Clinical audit on the management of acquired platelet disorders in Assiut University Children Hospital

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Introduction

Platelet disorders lead to defects in primary hemostasis and produce signs and symptoms different from coagulation factor deficiencies (disorders of secondary hemostasis). Primary hemostatic disorders are characterized by prolonged bleeding time and the characteristic physical examination in the form of mucocutaneous bleeding. Initial laboratory evaluations for patients with acquired platelet disorders include complete blood count and peripheral blood smear. Treatment of patients with suspected acquired platelet disorder is generally specific to the underlying disorder.

Patients and methods

Data of children older than 1 month of age admitted with bleeding tendency caused by acquired platelet disorders during 1 year were collected and analyzed, and clinical management was compared with the standard management guidelines according to the American Society of Hematology 2011.

Results

In all patients, complete blood count was performed, whereas peripheral blood smear was performed only for 46 (60.5%) cases. Bone marrow examination was performed in 30 (39.5%) cases and it was indicated in 23 (77%) of these cases, whereas in seven (23%) cases it was done for purpose of research with guardian consent. Medical treatment (in the form of intravenous immunoglobulin and/or corticosteroids) was provided to seven (9%) cases in whom treatment was not indicated. Intravenous immunoglobulin was administered to 22 (29%) cases, and there was a delay in its administration to 10 (45%) of these cases because of unavailability. Corticosteroids were administered to 26 (34%) cases; eight (31%) of these cases received it for a longer time than indicated. Anti-D immunoglobulin was not administered to any patient.

Keywords:

aplastic anemia, hemolytic uremic syndrome, immune thrombocytopenia, intravenous immunoglobulin, purpura

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Introduction

The platelets arise from the fragmentation of the cytoplasm of megakaryocytes in the bone marrow and circulate in blood as disc-shaped anucleate particles for 7–10 days. The primary physiological role of platelets is to support hemostasis at sites of vascular injury by forming platelet plugs that arrest blood loss [1].

Platelet disorders lead to defects in primary hemostasis and produce signs and symptoms different from coagulation factor deficiencies (disorders of secondary hemostasis). The body's reaction to vessel wall injury is rapid adhesion of platelets to the subendothelium. The initial hemostatic plug, composed primarily of platelets, is stabilized further by a fibrin mesh generated in secondary hemostasis. The arrest of bleeding in a superficial wound, such as the bleeding time wound, almost exclusively results from the primary hemostatic plug. Hence, primary hemostatic disorders are characterized by prolonged bleeding time, and the characteristic physical examination findings are petechiae and purpura [2]. Low platelet count is thrombocytopenia and is because of either decreased production or increased destruction. A disorder of platelet function is thrombocytopathy [3].

The causes of acquired platelet disorders can be classified according to the pathologic mechanism involved in thrombocytopenia and platelet function disorders; each of these have congenital and acquired causes.

Acquired platelet disorders include a group of diseases in which immune thrombocytopenia (ITP) is the most common cause.

Clinical manifestations of platelet disorders include bleeding manifestations and manifestations of the underlying disease.

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Initial laboratory evaluations for patients with acquired platelet disorders include complete blood count (CBC) and peripheral blood smear, whereas further evaluation may include blood urea, serum creatinine, liver function tests, erythrocyte sedimentation rate, C-reactive protein, and/or bone marrow studies for children with lymphadenopathy, splenomegaly, systemic symptoms, or other abnormalities in the CBC or peripheral blood smear [4].

Treatment of patients with suspected acquired platelet disorder is generally specific to the underlying disorder.

In thrombocytopenia, treatment can include corticosteroids, intravenous immunoglobulin (IVIG), intravenous anti-D immunoglobulin, and/or transfusion of platelets [5].

In platelet function disorders, treatment can include desmopressin, antifibrinolytic agents, and/or transfusion of platelets [6].

An accurate diagnosis of the cause of acquired platelet disorder is essential to inform appropriate management.

Assiut University Children Hospital is a tertiary hospital in Upper Egypt where cases with acquired platelet disorders are referred for management.

Self-assessment and evaluation is a policy of the hospital for improvement of quality.

Aim

The aim of this study is to assess the quality of care and management of acquired platelet disorders in Assiut University Children Hospital in 1 year.

Patients and methods

All children older than 1 month of age with bleeding tendency caused by acquired platelet disorders admitted in Assiut University Children Hospital were recorded over 1 year (from 1 May 2015 to 30 April 2016).

Inclusion criteria

Children older than 1 month of age admitted to Assiut University Children Hospital with bleeding tendency caused by acquired platelet disorders were included.

Exclusion criteria

(1) Bleeding tendency not caused by platelet disorders (e.g. vasculitis, coagulation factors deficiencies)

- (2) Bleeding tendency because of inherited platelet disorders (e.g. Bernard–Soulier syndrome, Glanzmann's thrombasthenia)
- (3) Neonates
- (4) Malignancies (e.g. leukemias, lymphomas).

Tools of study

An observational checklist based on the American Society of Hematology guidelines 2011 developed by the investigators was used to decide the management plan for the treatment of the acquired platelet disorders.

Ethical approval

Institutional Review Board, Faculty of Medicine, Assiut University approved the study.

Results

This study was carried out on 76 cases with acquired platelet disorders admitted to Assiut University Children Hospital over 1 year in the period from the 1 May 2015 to the 30 April 2016. Their ages ranged from 2 months up to 17 years.

The duration of admission ranges from 2 days up to 26 days, with an average admission duration of 7.5 days.

The results of the study are shown in Tables 1-3 and Fig. 1.

CBC was performed for all cases, whereas a peripheral blood smear was performed only for 46 (60.5%) cases.

Bone marrow examination was performed in 30 (39.5%) cases, and it was indicated in two (77%) of three cases, whereas in seven (23%) cases, it was done for the purpose of research with guardian consent.



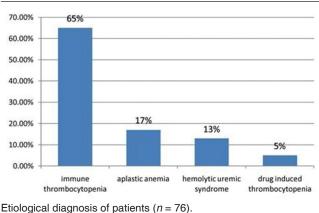


Table 1 Complete blood count and peripheral blood smear in the studied cases

Items	n (%)
Pancytopenia	11 (14.5)
Bicytopenia	32 (42)
Isolated thrombocytopenia	33 (43.5)
Large platelets	8 (10.5)

Table 2 Other investigations for the studied cases

Items	n (%)
Renal function tests	40 (52.5)
Liver function tests	33 (43.5)
Antinuclear antibodies	4 (5)
Abdominal ultrasound	21 (27.5)
Bone marrow examination	30 (39.5)
Indicated	23 (77)
Nonindicated	7 (23)

Table 3 Treatment used in the studied cases

Items	n (%)
Observation only	20 (26.5)
Intravenous immunoglobulin	22 (29)
Corticosteroids	26 (34)
Platelet concentrate	1 (1.5)
Platelet-rich plasma	24 (31.5)

Medical treatment (in the form of IVIG and/or corticosteroids) was administered to seven (9%) cases in whom treatment was not indicated. IVIG was administered to 22 (29%) cases and there was a delay in its administration to 10 (45%) of these cases. Corticosteroids were administered to 26 (34%) cases; eight (31%) of these cases received it for a longer time than indicated.

Anti-Dimmunoglobulin was not administered to any case.

Discussion

In this study, peripheral blood smear was not performed in 30 (39.5%) cases; however, it is considered a necessary evaluation for all cases with acquired platelet disorders.

Bone marrow examination may be indicated in the presence of abnormalities in the history, physical examination, or the CBC (other than thrombocytopenia) and peripheral blood smear. In this study, bone marrow examination was performed for 30 (39.5%) cases and it was indicated for 23 (77%) of these cases, whereas in seven (23%) cases, it was done for the purpose of research with guardian consent.

A single dose of IVIG should be used as a first-line treatment for ITP, especially when a more rapid increase in platelet count is required. In this study, IVIG was administered to 22 (29%) cases with ITP and there was a delay in its administration to 10 (45%) of these cases because of unavailability.

Medical treatment should not be provided for all cases with ITP unless indicated according to guidelines. In this study, seven (9%) cases received medical treatment such as corticosteroids and IVIG, whereas observation is the only requirement, which can be explained by they are far away from our center.

Treatment with anti-D immunoglobulin is considered the first-line therapy in Rh+ nonsplenectomized children with ITP if there is no decline in blood hemoglobin or evidence of autoimmune hemolysis. No cases received anti-D immunoglobulin in this study, which can be attributed to unavailability of the drug.

Corticosteroids were administered to 26 (34%) cases; eight (31% of these) cases received it for a longer time than indicated, whereas there is no evidence to support the use of corticosteroids for longer courses compared with guidelines, but this can be explained by the noncompliance of the patient in the follow-up.

In this study, no cases with acquired platelet function disorders were registered.

Conclusion

In this audit, we analyzed the management protocol of acquired platelet disorders followed in Assiut University Children Hospital with the aim of improving the ultimate management of patients admitted with acquired platelet disorder. The results showed some deviations from the guidelines in terms of investigations and treatment administration. Shortage of supply, noncompliance, and difficult follow-up may lie behind these shortages.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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