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Too tight for comfort – A rare case of hypertensive microangiopathic haemolytic anaemia

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Abstract

A 43-year-old man presented to the emergency department with a one-month history of stiffness and swelling to his hands and fatigue with progressive headache and blurring of vision. Examination revealed a blood pressure 260/160mmHg, a generalised non-blanching petechial rash and bilateral skin tightening of the hands and face. Fundoscopy showed macular oedema and retinal detachments. Investigations revealed normocytic anaemia (haemoglobin 85g/dL), thrombocytopenia (platelets 25x10 9 /L), blood film showing schistocytes, acute renal impairment (creatinine 380 umol/L, eGFR 55ml/min) and elevated lactate dehydrogenase of 1260 U/L. Haemolysis creen demonstrated low haptoglobin, reticulocytosis, negative direct Coombs test and normal ADAMTS13 level. Management of the hypertensive emergency was refractory to multiple agents prior to the introduction of captopril: A combination of intravenous glyceryl trinitrate, hydralazine, esmolol and labetalol was used. The presentation was a scleroderma renal crisis with microangiopathic haemolytic anaemia.

This case was challenging as a first presentation of undiagnosed scleroderma in renal crisis.

Early treatment of hypertension is essential to prevent irreversible renal impairment; up to 40 % of patients may require dialysis. Secondary MAHA occurs in up to 60 % of patients.

Emergency management decisions include managing hypertension, the need for plasma exchange, anti-complement therapy or dialysis. Platelet transfusion is relatively

contraindicated as it perpetuates microangiopathic thrombosis and should be reserved for life-threatening bleeding.

The key to optimal management of SRC is recognition of pathognomonic clinical signs and early institution of ACEI. MAHA is a common occurrence requiring a framework of approach to exclude alternative aetiologies which may require unique management modalities.

Introduction

A 43-year-old man presented to the emergency department with a one-month history of stiffness and swelling to his hands and fatigue with progressive headache and blurring of vision. His medical history was relevant for heavy alcohol intake and smoking. Examination revealed severe hypertension (blood pressure 260/160mmHg), a generalised non-blanching petechial rash and bilateral skin tightening of the hands and face. Fundoscopy showed macular oedema and retinal detachments, consistent with severe hypertensive retinopathy. Initial investigations revealed normocytic anaemia (haemoglobin 85g/dL), associated severe thrombocytopenia (platelets $25x10^9$/L) and a blood film showing schistocytes. In conjunction, there was acute renal impairment (creatinine 380 umol/L, eGFR 55ml/min), elevated lactate dehydrogenase (LDH) of 1260 U/L and unconjugated bilirubin 64 umol/L. The coagulation profile was normal. Subsequently, a haemolysis screen demonstrated low haptoglobin, reticulocytosis, a negative direct Coombs test and a normal ADAMTS13 level.

A CT brain was performed which showed bilateral hypodense areas in the region of bilateral basal ganglia which was consistent with ischemia from hypertensive angiopathy later confirmed on an MRI.

Acute management was aimed at treating the hypertensive emergency, which was refractory to multiple agents prior to the introduction of the ace inhibitor (ACEI) captopril: A combination of intravenous glyceryl trinitrate, hydralazine, esmolol and labetalol was used, to aim a systolic blood pressure of < 160 mmHg.

The presentation was consistent with a scleroderma renal crisis (SRC) with secondary microangiopathic haemolytic anaemia (MAHA) in the context of severe hypertension. Further investigations confirmed this diagnosis (RNA polymerase 2 positive with SCL 70 negative). The creatinine peaked at 750 umol/L, however with the introduction of regular ACEI captopril and immune suppression with mycophenolate mofetil, the renal impairment and haemolysis resolved. The major differential in this situation was a haemolytic uraemic syndrome, although there was no history of bloody diarrhoea.

Discussion

This case was challenging as a first presentation of undiagnosed scleroderma in renal crisis. Early treatment of hypertension is essential to prevent irreversible renal impairment as up to 40 % of patients may require dialysis**[1].** The optimal medication is ACEI; the most experience has been with captopril **[2].** Secondary MAHA occurs in up to 60 % of patients in this setting **[1]**.

MAHA is characterised by thrombosis of the microvasculature leading to haemolysis via shearing of red cells and end-organ dysfunction due to ischemia. The presence of severe thrombocytopenia and red cell fragmentation on the blood film is characteristic, although the onset of fragmentation can be delayed and therefore a haemolysis screen is essential. Evidence of haemolysis includes an elevated unconjugated hyperbilirubinaemia, LDH and reticulocyte count and low haptoglobin. The direct Coombs test is negative and the coagulation profile is usually normal unless disseminated intravascular coagulation is present **[3]**.

Primary causes of MAHA include thrombotic thrombocytopenic purpura (TTP) and haemolytic uremic syndrome (HUS). Secondary causes include malignant hypertension, pregnancy-related (HELLP syndrome and eclampsia), drug-induced (eg. calcineurin inhibitors), rheumatic disease (eg. scleroderma renal crisis), intravascular foreign bodies, vasculitides and malignancy**[3]**. Considering different causes of MAHA is important because SRC can masquerade as TTP and patients have been mistakenly treated for TTP **[4]**.

The emergency management decisions include managing hypertension if present, the need for plasma exchange (for TTP), anti-complement therapy (atypical HUS) or dialysis. Additionally, platelet transfusion is relatively contraindicated in thrombotic microangiopathy as the associated thrombocytopenia is a result of platelet aggregation and consumption and therefore should be reserved for cases with life-threatening bleeding**[5]**.

Conclusion

The key to optimal management of SRC in the emergency department is recognition of pathognomonic clinical signs and early institution of ACEI **[4]**. Secondary MAHA is a common occurrence requiring a framework of approach to exclude alternative aetiologies which may require unique management modalities in conjunction with nephrologists and haematologists.

Conflict of Interest: None

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