collagen</td>
<td>Irreversible aggregation</td>
<td>irreversible aggregation</td>
<td>normal</td>
</tr>
<tr>
<td>Ristocetin</td>
<td>Irreversible aggregation</td>
<td>Irreversible aggregation</td>
<td>normal</td>
</tr>
</tbody>
</table>

*The platelet aggregation tests were done manually. In Case 2, there was fine aggregation with ADP after 4 seconds (normal control showed immediate aggregation with ADP). In Case 3, there was no aggregation at all in response to ADP.*

Apart from the ecchymosis, these children are usually well as illustrated by our patients. However Suvette et. al., reported epistaxis and gum bleeding in 48.4% and 12.9% of their patients respectively.

The bleeding time was prolonged in all three patients. However, the full coagulation profile and platelet aggregation tests were undertaken only in Cases 2 and 3. In both these cases, platelet aggregation with ADP was abnormal. Case 2 had a prolonged lag phase and showed fine aggregation. Case 3 had no aggregation with ADP.

Platelet aggregation with collagen and ristocetin was normal. This contrasts with the Thai experience in which platelet aggregation to ADP, thrombin and collagen were found to be abnormal in 67.7%, 79.6%, and 87.1% of cases respectively; these tests returning to normal within six months to a year.

The hypochromic picture in Case 1 was thought to be due to Alpha thalassaemic trait. This appears to be a coincidental finding. The percentage of eosinophils in our cases varied from 11% to 19%. In the Thai series, this ranged from 3% to 69% with a mean of 26.17%.

The brothers’ stools were positive for hookworm ova, although in our first patient, no ova or cyst could be identified in the stool. In the Thai series, 58% of the stools were positive for various parasites, particularly *Ascaris, Enterobius* and *Ankylostoma*.1–3

It is essential to recognise this benign condition which clinically appears to be a good mimicker of Idiopathic Thrombocytopenic Purpura. This was proven in our first patient. Our last two cases illustrate the necessity to screen the family members for the same problem.

The syndrome of APDE runs a benign course and resolves spontaneously within six months to a year. No specific treatment has been found to be useful except that perhaps there is a place for a course of antihelminthics. It must be stressed in particular that blood component therapy has not been found useful in the treatment of these cases. All that is required is to reassure the patient and their parents.

While the incidence of this disorder in Malaysian children is not known, it is considered to be the most common cause of purpura in Thai children. It is possible that our experience may prove to be similar to that of our neighbours.

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### REFERENCES

