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Case Report

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Grey Platelet Syndrome and Pregnancy: A Case Report and Literature Review

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Abstract

Grey platelet syndrome is an uncommon hereditary platelet disorder, and is characterized by thrombocytopenia and platelet dysfunction with a specific absence of alpha-granules. Electronic microscopy is a quick test that can confirm the diagnosis. We present perinatal results of a patient with diagnosed grey platelet syndrome. We reviewed the case of a patient with grey platelet syndrome at Instituto Nacional de Perinatologia. She is 26 years old, with 21 weeks' gestation. It is noted in initial laboratories a platelet amount of 64,000/mm^3, and grey platelet syndrome is suspected, so peripheral blood smear is carried out which showed pale platelets, and electronic microscopy was performed to confirm the disorder, which showed the absence of platelet alpha-granules. A female newborn was delivered at 38.4 week of gestation by abdominal cesarean section, weighing 2,588 g, with platelet count at birth of 119,000/mm^3. There is no general consensus of treatment in patients with grey platelet syndrome, and management must be multi-disciplinary between obstetrics, hematology and anesthesiology services.

Keywords: Pregnancy; Thrombocytopenia; Syndrome; Grey platelet

Introduction

Grey platelet syndrome is an uncommon hereditary platelet disorder first described in 1971 [1]. It is characterized by thrombocytopenia and platelet dysfunction with absence of alpha-granules, resulting in grey color platelets in a peripheral blood smear with Wright stain. Pregnancy thrombocytopenia is not unusual, present in 6-15% of pregnant women by the end of the pregnancy; the most common causes are pregnancy itself (74%), pre-eclampsia (21%) and immune thrombocytopenic purpura (4%) [2]. Clinically patients present ecchymosis and mucocutaneous bleeding generally after surgical procedures [3, 4]. Most cases are diagnosed in childhood or early adolescence by a history of bleeding events. Common laboratory values are thrombocytopenia and platelet abnormalities in a peripheral blood smear; electronic microscopy is a quick test that can confirm the diagnosis [5]. Actually, there are 100 published cases and only three of them present during pregnancy [5, 6].

Case Report

Clinical record of a patient with grey platelet syndrome during pregnancy was reviewed, who was treated at Instituto Nacional de Perinatologia.

A 26-year-old patient without important personal and family background was reviewed. She has a prior pregnancy in 2013 delivered vaginally at 40 weeks of gestation apparently without complications. At the interview we noted abnormal uterine bleeding with menstrual periods increased in volume and frequency. Patient started prenatal care at Instituto Nacional de Perinatologia at 21 weeks of gestation. Among initial laboratory values, platelet count was 64,000/mm^3, and serial platelet counts varied between 55,000 and 64,000/mm^3. Associated clinical data do not present like hypertension or proteinuria. Grey platelet syndrome is suspected so that peripheral blood smear was conducted, which showed pale platelets (Fig. 1); electronic microscopy study was performed to confirm the diagnosis, which showed absence of alpha-granules (Fig. 2). An infant was delivered by abdominal cesarean section at 38.4 weeks of gestation; prior surgery one unit platelet apheresis was transfused. A female newborn was obtained weighing 2,588 g, and platelet count at birth was 119,000/mm^3.

Discussion

Grey platelet syndrome diagnosis is based on the absence of alpha-granules visible in an electron microscopy. A classic micrography shows virtually empty alpha-granules [7], such as

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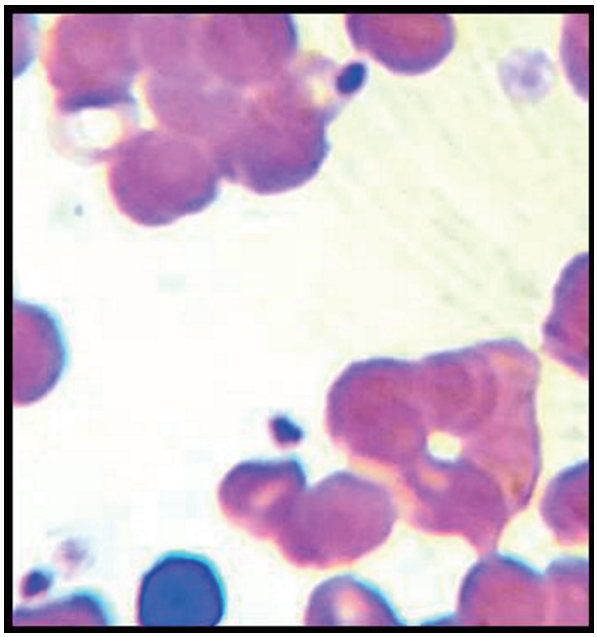

Figure 1. Pale platelets in peripheral blood smear.

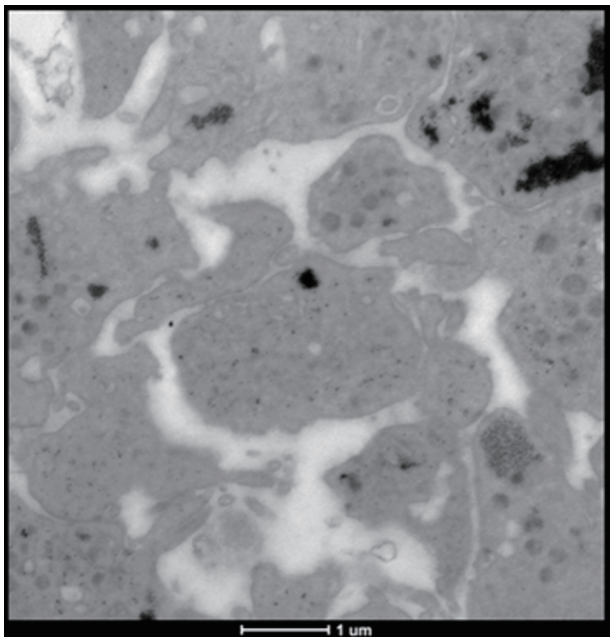

Figure 2. Electronic microscopy with absence of alpha-granules in platelets.

the one realized in our patient. Alpha-granules contain abundant proteins including fibrinogen, VIII factor and V factor [8]. In most cases of grey platelet syndrome, there is a bleeding episode from moderate to severe, myelofibrosis and splenomegaly, like so rising levels of vitamin B12, clinical and biochemical features were not observed in our patient. Bleeding mechanism is not clearly elucidated, in the studied patient, and thrombocytopenia and abnormal uterine bleeding were observed. There is no general consensus about treatment, but opinions include desmopressin and platelet apheresis transfusion prior to surgical procedures, even when platelet count is above 10,000/mm^3. Epidural and spinal anesthesia could not be recommended without demonstration of a normal bleeding time [5, 9], such as the one in the present case.

Conclusions

The incidence of congenital thrombocytopenia in pregnant women is unknown. Pregnancy course and delivery with congenital thrombocytopenia have not been described and there are no existent clear guides for management of these patients. The situation that is confronted is the risk of bleeding during pregnancy. Platelets must be prophylactically transfused to maximize hemorrhage control, using a one-donor-only platelet apheresis in order to reduce the alloimmunization risk. Close attention to any medication that can affect platelet function is suggested. Management must be multi-disciplinary between obstetrics, hematology and anesthesiology services.

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Financial Disclosure

None.

Conflict of Interest

None.

Informed Consent

The signed informed consent from the patient was obtained.

Author Contributions

All the authors actively participated in the present work.

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