Haematological Emergencies

Acute and General Physician Study Day
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Outline of presentation

• 5 clinical case studies
• Common and uncommon haematological emergencies
• Guidelines listed where available
Patient 1

03:40am – review by foundation doctor

19 yr old male with homozygous Sickle Cell Disease (Hb SS)

- Admitted with 1 week history of generalised pain not responding to oral analgesia
- Admission precipitated by acute chest pain and limb pain
- PH of CVA (residual mild L hemiparesis) aged 12 and epilepsy
- On regular transfusion protocol and s.c. desferrioxamine
Examination (post analgesia)

- Afebrile - temp 36.2
- Pain score 5/10
- P96, BP 126/82, RR 24, O2 sats (air) 92%
- Chest: few R basal crackles
- Distended tender abdomen. BS present
Investigations and initial management

- CXR and AXR ordered
- FBC: WBC 19.3, Hb 7.6, Plats 214, retics 19%
- Sickle screen (HbS 71%)
- D Dimer: 1661ng/ml
- CRP: 5mg/l
- LFTs: ALP375, Bil 156
- Septic screen sent

• Started on Morphine PCA, IV fluids
• Penicillin V 250mg bd + folic acid continued
Five hours later

08:30 am

• Tired, chest pain worsening
• $O_2$ sats (on air) 80%, RR 33/min
• CXR: L basal pneumonia and interstitial shadowing
• ABG(air): $PaCO_2$ 5.1 KPa, $PaO_2$ 8.1 KPa
What is the diagnosis?

A. Pulmonary embolus
B. Pneumonia
C. Acute chest syndrome
D. Painful sickle cell crisis
Subsequent management

• Transferred to HDU for CPAP
• Manual exchange transfusion
  – 3 cycles of 2 units
  – Aim HbS < 30%, Hb not > 12g/dl
• Post exchange %HbS 27.9, Hb 8.2g/dl
• Further top up transfusion 2u
• Remained on CPAP and opiates for 4 days
• Discharged home 5 days later
Acute Chest Syndrome (ACS)

1/3 SCD patients have 1 or more chest syndromes

• Definition:

  New infiltrate on CXR
  +
  Acute respiratory symptoms

• Risk factors:
  – Age (children : adults 3:1)
  – Low fetal Hb levels
  – High baseline Hb level
  – Bronchoreactive long disease
Aetiology

- Infarction
  - vascular sickling

- Infection
  - bacterial (atypical)
  - viral

- Fat embolism
Clinical Features

• 15% require intubation
• 11% develop neurology events & multi-organ failure
• Mortality 3% overall, 9%>18y
  – ↑ Morbidity + mortality is associated with:
    • multilobar pneumonia
    • platelets < 200,00 x 10⁹/l
    • previous history of cardio-pulmonary disease
Management

Expectant:
- patients with chest pain or fever
- previous history of chest syndromes
- respiratory signs/symptoms

- CXR
- O₂ saturation monitoring (AIR) → ABG if <92%
- Encourage breathing (analgesia (PCA) v narcosis)
Treatment of ACS

- Routine analgesia and fluids (do not overhydrate)
- Antibiotics: cefuroxime + clarithromycin
- Monitor PaO₂ on air
- PaO₂ < 9.5KPa (70mm Hg) consider CPAP
  - < 8.0KPa (60mm Hg) CPAP + exchange transfusion
  - < 7.5KPa (55mm Hg) Ventilate
Take home messages

• Watch for ACS in acute sickle cell crisis especially if chest pain, respiratory symptoms or fever
• Frequent review necessary as ACS often develops after admission
• Monitor O2 sats on air in SCD patients

Howard et al, British Journal of Haematology, 2015, 169, 4, 492-505
Guideline on the management of Acute Chest Syndrome in Sickle Cell Disease
Patient 2

- 54 year woman
- Diagnosis of high grade lymphoma
- Received chemotherapy 1 week ago
- Presents to A+E with a temperature of 38°C
What antibiotics are you going to give?

A. Tazocin (piperacillin with tazobactam) and Gentamicin
B. Tazocin and Amikacin
C. Tazocin alone
D. Co-amoxyclyclav
Chemotherapy and neutropenia

- Variety of regimens
  - Ranges from “high dose” chemotherapy before stem cell transplant to low dose out-patient regimens
  - Blood counts often reach a nadir approx 7 days post chemotherapy (depends on regimen)
  - Patients should carry an “alert card”
  - Fluoroquinolone prophylaxis used frequently
Neutropenic sepsis

• Neutrophil count $< 0.5 \times 10^9/l$ and either
  – Temp higher than 38°C
  – Other signs or symptoms consistent with clinically significant sepsis

Nice Guidance, Sept 2012
Investigation and Management

• Offer empiric antibiotic therapy immediately
  – Beta lactam monotherapy
  – Add aminoglycoside only if patient-specific or local microbiological indications

• History and examination

• FBC, U+Es, LFTs, albumin, CRP, lactate and blood culture, line cultures, urine dipstick, chest X-ray, throat swabs

Nice Guidance, Sept 2012
Case 3

• 62 year old man attends GP
• No significant past medical history
• History of back pain, constipation and urinary retention
What condition are you worried about?

A. Muscular back pain
B. Osteoporosis
C. Spinal cord compression
What percentage of patients with SCC do not have a history of cancer?

A. 4%
B. 10%
C. 23%
D. 56%
Why is SCC often missed?

• Low back pain is common
• 23% of patients with spinal cord compression have no previous history of cancer
• One study showed took 2 months to make diagnosis

http://www.bmj.com/content/342/bmj.d2402
MRI Scan – spinal cord compression
Why does it matter?

• Patients with motor dysfunction progress to complete paralysis in the absence of intervention.
• Almost half of patients cannot walk by the time of diagnosis.
• Neurological status at the time of diagnosis, particularly motor function, has been shown to correlate with prognosis.
• Treatment before paralysis is clinically and cost effective.
Clinical Features

• Pain (83-95% of patients)
• Thoracic pain (70% of SCC is thoracic)
• Upper motor neurone signs
• Anaesthesia
• Paraesthesia
• Bladder dysfunction (40-64%)
• Bowel dysfunction
Spinal Cord Compression – key points

- History and examination
- Early MRI scan
- Urgent steroids
- Refer to acute oncology team
  - Radiotherapy
  - Surgery
  - Supportive care

Patient 4

• 25 year old female
• Presented to A+E with fever and headache
• FBC shows platelet count of 15 x 10⁹/l
What is the most likely diagnosis?

A. APML
B. Sickle cell crisis
C. TTP
D. B12 deficiency
TTP—clinical features

Key features

- Thrombocytopenia
- MAHA
- Neurological impairment
- Renal impairment
- Fever

Not present in around 35% of patients

Not prominent features
Management

• Commence treatment based on routine history, examination and blood film results
  • Send virology screen and ADAMTS13 assay (severely reduced in TTP)

• Plasma exchange
  • Initiate within 4-8 hours
  • Reduces mortality from 90% to 10-20%
  • Large volume plasma infusions indicated if delay in plasma exchange

Scully et al, BJH, 2012, Guideline on diagnosis and management of TTP and other thrombotic microangiopathies
Patient 5

- 27 year old male
- Presents to A+E with bruising and fever
- Hb 92g/l WCC 1.9 x 10^9/l (neutrophils 0.3 x 10^9/l) Platelets 38 x 10^9/l
- Prothrombin time 23 secs (11-14 secs) APTT 50 seconds (28-37 seconds)
Blood film
What condition are you most worried about?

A. Acute myeloid leukaemia
B. Acute lymphoblastic leukaemia
C. Acute promyelocytic leukaemia
D. Thrombotic thrombocytopenic purpura
Acute promyelocytic leukaemia

• Needs urgent treatment with ATRA (all-trans retinoic acid)
• High early mortality due to haemorrhage
• Good long term prognosis
Summary – Haem emergencies

• Acute chest syndrome in sickle cell disease
• Febrile neutropenia
• Spinal cord compression
• Thrombotic thrombocytopenic purpura
• Acute promyelocytic leukaemia