MEDICINE 2019 – RCP ANNUAL CONFERENCE

CLINICAL CASE

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SETTING THE SCENE

• Who?
• What?
• Where?
• When?
REFERRAL DETAILS

- 83M
- Severe rheumatoid arthritis (on pred 2.5mg), gout, IHD, BPH
- Generally unwell with rash on arms
- Treated for UTI ~1/52 earlier with Nitrofurantoin

- FBC: Hb 48, MCV 82.6, Plts 319, WCC 13.9
- Coag screen: PT 13.8, APTT 88.2

- eGFR 73
- LFTs normal
- CRP 36
Lupus antibody

Drugs

DIC

Contaminated sample
NEXT STEPS

• Top to toe examination

• Fibrinogen → 3.17

• Haematinics
  • Ferritin 87
  • B12 284
  • Folate 8.2

• Haemolysis screen
  • DCT +++
  • Reticulocyte count 83
  • Bilirubin 14
  • LDH 274
  • Haptoglobin 3.2
URGENT FVIII RESULT = 1% (70 – 150%) 

• **Acquired Haemophilia A**

• Rare, but potentially life-threatening
• Median age 75 – 80 yrs
• Time & temp dependent inhibitor
• Pattern of bleeding
• Frequently have underlying malignancy

• Mortality 8-42%*

*Green & Lechner, 1981; Morrison et al, 1993; Hayet al, 1997; Delgado et al, 2003; Collinset al, 2007*
INITIAL MANAGEMENT

1. Stop the bleeding ➔ FVIII bypassing agent

2. Suppress the antibody ➔ immunosuppression
FEIBA MECHANISM OF ACTION

- FEIBA contains Factors II, VII, IX, X and variable amounts of their activated forms; small amounts of FVIII too.
Delays >12 days in a quarter of patients*

*Knobl et al, 2012
REFERENCES

- Diagnosis and management of acquired coagulation inhibitors: a guideline from UKHCDO. A United Kingdom Haemophilia Centre Doctors’ Organization (UKHCDO) guideline approved by the British Committee for Standards in Haematology. Collins et al. [accessed 16th April 2019]

KEY FLAGS TO PICK UP

• Unexplained, isolated & very prolonged APTT
• Extensive subcutaneous bruising
• No prior bleeding hx

• Contact your local haemophilia centre

• Acquired Haemophilia