Balancing the Risks – a Difficult Case of Sickle Pregnancy

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Mrs AA 30y

- Sickle cell anaemia (Hb SS)
- Arrived in UK August 2010
- Sept 10 uncomplicated VOC sickle crisis

Clinic review:

- Mild exertional dyspnoea
- Infrequent admissions in Nigeria
- Never transfused

Investigations

- Baseline Hb 6-7g/dl
- Red cell genotype sent
- Respiratory function tests

November 2010

- Painful crisis leg and shoulder
- 6/40 pregnant
- Hb 6.9g/dl
- Management: Morphine PCA and fluids
- Day 2:
  - O_2 sats 92% room air. $P0_2$ 9.14KPa
  - Chest X-ray: shadowing and atelectasis R base
  - Hb 5.6g/dl

- What would you do next:

1. Transfuse and standard care
2. Standard care only

- Transfused 2 units Rhesus and Kell matched blood
- Post transfusion Hb 8.2g/dl
- Day 4 post transfusion Hb 8.9g/dl discharged

- 10/40 delayed miscarriage: ERPC

Feb 2011

- Admission with painful crisis
- Hb 6.6g/dl falling to 5.3g/dl next day
- Pyrexial, increasing respiratory rate and pulse
- O_2 sats 78% air
- Bilateral basal crackles

- Diagnosis: Acute Sickle Chest Syndrome

Management:

- Transfused 3 units

- post transfusion PaO_2 6.8kpa (2/3/11)
  - Red cell exchange 6 units (Hb S 26% post)
- 48h pain free. O_2 sats 96%air
  - Hb post exchange 10.5g/dl

8/3/11 (day 6 post tx) Readmitted

- Jaundice, haemoglobinuria
- Febrile
- Hb 7.3 g/dl

Diagnosis: Delayed haemolytic transfusion reaction

Investigations:

- Direct antiglobulin test weakly positive
- Blood cultures: no growth
- Antibody screen negative (weak enzyme only; prob non sig)
- Eluate: weak non specific antibody detected by IAT
- Retics 8.6%
- MSU E Coli

(Transfusion 2/3/11)

Management

Date	8/3	9/3	10/3	10/3	11/3*	12/3	13/3	15/3	16/3
Hb (g/dl)	7.3	5.6	5.1	4.3	4.0	4.0	4.4	5.7	6.4
Retics x109/l	8.6		18.1		24.6	31.6	42		

HbS 37%

* Management ?

1. Observation only
2. Transfusion
3. Ivlg
4. Steroids
5. 2 and 3
6. 2 and 4
7. 2,3 and 4

Hyperhaemolysis management

- IvIg 0.4g/kg/day
- Prednisolone 60mg/kg/day decreasing over next 4 weeks

- No antibody identified
- ? antiJKb as culprit

Clinic review

- Increasing breathlessness
- PFTs Restrictive defect 62% predicted
- Wanting to get pregnant

- Referred to pre-conceptual clinic
- DNA x1
- Next OPA 14/40 pregnant!
- Partner Hb A only. Red cell genotype performed

Pregnancy:

- Increasing SOB at rest and on minimal exertion
- Hb maintained 6.5-7g/dl
- Sickle pains from 28/40
- Fetal growth good
- Increasingly not coping
- Plan elective LSCS 33/40 with IvIg and steroids beforehand

33/40 admitted from clinic

- In sickle crisis Hb 5.6g/dl
- Steroids and IvIg given as per plan
- 24/11/12 LSCS baby girl wt 2.2kg
- Post delivery 2unit RBC transfused (post Tx Hb 6.4g/dl)
- Post delivery hypertension: labetalol
- Wound haematoma required drainage
- Persistent PUO: abdominal lymphadenopathy
- Biopsy consistent with TB (culture pos)
- Quadruple anti TB treatment (completed June2012)

Summary

- Sickle cell anaemia
- History of chronic sickle lung and Hyperhaemolysis
- High risk pregnancy
- Successful outcome for mother and child
- Further pregnancy discouraged!

Discussion points

Management of hyperhaemolysis in sickle cell disease

Transfusion in sickle pregnancy