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Short Communication

Paroxysmal Nocturnal Hemoglobinuria Clinical Manifestations in Pregnancy

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DESCRIPTION

Paroxysmal Nocturnal Hemoglobinuria (PNH) during pregnancy is linked to higher rates of morbidity and mortality. Studies conducted in the past indicate that the outcome has improved since the introduction of the complement inhibitor eculizumab. Every patient was registered with the international PNH registry and prospectively tracked. In 9 individuals with conventional PNH, we found 16 pregnancies, and in 2 patients with Aplastic Anaemia (AA)-PNH, we found 2 pregnancies [1]. In classical PNH, 13 pregnancies were supported by eculizumab. Six pregnancies experienced breakthrough hemolysis, requiring an increase in the biweekly eculizumab dose from 900 mg to 1,200-1,800 mg. six foetuses had red blood cell transfusions, and two received platelet transfusions. Two pregnancies were complicated with cholecystitis and the Budd-Chiari syndrome. Four of 13 pregnancies supported by eculizumab ended in spontaneous abortion or stillbirth, and one was prematurely terminated due of foetal trisomy 21. The three pregnancies that eculizumab did not support all ended in miscarriage. Preterm delivery rates were 50%, none of the patients passed away, and all but one patient's postpartum experience went smoothly. In patients with AA-PNH, both pregnancies progressed well.

Clinical manifestations

Anaemia: Hemolysis and bone marrow failure may coexist to cause anaemia in PNH, which is frequently complex. In classical PNH, intravascular hemolysis frequently coexists with moderate to severe anaemia, an increased reticulocyte count, and an elevation in Lactate Dehydrogenase (LDH) of up to 10-fold. PNH granulocytes are frequently seen in high concentrations (>50%) in patients with classical PNH. PNH typically refers to acquired aplastic anaemia when used in conjunction with other primary marrow diseases. These individuals usually have hypo cellular bone marrows, more severe thrombocytopenia, small PNH clones, lower reticulocyte counts, and minimal or no rise in LDH levels since the anaemia in these patients is predominantly caused by bone marrow failure. By definition, patients with subclinical PNH are asymptomatic, have normal or almost normal blood counts, and have few PNH (often less than 10%) granulocytes. These patients frequently have mild aplastic anaemia as their diagnosis or have recovered hematopoiesis following treatment for acquired aplastic anaemia. Relapse of their aplastic anaemia may be accompanied by expansion of the PNH clone and PNH symptoms [2].

Thrombosis: The most frequent cause of death in PNH is thrombosis, which causes significant morbidity. With PNH, thrombosis can happen anywhere; however venous thrombosis happens more frequently than arterial. Hepatic vein thrombosis (Budd-Chiari syndrome) is the most frequent site of thrombosis in PNH, with intraabdominal (hepatic, portal, mesenteric, splenic, etc.) and cerebral (sagittal and cavernous sinus) veins being other prominent locations [3]. In addition, cutaneous thrombosis, pulmonary emboli, and deep vein thrombosis are rather prevalent. In PNH, thrombophilia is multifactorial. Pro-thrombotic micro particles result from PNH platelets lacking the complement regulating proteins CD55 and CD59 that are GPI-anchored. Nitric Oxide (NO), which has been linked to nitric oxide scavenging and platelet activation and aggregation, is scavenged by high amounts of free haemoglobin. Complement activation also plays a role in PNH patients' pro-thrombotic propensity. In particular, by producing inflammatory cytokines including interleukin-6, interleukin-8, and tumour necrosis factor, C5a may lead to pro-inflammatory and pro-thrombotic activities.

Smooth muscle dystonia: Common symptoms of classical PNH include abdominal pain, esophageal spasm, dysphagia, and erectile dysfunction. These symptoms are a direct result of intravascular hemolysis and the release of free haemoglobin. Haptoglobin, CD163, and hemopexin typically remove free haemoglobin. Due to the overstimulation of these cleaning mechanisms in PNH, excessive concentrations of free haemoglobin accumulate in the plasma, depleting NO as a result. An effective NO scavenger is free haemoglobin, which reacts quickly and irreversibly with NO to produce nitrate and methemoglobin. Proper production of NO by endothelial cells keeps smooth muscle relaxed and prevents platelet activation and aggregation [4]. The scavenging of free haemoglobin results in a deficit of NO, which in turn leads to platelet activation and a dysregulation of smooth muscle tone. Therefore, patients with a

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large PNH clone size have a higher prevalence of smooth muscle dystonias.

CONCLUSION

Treatment for PNH in the past was primarily supportive. In order to treat the patients' repeated hemolysis and anaemia, blood transfusions and iron supplements were administered. To avoid thrombosis, anti-thrombosis prophylaxis was given to them. An allogeneic bone marrow transplant was available for bone marrow issues. Hemolysis mediated by complement and persistent deregulation of the alternative complement pathway are the main causes of PNH. Loss of anchoring proteins like CD55 and CD59, which frequently occurs, allows cells to hemolyze and can result in issues like thrombosis, which raises the risk of morbidity and mortality. Eculizumab, ravulizumab, and allogeneic hematopoietic stem cell transplantation are hence the mainstays of contemporary therapy for PNH. These medications target alternative complement pathways.

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