

BACKGROUND

- **Thrombotic thrombocytopenia purpura (TTP)** is an acute, rare, potentially life-threatening disorder that presents with **thrombocytopenia, haemolytic anaemia** and clinical consequences of microvascular thrombosis such as **stroke**.
- The exact cause is unknown but it is associated with a **deficiency of ADAMTS13 enzymes**.
- Immune mediated TTP is more common and can present In pregnancy. It is associated with pregnancy loss.

CASE



- 23-year-old female
- Parity 0
- Caucasian
- Mental health issues requiring medication
- BMI 29
- Smoker

Patient Timeline

28 weeks gestation
Routine bloods

- Low platelet count of 26
- Bruising on abdomen and legs
- **Diagnosis of Idiopathic Thrombocytopenia Purpura (ITP) given**
- Oral prednisolone prescribed
- Follow-up with Obstetric Haematology team planned

28 + 3

- Presented to Maternity Triage with reduced fetal movement, dizziness and blurred vision
- Discharged home after normal CTG and improvement in symptoms

29 + 3

- Presented to Accident & Emergency with headache, visual disturbances and transient left sided weakness
- No vaginal bleeding, abdominal pain or reduced foetal movement reported
- Referred to Obstetrics and promptly discussed with Haematology due to **deranged bloods**
- **Diagnosis of HELLP (Haemolysis, elevated liver enzymes, low platelets) syndrome suggested**

29 + 4

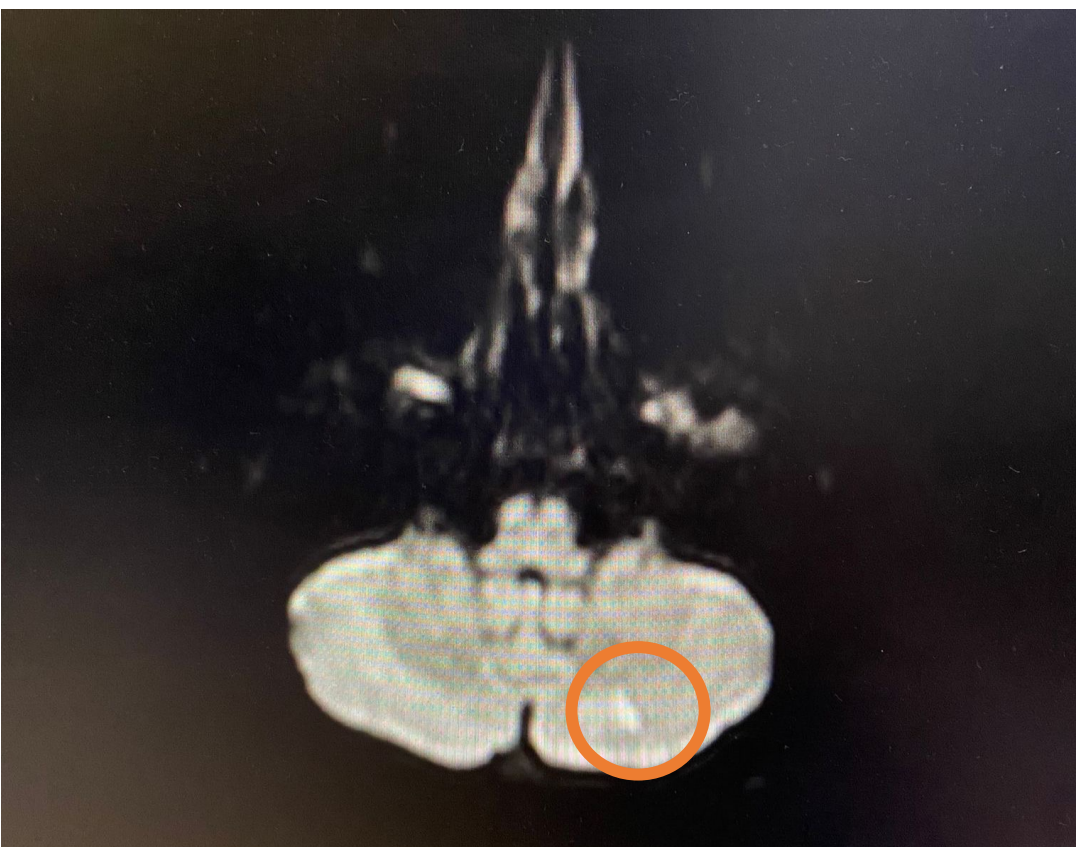
- Patient re-examined: 4/5 power left upper and lower limb, GCS 15/15
- **MRI imaging arranged**

CLINICAL FINDINGS

Bloods	Results
HB	74g/L
PLATELETS	15 *10 ⁹ /L
WHITE BLOOD CELLS	32 *10 ⁹ /L
LACTATE DEHYDROGENASE	1578 U/L

ALANINE TRANSFERASE	76 U/L
RECTICULOCYTE	275 *10 ⁹ /L
RENAL FUNCTION	NORMAL
CLOTTING	NORMAL

Bloods suggested **haemolytic anemia, thrombocytopenia and deranged liver function tests**



MRI head revealed **acute infarct in the left frontal lobe and left cerebellum likely embolic**

DIFFERENTIAL DIAGNOSES AND MDT INVOLVEMENT

- Differential diagnoses at this stage included: **atypical pre-eclampsia, haemolytic uremic syndrome and disseminated intravascular coagulation**.
- A multi-disciplinary team (MDT) discussion involving medics, stroke specialists, intensive care team, obstetricians, haematologists, obstetric physicians and anaesthetists helped to optimise care and eventually proposed the diagnosis of **TTP** based on the triad of **thrombocytopenia, haemolytic anaemia and ischemic stroke**.

OUTCOME

- The patient was transferred to a tertiary centre for **diagnostic ADAMTS13 testing and ultimately, plasma exchange**.
- The pregnancy continued until 33+5, when she underwent an emergency caesarean section for foetal concerns. The procedure was uneventful.
- Monoclonal antibodies, rituximab, were given postnatally.
- The patient remains under surveillance and has recovered well.

DISCUSSION

- TTP is characterised by widespread intraluminal and sub-endothelial platelet thrombin in capillary and pre-capillary arterioles.
- It is diagnosed by a **deficiency of ADAMTS 13** and the presence of antibodies against it.
- ADAMTS13 is an enzyme which breaks down Von Willebrand factor. Without this enzyme, VWF stays in circulation and can increase platelet coagulation and blood cell destruction.
- It occurs in 1 in 25,000 -100,000 pregnancies, mostly in the third trimester.
- This condition carries a 90% mortality rate if untreated.
- Symptoms often consist of the **pathogenic triad: thrombocytopenia, macroangiopathic haemolytic anaemia and neurological symptoms**.
- 40% of these patients present with **additional symptoms of renal impairment and high temperature, making up the full pentad**.
- **Treatment includes plasma exchange or plasma infusion or plasma exchange that aims to correct severe deficiency of ADAMTS 13.**
- Immunosuppressive therapy is also used, including steroids or monoclonal antibodies in the post natal period.
- Delayed management may have significant sequelae such as placental infarction, foetal growth restriction, pregnancy loss and maternal mortality and morbidity.

CONCLUSION/ FUTURE PLANNING

- This case highlights the **importance of MDT involvement** to achieve the correct management and diagnosis of patients in a **time-sensitive manner**.
- TTP patients require **pre-pregnancy counselling** to plan care under specialist centres, provide health education and prognostic information and to allow optimisation of the condition prior to conception.

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