Acquired Platelet Dysfunction with Eosinophilia

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Acquired platelet dysfunction with eosinophilia or nonthrombocytopenic purpura with eosinophilia is an acquired bleeding disorder of unknown etiology associated with platelet dysfunction and eosinophilia. It was first described by Mitrakul[1] and Suvatte in 1975[2]. In 1977 Isarangkura[3] named this disease "acquired platelet dysfunction with eosinophilia (APDE)" and described the abnormal platelet morphology found in this disease. A few papers, mostly from tropical countries,[4-15] have reported this bleeding entity.

Clinical manifestations.
It is one of the common causes of purpura in children in Thailand[2]. It occurs mostly in children age 1-12 years and may be seen in adolescents and adults. The most common age group is 5-8 years It affects both sexes. The leading symptom is spontaneous bruising on the extremities off and on for a duration of weeks or months. The purpura is shown as purpuric spots or medium size ecchymoses. Some patients may have mucosal bleeding, i.e. epistaxis, gum bleeding. Spontaneous intracranial hemorrhage has not been reported[1-16]. With the exception of bleeding, the patients are in good health with no history of previous bleeding tendency (in the patients and their families), and the patient has no history of recent drug intake. It should be noted that the bleeding symptoms in most patient are mild, transient with spontaneous recovery, i.e. much less severe than that seen in idiopathic thrombocytopenia purpura. None of them require hospitalization or bleed spontaneously to death except due to severe trauma or accidents.

Laboratory findings.

1. Eosinophilia is found in most cases (83%)[16] and persists only few weeks after the onset. The eosinophil count varies from 594-28,566/µL or 3-69% of total WBC[2].
2. Mild leukocytosis was found in 80% of the cases with the range of 5,600-35,000/µL[16]. Eosinophilia and mild leukocytosis are observed transiently during the early onset which may be the response to some immune-mediated response. If the patient presents a few weeks after onset, eosinophilia usually is not seen.
3. The platelet count is normal. Only few cases (3%)[2] show mild transient thrombocytopenia.
4. Platelet morphology is one of the pathognomonic signs to make the diagnosis of this disease using simple tests. Wright's stained blood smears shows pale stained platelets, fewer granules in the cytoplasm, good cell membrane appearance and reduced or no clumping of platelets. The abnormal morphology of platelets was found in 30-80% of total platelets in blood smears[3]. The amount of abnormal morphology correlates with the severity of clinical bleeding[16]. When the bleeding symptoms improve or recover, platelet abnormalities decrease or become normal.
5. Bleeding time is prolonged in about 60% of patients[2,16].
6. Clot retraction is normal in all cases[2,16].
7. Platelet adhesiveness is abnormally low in 60% of patients[2]
8. Platelet factor 3 release is abnormal in 50% of patients[2].
9. Platelet aggregation in response to stimulation by ADP, thrombin, and collagen is decreased, but the response to ristocetin is normal [2,16].
10. Immunoglobulins are in the normal range including IgG, IgA, IgM. However, IgE is increased.
11. Stool examination shows common parasites, e.g. ascaris, hookworm, enterobius, etc., in 50-60% of cases[2].

Etiology and pathogenesis.
The pathogenesis is still unknown. Platelet function tests show variable storage pool defects[12]. It has been speculated that the high IgE is in response to parasite causes mediated mast cell degranulation and leads to in-vivo platelet activation. Stool helminths are observed in about 50-60%[2]. Stool parasites may be involved in the pathogenesis[2,12]. This should be further explored.

Management.
Since the etiology is unknown, the management is symptomatic and supportive:

1. Patients should be educated to avoid trauma and injury, and an identification card should be given to all patients. Emphasize the need for patients to inform physicians when undergoing surgery that an infusion of platelet concentrates is necessary.
2. Transfusion of platelet concentrate 0.2-0.4 unit/kg/dose every 2-3 days until the surgical wound is healed[3]. Fresh frozen plasma or cryoprecipitate does not correct the hemostatic defect[5,18].
3. Common intestinal parasites are usually removed by giving antihelminthic drugs.
4. Reassure the parent about the prognosis and alert them to accidents.

Course and prognosis.
The course usually lasts for 2-6 months but sometimes for 2-3 years. Bleeding symptoms may recur.

The prognosis is good unless severe trauma or accidents particularly head injury with intracranial hemorrhage

is associated. Mortality has not been observed in over 150 reported cases[1-18].

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