Case Report

Successful use of the cell separator hemonetics multicomponent collection system+ for therapeutic thrombocytapheresis in a low body weight child of essential thrombocythemia

Abhay Singh, Soniya Nityanand[1], Atul Sonker, Sanjeev Kumar[1]

Departments of Transfusion Medicine and [1]Hematology, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

Access this article online

Website: www.ajts.org

DOI: 10.4103/0973-6247.162722

Quick Response Code:

Correspondence to:
Dr. Abhay Singh,
Department of Transfusion Medicine, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India.
E-mail: abhaysgpgi@gmail.com

Abstract:

In children, essential thrombocythemia (ET) is extremely rare with an incidence of 1/million. Since thromboembolic complications are more common than hemorrhagic manifestation, immediate thrombocytapheresis by an automated cell separator can prevent untoward consequences in the form of cerebrovascular, coronary or peripheral vascular occlusive events. Due to varied options of automated cell separators, selecting an appropriate cell separator in such acute emergency situation can be confusing for a treating physician, especially if the patient is a child of low body weight. We present here the successful use of hemonetics multicomponent collection system (MCS+) for therapeutic platelet reduction (TPR) in a 12-year-old male child of 28 kg with extreme thrombocytosis (TS) (3072 × 10^9/l) due to ET. A total of three procedures were performed without priming of the machine with allogenic blood. We observed hemonetics MCS+, best suited for TPR even in children with low body weight.

Key words:

Automated cell separators, essential thrombocythemia, therapeutic platelet reduction

Introduction

Prevalence of essential thrombocythemia (ET) in the general population is approximately 30/1,00,000 and the reported annual incidence rates range from 0.59 to 2.53/1,00,000 inhabitants. The median age at diagnosis is 65-70 years with the disease affecting women more than men (2:1).[1] Thrombocytosis (TS) or elevation in the peripheral blood platelet count to values >400,000/μL is common in infancy and childhood, occurring in 3-13% of children.[2] Extreme TS (platelets >1,000,000/μL) is rare, occurring in <2% of children,[3] but may be common in critically ill children.[4] Primary TS is even rarer in childhood, which can be familial or essential. In children ET is extremely rare with an incidence of 1/million, that is 60 times lower than in adults.[5]

Essential thrombocythemia an acquired myeloproliferative disorder, characterized by a sustained elevation in the platelet count (≥450 × 10^9/L) which may present with thrombotic and hemorrhagic events during its clinical course.[1,6] Thromboembolic complications are more common than hemorrhagic manifestation with bleeding episodes only in cases of extreme TS. Arterial thrombosis occurs more frequently than venous episodes leading to cerebrovascular, coronary or peripheral vascular occlusive events.[7] Patients presenting with vasomotor disturbances in the form of headaches, visual disturbance, atypical chest pain, digital paresthesia should be kept under high suspicion of thromboembolic complication and urgent planning for mechanical platelet removal should be sorted to. Immediate thrombocytapheresis by an automated cell separator can prove as a life saving measure for these patients. The present day scenario provides us with varied option of automated cell separators, but selecting an appropriate cell separator in such acute emergency situation can add to the dilemma of treating physician, especially if the patient is a child of low body weight. We, therefore, present here our experience of successfully using automated cell separator hemonetics multicomponent collection system (MCS+) for therapeutic platelet reduction (TPR) in a child of low body weight suffering from ET.

Case Report

A 12-year-old male child was referred to the hematology unit of our hospital presenting with intense headache and abnormally elevated platelet count. Physical examination revealed patient to be

of 28 kg and well oriented to time, place and person. There was no evidence of any superficial bleeding in the form of echymotic patches or deep seated bleeding. All sensations were intact and on abdominal examination no hepatosplenomegaly was detected. Urgent laboratory investigation was planned in the form of complete blood counts and radiological evaluation of head and abdomen. Computed tomography (CT) head and ultrasound sonography abdomen detected no anomaly. No evidence of deep vein thrombosis or venous occlusion was detected on Doppler studies. On laboratory blood investigation extremely high platelet count (3072×10^9/l) was observed, rest all parameters appeared to be within normal limits [Table 1]. Further investigation were planned to confirm the diagnosis and in the meantime an urgent thrombocytapheresis was planned to prevent any impending thromboembolic episode, for which the patient was referred to Transfusion Medicine Department.

On further evaluation total blood volume of patient was calculated as 2280 ml (80 ml/kg body weight)[8] and after assessing the central venous line, MCS+ automated cell separator (Hemonetics Corp, Braintree, MA, USA) with discontinuous flow centrifugation technology was selected among the several apheresis machines available in Transfusion Medicine department, for performing immediate bed side TPR. Closed system disposable kits, with 125 ml bowl capacity (kit number: REF0961E, lot number: WW13075) with anticoagulant citrate dextrose solution A (ACD-A) as the anticoagulant solution were used for the procedure. Per procedure 8-10 cycles of blood circulation was performed using a 12 Fr (French) central venous double lumen dialysis catheter maintaining anticoagulant ratio of 1:8 (1 ml ACD-A with 8 ml blood). Blood flow rate was maintained at 70 ml/min. In order to avoid any hypocalcemic complications, 100 ml of 10% calcium gluconate (10 ml calcium gluconate in 100 ml normal saline) was given intravenously at a slow rate throughout the procedure. Two units of fresh frozen plasma (200 ml each) were also infused using separate venous line throughout each procedure in order to maintain normal hemodyanamic status of the patient. Extra corporeal volume was set at 15% of the total body volume for each procedure following the manufacturer's instructions. No priming of the machine with allogenic blood was carried out before any of the procedure. A postprocedure peripheral blood count was obtained 1 h and 24 h after every TPR. After each TPR a rebound increase in circulating platelet was observed [Figure 1]. A total of three procedures were performed within 7 days. About 30%, 39.2% and 40.9% of circulating platelets were removed in each of the respective procedures. The whole blood processed ranged from 2164 ml to 2362 ml with a total ACD-A infusion and total time taken ranging from 249 ml to 277 ml and 103 min to 142 min. Platelet counts at the end of third procedure were 1087×10^9/l [Table 2]. A decreasing trend of platelet count started to appear as the patient was maintained on hydroxyurea (1 g/day) and aspirin (80 mg/day) from the start of treatment [Figure 1].

Table 1: Patient characteristics

Laboratory parameters	Values
Complete blood counts	
Hemoglobin (g/dl)	11.6
TLC (10^9/l)	18.3
Platelet (10^9/l)	3072
HCT (%)	38.3
MCV (fl)	79.3
MPV (fl)	8.7
Clinical chemistry	
LDH (U/l)	1181
SGOT (U/l)	103
SGPT (U/l)	124
Serum creatinine (mg/dl)	0.44
Serum Ca^{2+} (mg/dl)	9.65
Na^+ (mmol/l)	137
K^+ (mmol/l)	6.3
Serum iron (μg/dl)	70.1
Serum ferritin (μg/l)	88.31
Transferrin saturation (%)	28.9
Serum transferrin (mg/dl)	327
Total iron binding capacity (μg/dl)	309.7
Bone marrow examination	
Myeloid and erythroid cells adequate and showing normal maturation, megakaryocytes increased in number and found in clusters with hyperlobation	
Genetic assays	
BCR-ABL	Negative
JAK2V617F	Negative
Others	
TPO (pg/ml)	78.9
PT (s)	12.4
APTT (s)	31.7
INR	1.17

MCV: Mean corpuscular volume; MPV: Mean platelet volume; SGOT: Serum glutamic oxaloacetic transaminase; SGPT: Serum glutamic pyruvic transaminase; APTT: Activated partial thromboplastin time; TLC: Total leucocyte count; HCT: Hematocrit; LDH: Lactate dehydrogenase; PT: Prothrombin time; INR: International normalized ratio; TPO: Thrombopoietin

Discussion

In patients presenting with extreme TS the clinicians should act aptly to prevent any thromboembolic, as well as hemorrhagic consequences. ET being a diagnosis of exclusion, urgent mechanical removal of platelets should be planned even before the diagnostic evaluation of such cases in order to prevent complications. Very few cases of successfully using automated cell separator for mechanical platelet removal in pediatric and adult patients of TS have been reported.[9,10] This may be because of physicians considering the activation of platelet and coagulation factors due to blood flowing through these apheretic devices but Ullrich *et al*.[11] have very well demonstrated the absence of platelet or coagulation activation by cell separators used for thrombocytapheresis.

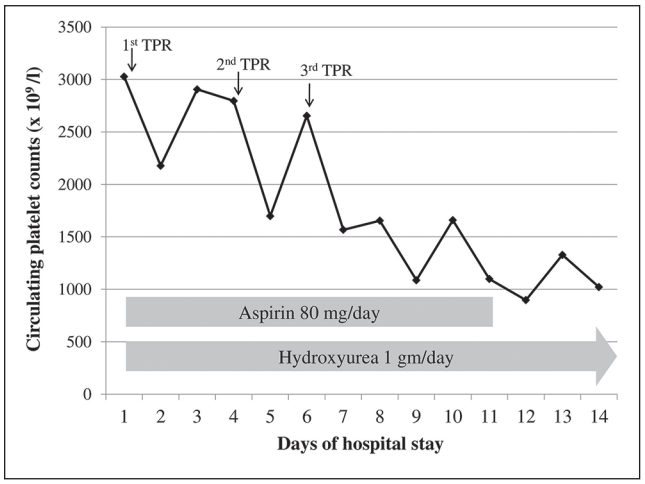

Figure 1: Effect of therapeutic platelet reduction on circulating platelet counts with Y-axis showing platelet counts and X-axis showing days of hospital stay

Table 2: Peripheral blood counts pre and post each TPR procedure

Parameters	First TPR			Second TPR			Third TPR		
	Precounts	1 h postcounts	24 h postcounts	Precounts	1 h postcounts	24 h postcounts	Precounts	1 h postcounts	24 h postcounts
Hemoglobin (g/dl)	11.6	11.1	11.9	10.8	10.9	11.1	12.1	11.9	11.1
Platelet (10^9/l)	3072	2178	2906	2796	1698	2238	2654	1567	1087
TLC (10^9/l)	18.3	12.4	13.1	9.8	9.1	8.7	10.2	9.8	11.3
HCT (%)	38.3	36.5	38.4	35.4	36.4	36.2	40.1	38.9	39.7

TPR: Therapeutic platelet reduction; TLC: Total leucocyte count; HCT: Hematocrit

McCarthy *et al.*[12] have reported the use of platelet apheresis machine in 11 years child successfully, wherein the platelet count was not as high as found in the present case. A rebound phenomenon of increase in platelet counts is often seen after therapeutic thrombocytapheresis, we did observe such phenomenon in a case reported earlier.[9] This rebound phenomenon may increase the frequency of TPR procedures. In the present patient, three procedures were done though earlier we had reported a patient requiring seven procedures. Until date, no consensus exists for the safety of TPR in children of ET regarding the frequency and timing of procedures with number of cycles to be performed in each procedure, choice of machine, type and dose of anticoagulant to be used. Here the patient was effectively managed by three procedure rather than seven procedures, the reasons for which are obscure, which needs further evaluation as this may be dependent on age of onset of disease or body response to cytoreductive therapy. The previous patient requiring seven TPR was nonresponsive to cytoreductive therapy for 2 months from start of therapy, and the present case responded to hydroxyurea within 14 days.

Proper selection of apheretic machine in such emergency situation can add to the safety of the procedure. We observed hemonetics MCS+, due to its option of controlling extracorporeal volume according to body weight of patient and compatibility with pediatric disposable kits, to be appropriately suitable for these patients. We here successfully used this machine in a patient of 28 kg, which was far below the lower acceptable limit for body weight of 40 kg. This experience of ours further proves the efficacy and safety of hemonetics MCS+ for therapeutic thrombocytapheresis even in low body weight patient.

References

1. Brière JB. Essential thrombocythemia. Orphanet J Rare Dis 2007;2:3.
2. Sutor AH. Thrombocytosis in childhood. Semin Thromb Hemost 1995;21:330-9.
3. Yohannan MD, Higgy KE, al-Mashhadani SA, Santhosh-Kumar CR. Thrombocytosis. Etiologic analysis of 663 patients. Clin Pediatr (Phila) 1994;33:340-3.
4. Denton A, Davis P. Extreme thrombocytosis in admissions to paediatric intensive care: No requirement for treatment. Arch Dis Child 2007;92:515-6.
5. Mantadakis E, Tsalkidis A, Chatzimichael A. Thrombocytosis in childhood. Indian Pediatr 2008;45:669-77.
6. Harrison CN, Bareford D, Butt N, Campbell P, Conneally E, Drummond M, *et al*. Guideline for investigation and management of adults and children presenting with a thrombocytosis. Br J Haematol 2010;149:352-75.
7. Tefferi A, Elliott M. Thrombosis in myeloproliferative disorders: Prevalence, prognostic factors, and the role of leukocytes and JAK2V617F. Semin Thromb Hemost 2007;33:313-20.
8. Pearson H. Blood and blood forming tissues. In: Rudolph C, Rudolph A, editors. Rudolph's Pediatrics. 21st ed. New York: McGraw-Hill; 2003. p. 1521.
9. Singh A, Chaudhary R, Nityanand S. Successful management of acute bleeding in essential thrombocythemia using automated cell separator. Transfus Apher Sci 2014;50:56-8.
10. Das SS, Bose S, Chatterjee S, Parida AK, Pradhan SK. Thrombocytapheresis: Managing essential thrombocythemia in a surgical patient. Ann Thorac Surg 2011;92:e5-6.
11. Ullrich H, Kadar J, Waxenberger Y, Hohe R, Saueressig C, Heyder M, *et al*. Therapeutic thrombocytapheresis in patients with myeloproliferative diseases with the cell separators Fresenius AS 104 and Cobe Spectra: Biocompatibility and safety. Beitr Infusionsther 1992;30:311-4.
12. McCarthy LJ, Graves VL, Eigen H, McGuire WA. Platelet apheresis for extreme thrombocytosis in an 11-year-old girl with CML. Transfus Med 1991;1:187-9.

Cite this article as: Singh A, Nityanand S, Sonker A, Kumar S. Successful use of the cell separator hemonetics multicomponent collection system+ for therapeutic thrombocytapheresis in a low body weight child of essential thrombocythemia. Asian J Transfus Sci 2015;9:207-9.

Source of Support: Nil, **Conflicting Interest:** None declared.