TTP: confusion, and bleeding, finding a needle in haystack seems impossible.

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Introduction:

Thrombotic thrombocytopenic purpura (TTP) was initially characterized by a pentad of symptoms, including thrombocytopenia, microangiopathic hemolytic anaemia (MAHA), fluctuating neurological signs, renal impairment, and fever, typically with a gradual onset. However, the complete pentad may not always be evident; approximately 35% of patients may lack neurological signs initially, and features such as renal abnormalities and fever may not be prominent. Updated diagnostic criteria emphasize considering TTP when both thrombocytopenia and MAHA are present.

Case report:

A 69-year-old male patient presents at the Emergency Department (ED) with acute confusion and weakness. This is morning he has nearly collapsed while trying to move from the bathroom around 5:00 AM and he was unable to take his morning oral medication for diabetes type 2 and prednisolone for his recent diagnosis of Giant cell arteritis. His urine was dark and foul-smelling. Upon examination, the patient has a Glasgow Coma Scale (GCS) score of 13. His vital signs are recorded as follows: blood pressure 124/72 mmHg, heart rate 96 beats per minute, respiratory rate 21 breaths per minute, oxygen saturation 96% on room air, and a temperature of 36.7°C. Dark urine with blood is detected in a dipstick test, and the patient seems agitated.
The blood glucose level is greater than 30 mmol/L. Initial venous blood gas (VBG) analysis shows a pH of 7.34, HCO3 of 19 mmol/L, potassium (K) of 7.1 mmol/L, sodium (NA) of 127 mmol/L, lactate of 9.7 mmol/L, and ketones of 0.1 mmol/L.
The ECG has revealed sinus tachycardia at 110 beats per minute with no hyperkaliaemic changes. However, a repeat VBG has indicated a potassium level of 7.1, prompting the initiation of an anti-hyperkaliemia bundle. During treatment, the patient has suffered a generalized tonic-conic seizure lasting 2 minutes, which was self-limiting but has caused a decrease in the Glasgow Coma Scale (GCS) from 13/15 to 8/15. A repeat VBG has revealed severe acidosis with a pH of 6.9, HCO3 of 10 mmol/L, potassium of 4 mmol/L, base excess (BE) of -20 mmol/L, lactate of 17 mmol/L, and ketones of 0.1 mmol/L. Upon reassessment, a bleeding wound in the lower lip and small petechiae in both cubital fossae are noted. The patient underwent intubation. A CT brain scan was conducted, and it showed no abnormalities.
Subsequent blood results reveal a creatinine level of 312 µmol/L, a glomerular filtration rate (GFR) of 17 mL/min, urea concentration of 16 mmol/L, indicating acute kidney injury (AKI) at stage 3. The bilirubin is measured at 54 µmol/L, Alanine aminotransferase (ALT) at 111 U/L, hemoglobin (HB) at 111 g/L, platelet count (PLT) at 22 x 10^9/L, white blood cell count (WBC) at 10 x 10^9/L, and a reticulocyte count of 125 x 10^9/L. Additionally, the international normalized ratio (INR) is recorded as 1.1. The blood glucose is 41 mmol/L. The blood film revealed fragmented red blood cells (RBCs), Schistocytes and microangiopathy (Figure 1.)

TTP remains a diagnosis based on clinical history, examination of the patient and the blood film. ADAMTS 13 assays help to confirm the diagnosis and monitor the course of the disease and possible need for additional treatments. Our patient ADAMTS 13 was less than 0.2 IU /dl and plasma exchange was initiated.

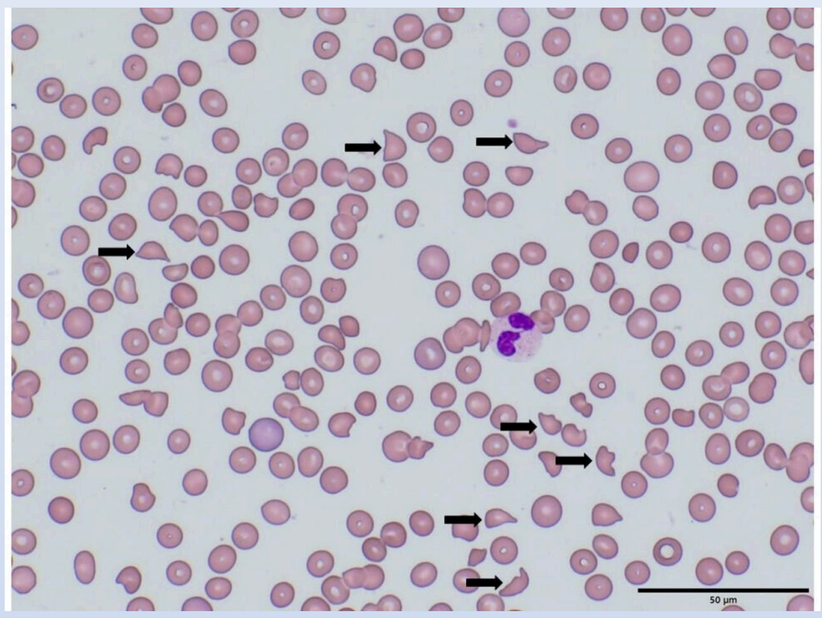

Image Source: *Peripheral blood smear of a patient with microangiopathic haemolytic anemia with schistocytes in HUS (arrows).* ResearchGate. https://www.researchgate.net/figure/Peripheral-blood-smear-of-a-patient-with-microangiopathic-haemolytic-anemia-with_fig2_261547808

Figure s

Disscssion:

The diagnosis of TTP should be treated as a medical emergency.

The primary identification of TTP should rely on the patient's clinical history, examination, and standard laboratory assessments, which encompass the review of a blood film.

Due to the substantial risk of avoidable early fatalities in TTP, it is advisable to commence treatment with plasma exchange (PEX) at the earliest opportunity, ideally within 4–8 hours. This is applicable irrespective of the presentation time during the day, provided a patient exhibits microangiopathic hemolytic anemia (MAHA) and thrombocytopenia without any alternative clinical cause

Reference

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