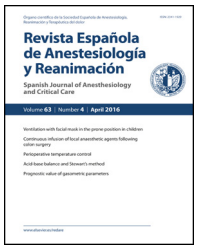
Rev Esp Anestesiol Reanim. 2018;65(6):351–355

Revista Española de Anestesiología y Reanimación

www.elsevier.es/redar



CASE REPORT

Atypical hemolytic uremic syndrome: An unusual postoperative complication☆

S. Mota*, **C. Filipe**, **A.L. Almeida**

Departamento de Anestesiología, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Received 17 October 2017; accepted 19 December 2017
Available online 1 June 2018

KEYWORDS
Atypical haemolytic uremic syndrome;
Complement factor H;
Postoperative period;
Plasma exchange

Abstract
Introduction and objectives: Thrombotic thrombocytopenic purpura and atypical haemolytic uremic syndrome (aHUS) are acute, rare, life-threatening thrombotic microangiopathies that require swift management. We report a case of acute microangiopathic haemolytic anaemia (MAHA) presenting in perioperative setting.
Clinical case: After hepatic pericystectomy for hydatid cyst, a 46-year-old female developed MAHA, thrombocytopenia and acute renal failure in the immediate postoperative period. ''aHUS'' was considered and immediate plasma exchange was initiated. Plasma exchange was performed for 2 weeks with remission of renal dysfunction. Further evaluation of genetic mutations and immunological causes for MAHA were sought. Mutations in complement factor H associated with factor H deficiency were identified, which are associated with increased risk of aHUS.
Conclusion: MAHA is a rare postoperative condition, requiring rapid differential diagnosis and treatment. Anaesthetists should bear in mind aHUS as a possible cause of MAHA, especially concerning immediate care for these patients.
© 2018 Sociedad Española de Anestesiología, Reanimación y Terapéutica del Dolor. Published by Elsevier España, S.L.U. All rights reserved.

PALABRAS CLAVE
Síndrome urémico hemolítico atípico;
Factor H del complemento;
Periodo postoperatorio;

Síndrome urémico hemolítico atípico: una complicación postoperatoria infrecuente

Resumen
Introducción y objetivos: La púrpura trombocitopénica trombótica y el síndrome urémico hemolítico atípico (SHUa) son microangiopatías trombóticas agudas, infrecuentes y potencialmente fatales que requieren una gestión rápida. Reportamos un caso de anemia hemolítica microangiopática aguda (MAHA) que se presentó en el entorno perioperatorio.

☆ Please cite this article as: Mota S, Filipe C, Almeida AL. Síndrome urémico hemolítico atípico: una complicación postoperatoria infrecuente. Rev Esp Anestesiol Reanim. 2018;65:351–355.

* Corresponding author.
E-mail address: jordana_mota_717@hotmail.com (S. Mota).

2341-1929/© 2018 Sociedad Española de Anestesiología, Reanimación y Terapéutica del Dolor. Published by Elsevier España, S.L.U. All rights reserved.

Intercambio de plasma

Caso clínico: Tras la realización de periquistectomía hepática debida a quiste hidatídico, una mujer de 46 años desarrolló MAHA, trombocitopenia e insuficiencia renal aguda durante el periodo postoperatorio inmediato. Se consideró la posibilidad de SHUa, iniciándose intercambio de plasma inmediato. Se realizó intercambio de plasma durante 2 semanas, con remisión de la disfunción renal. Posteriormente se evaluaron las mutaciones genéticas y las causas inmunológicas de MAHA. Se identificaron mutaciones en el factor H del complemento asociadas a deficiencia del factor H, que están asociadas a un incremento del riesgo de SHUa.
Conclusión: MAHA constituye una situación postoperatoria infrecuente, que requiere un rápido diagnóstico diferencial y tratamiento. Los anestesiólogos deberán considerar el SHUa como causa posible de MAHA, especialmente en lo referente al cuidado inmediato de estos pacientes.
© 2018 Sociedad Española de Anestesiología, Reanimación y Terapéutica del Dolor. Publicado por Elsevier España, S.L.U. Todos los derechos reservados.

Introduction

Thrombotic thrombocytopenic purpura (TTP) and atypical haemolytic uremic syndrome (aHUS) are acute, rare, life threatening thrombotic microangiopathies that require rapid diagnosis and treatment. These conditions are defined by erythrocyte fragmentation and elevated levels of lactate dehydrogenase (LDH) and thrombocytopenia, with renal involvement primarily present in aHUS and neurological and cardiological sequelae in TTP.[1,2]

The cornerstones of prompt treatment for most cases of TTP and aHUS are plasma exchange (PEX) and monoclonal therapy.[1–4] HUS is a rare disease with an overall incidence of 1–2 cases per 100,000[3]; it can be subdivided into typical and atypical forms. Typical HUS is a syndrome of thrombocytopenia, renal dysfunction, and microangiopathic haemolytic anaemia (MAHA) associated with Shiga toxin-producing *Escherichia coli*. The atypical form is not related to Shiga toxins and accounts for around 5–10% of all cases of HUS.[2–5] AHUS has a poorer prognosis, with death rates as high as 25% and progression to end-stage renal disease in half of all patients. In the typical form, in contrast, 3% of patients develop end-stage renal disease, and 25% present reduced renal function.[3,4]

AHUS is currently thought to be a consequence of dysregulation of the complement system associated with mutations causing excessive complement activation on the surface of the renal microvasculature. However, further environmental and possible genetic triggers are needed for disease presentation.[1,5–7] Approximately 60% of individuals with aHUS also have an inherited and/or acquired abnormality affecting components of the complement pathway. These include mutations of genes encoding complement regulators (factor H, factor I, CD46 and thrombomodulin) and/or complement activators (C3 and factor B).[1,5,7]

We present the case of a patient presenting with MAHA, and describe the differential diagnosis, its therapeutic implications, and follow-up.

Clinical case

A 46-year-old female, ASA II, with a history of vertical sleeve gastrectomy without complications, and trazodone 50 mg and alprazolam 0.25 mg for depression, was scheduled for pericystectomy for hydatid cyst of the liver. The preoperative analytical workup was unremarkable, except for slight normocytic normochromic anaemia (haemoglobin 10.2 g/dL). This was not considered relevant for the present surgical indication, and a study of its aetiology was postponed for after surgery.

Surgery lasted for 4 h and 30 min, with no intraoperative complications or observations of note, and the patient was transferred to the post-anaesthesia care unit.

In the immediate postoperative period, she became oliguric, with decreased urinary output in the following hours, and dark urine. She was calm, cooperative, hemodynamically stable, and with no clinical signs of haemorrhage. Seven hours later, we noticed a progressive fall in haemoglobin levels (Table 1). Fluid therapy and blood transfusion were initiated. Although the patient remained asymptomatic, haemoglobin levels continued to decline. When haemoglobin had fallen to below 7 g/dL, 1 unit of packed red cells (PRCs) was administered (between 7 and 8 h after surgery). Given the persistent decline of haemoglobin (Table 1), 3 more units were administered over a 17-h period from the end of surgery (total 4 units). No vasoactive drugs were needed. Blood panels showed decreased platelet count, increased creatinine values, increased LDH, and increased total bilirubin (Table 1). Coagulation studies were unremarkable. Urine analysis showed bilirubinuria, haemoglobinuria and proteinuria. Peripheral blood smear showed schizocytes, considered indicative of MAHA. Abdominal and renal ultrasound were unremarkable. The patient was asymptomatic, in other words, with no current or recent history of diarrhoea.

The surgical team denied intraoperative rupture of the hydatid cyst.

Given the coexistence of MAHA, thrombocytopenia and acute renal failure, aHUS was suspected, and PEX was initiated as soon as possible (within the first 24 h after surgery).

After PEX was started, there was no further need for PCRs. Kidney function initially improved, albeit only slightly, in response to therapy, but serum creatinine values remained high (Table 1). She was admitted for continuing haemodialysis and PEX for 2 weeks.

Further studies of genetic mutations and immunological causes of MAHA were requested. ADAMTS13 activity was normal. Anti-ADAMTS13 antibody, direct Coombs test, anti-ds-DNA antibody, anti-ANA antibody and anti-Scl70 antibody

Download English Version:

https://daneshyari.com/en/article/8622257

Download Persian Version:

https://daneshyari.com/article/8622257

Daneshyari.com