CASE REPORT
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Experience of kidney transplantation to a patient with Bernard Soulier syndrome: A case report

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ABSTRACT

Renal transplantation could be a challenging operation in patients with haemorrhagic diathesis, with predictable difficulties or even with unpredictable hurdles. Bernard Soulier Syndrome (BSS) is one of the ethiologies of the thrombocytopenia and it is a rare hereditary disease associated with defects of the platelet glycoprotein complex glycoprotein Ib/V/IX and characterized by large platelets, thrombocytopenia, and severe bleeding symptoms. Here, we present a challenging renal transplantation in BSS.

Keywords: Bernard Soulier syndrome, kidney transplantation, thrombocytopenia

INTRODUCTION

It is important to know coagulation parameters of a surgical patient, for avoiding uncontrollable bleeding. Platelet count is one of these parameters. Thrombocytopenia is defined as a platelet count below the lower limit of normal (<150.000/µL). Degrees of thrombocytopenia can be further subdivided into mild (100.000 to 150.000/µL), moderate (50.000 to 99.000/µL), and severe (<50.000/µL) (1). Severe thrombocytopenia may lead to a greater risk of bleeding. Low platelet levels and its clinical results are shown in Table 1. BSS is a disease characterized by prolonged bleeding time, thrombocytopenia, and extremely large platelets and has a prevalence of less than 1 in 1.000.000. We here in report the first case of a BSS patient undergoing a successful surgical procedure for kidney transplantation under the guidance of thromboelastography (TEG).

CASE REPORT

A 17-year-old, 172 cm, 55 kg-male patient, who had end stage renal disease due to vesicoureteral reflux, was admitted for preemptive living-related kidney transplantation. The patient was refused from many other transplantation centers, because of BSS. After routine evaluation of the patient, we planned to perform kidney transplantation with plateletpheresis transfusions by follow-up of thromboelastogram (Table 2). His preoperative platelet count was 26000/µL. The evening before surgery, two units of plateletpheresis transfusions were given in 15 minutes. The patient's platelet count was increased to 225000/µL, following plateletpheresis transfusions (Our aim was: preoperative platelet count >50000/µL). In the morning of surgery, five units of plateletpheresis were transfused in 30 minutes. In the operation period, nine units of plateletpheresis transfusions were given again in 60 minutes. Additionally, we also gave fresh frozen plasma (three units) to the patient to prevent uremic hemorrhagic diathesis. There was no bleeding complications in the peroperative period. In the postoperative period, two units of plateletpheresis transfusions were given prophylactically in 15 minutes for the first day, and one unit was given in 10 minutes for the second and third day. The patient's platelet count was between 125000 and 227000/µL, thromboelastogram values were in normal ranges, and there was no bleeding complication in the postoperative period. The patient was discharged on the fifth postoperative day with a creatinine level of 1.37

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Table 1. Thrombocytopenia classification and clinical presentation

Platelet count	Clinical results
>100.000/μL	No bleeding from thrombocytopenia
50-100.000/μL	May bleed longer than usual in response to surgery/trauma
20-50.000/μL	Bleeding from minor trauma
5-20.000/μL	May have spontaneous bleeding
<5.000/μL	Risk of life-threatening spontaneous bleeding

mg/dL. Renal function tests were normal and there were no BSS related other symptoms during the four years.

DISCUSSION

BSS is one of the ethiologies of the thrombocytopenia and it is a rare autosomal recessive coagulopathy. The syndrome was named by Dr. Jean Bernard and Dr. Jean Pierre Soulier (2). It is characterized by prolonged bleeding time, thrombocytopenia, increased megakaryocytes, and enlarged platelets. Thrombocytopenia is likely due to decreased platelet survival. This syndrome is associated with quantitative or qualitative defects of the platelet glycoprotein complex GPIb/V/IX (3). The incidence of BSS is estimated to be less than one case per million people, based on cases reported from Europe, North America, and Japan (4).

There are as yet no defined protocols for the perioperative management, which can be very complex and challenging in patients with coagulopathies, in particular BSS. When performing a literature search, our case was the first case of a Bernard-Soulier syndrome patient undergoing kidney transplantation. In a study about bleeding risk of surgery and its prevention in patients with inherited platelet disorders, Orsini S. et al recommended that prophylactic treatment is associated with a significant reduction of the bleeding frequency (5).

Following a routine evaluation of the patient, we planned to perform kidney transplantation with prophylactic treatment via plateletpheresis transfusions under the guidance of TEG. TEG is used for the diagnosis of bleeding-coagulation disorders and in determining efficacy of treatment by providing evaluation of coagulation parameters in many aspects in a short time. Table 3 summarizes the characteristics of plateletpheresis administrations in the preoperative, peroperative and postoperative period. Prior to surgery, platelet count should have a threshold of 50.000/μL. The patient's platelet count was between 125.000 and 227.000/μL, and thromboelastogram values were in nor-

Table 2. Patient's hemoglobin, platelet, creatinin, and thromboelastogram values

	Pre-op (POD -1)	Per-op (POD 0)	Post-op (POD 1)	Post-op (POD 2)	Post-op (POD 3)	Post-op (POD 4)	Post-op (POD 5)
Hemoglobin (g/dL)	10	7.2	10	9.2	9.1	9.1	9.1
Platelet (μL)	26000	225000	203000	227000	188000	156000	125000
Creatinine (mg/dL)	6.4	4.6	2	1.3	1.38	1.36	1.37
Thromboelastogram							
Platelet agregation (51-69 mm)		43.7	43.8	53.5	47.5	49	42.1
Activity of fibrinogen (55-78 degree)		52.5	58.9	43.5	52.4	54.9	44

POD: Postoperative day.

Table 3. Our plateletpheresis transfusion protocol

	Plateletpheresis transfusion protocol		
	Timing	**Units**	**Duration**
Preoperative	-The evening before surgery, -In the morning of surgery (Control platelet count: 225000/μL)	-2 units -5 units	-in 15 minutes -in 30 minutes
Peroperative	In the operation	9 units	in 60 minutes
Postoperative	-POD 1 -POD 2 -POD 3	-2 units -1 unit -1 unit	-in 15 minutes -in 10 minutes -in 10 minutes

POD: Postoperative day.

mal ranges during in the hospital stay. There was no intra- or postoperative bleeding complications. Following an uneventful postoperative recovery, the patient was discharged on the fifth postoperative day with a creatinine level of 1.37 mg/dL, and a platelet count of 125.000/µL.

CONCLUSION

Surgical bleeding risk in inherited platelet disorders is substantial, especially in inherited platelet function disorders, like BSS. We performed living-related kidney transplantation - a major surgical operation - to a BSS patient, by guidance of TEG and prophylactic treatment. Herein, we intend to highlight that TEG guidance and prophylactic treatment would allow effective operative management and is associated with a significant reduction of the bleeding frequency in inherited platelet disorders.

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OLGU SUNUMU-ÖZET
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Bernard Soulier Sendromlu hastaya böbrek nakli deneyimi: olgu sunumu

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ÖZET

Böbrek nakli ameliyatı, Bernard Soulier sendromu (BSS) gibi hemorajik diyatezi olan hastalarda oldukça zor bir ameliyattır. BSS, trombositopeninin etiyolojilerinden biridir ve trombosit glikoprotein kompleksi glikoprotein Ib / V / IX'in kusurlarıyla ilişkili ve büyük trombositler, trombositopeni ve ağır kanama semptomları ile karakterize nadir görülen kalıtsal bir hastalıktır. Bu çalışmada BSS tanılı bir böbrek nakli olgusu sunuyoruz.

Anahtar Kelimeler: Bernard Soulier sendromu, böbrek nakli, trombositopeni

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