中文題目：一位已移植 7 年的活腎臟移植患者懷孕時才誘發非典型溶血性尿毒症候群

英文題目：Atypical Hemolytic Uremic syndrome triggered by Pregnancy in a living kidney transplant patient who had been transplanted for 7 years

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Introduction

Atypical hemolytic uremic syndrome (aHUS) is a life-threatening rare disease with most characterized with microangiopathic hemolytic anemia, thrombocytopenia, and acute renal function initially and then may be progressive to extra-renal system thrombotic microangiopathy and induce multiple organ failure and death. The mechanism of the atypical hemolytic syndrome may part due to complete gene mutation related, while specific drug, infection, surgery, autoimmune disease, or pregnancy induced complete activity elevation which then triggered the atypical hemolytic syndrome out off control. Therefore, while patient diagnosis with atypical hemolytic uremic syndrome and asked for living kidney transplantation, we considered is important to confirm if the potential donor with the same complement gene variation or mutation to the recipient. If the potential donor with the same complement gene variation or mutation as the recipient, the donor may with risk of atypical hemolytic uremic syndrome while he or she donate the kidney during the surgery. Pregnancy is the most popular triggered factors in atypical hemolytic uremic syndrome which may induce multiple organ failure and include abortion. There are less living kidney transplantation cases with complement gene mutation but without triggered by transplantation surgery, tacrolimus, or mycophenolate sodium who then be diagnosed as atypical hemolytic uremic syndrome after triggered by pregnancy.

Case Report

A 36-year-old female with history of IgG4/FSGS induced ESRD s/p living kidney transplantation 7 years ago. Her usual immunosuppressive drugs are prednisolone, tacrolimus, and mycophenolate mofetil and her blood pressure are normal. During the regular followed up evaluation, the nephrologist notice she had unexplained renal function progression, microangiopathic anemia, thrombocytopenia, and new hypertension. Incidental pregnancy was notice and the patient was admission for graft kidney biopsy which confirmed as thrombotic microangiopathy (TMA). Her blood pressure became difficult control and TMA progression to extra-renal system and without response to plasma exchange. However, under impression of pregnancy as the major triggered factor for her which artificial abortion is indicated for saving the patient's life. However, artificial abortion also will trigger completement system, which means, if there is no hope from anti complement agent, the patient's may progress to a very critical condition. Since anti-complement therapy is very expansive, we tried to use one 1 single dose of Eculizumab (anti C5) for suppress the complement activity which let the thrombocytopenia improved to more than 50000/ul and thrombotic microangiopathy stable enough for artificial abortion. Just like what me predicted, the artificial abortion still triggered the atypical hemolytic uremic syndrome again which with small new stroke and anemia progression. Another 2 single dose of Eculizumab were given and the patient's condition finally stable and discharged without

dialysis. We confirmed she had CR1 mutation and her living kidney donor (her elder sister) really without the same mutation.

Discussion

Pregnancy is a high-risk time for the development of different kinds of TMA. Three major syndromes including thrombotic thrombocytopenic purpura (TTP), PE/HELLP syndrome, and aHUS should be sought in pregnancy-TMA. Also, in the setting of kidney transplantation, there are different possible triggers of post-transplant atypical hemolytic uremic syndrome, such as infections, the use of immunosuppressive drugs, and rejection. However, thrombotic microangiopathy could be life threatening which need to differentiate above condition as quickly as possible. In this case, we reviewed her disease history in detail and did complete survey to rule out every differential diagnosis of TMA.

Conclusion

Atypical hemolytic uremic syndrome is a life-threatening disease. It's may become stable while the complement system under balance condition which no sign of hemolysis from blood sampling. However, while any situation induces complement activation, the multiple organ failure due to the thrombotic microangiopathy will happen. There are lots of newly diagnosis atypical hemolytic uremic syndrome patients cannot have adequate or full dose of anti-complement therapy because of the financial problem. Here we present this case that even 1 dose of Eculizumab better than nothing. It is very important to remember, remove the triggered factor is also the major point for control aHUS, meanwhile, sometimes even low dose of anti-complement can help. Early diagnosis and early treatment for atypical hemolytic uremic syndrome can let the patient's multiple organ failure improved and back to their normal life.