

Case Studies

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Case 2



Case 2 Background

- Young female patient with HbSC
- 36/40 gestation
- Presented with fever, hypoxia, chest and bone pain
- Hb 9.5g/dL
- Previously transfused 4 months earlier

O D positive
R₁r (DCe/cde)
Negative antibody
screen

Is this most likely to be a sickle cell crisis? - Q

1. Yes

2. No

3. Not sure

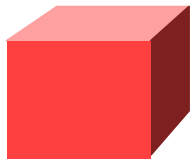


Is this most likely to be a sickle cell crisis? - A

1. Yes



2. No



3. Not sure



Treatment

- Diagnosis: sickle cell crisis
- Symptoms worsened and patient required emergency C/Section
- 2 unit transfusion post delivery
 - E-, K-
- Discharged Hb 10.1 g/dL

O D positive,
R₁r (DCe/cde)

10 days later...

Pre-delivery

- Fever, hypoxia, pain (chest; bone)
- Negative antibody screen
- Hb 9.5g/dL
- Previously transfused 4 months earlier
- Hb 10.1g/dL on discharge

10 days post delivery

- Fever, hypoxia, pain (chest; bone)
- Negative antibody screen
- Hb 6.7g/dL
- Previously transfused 10 days earlier

Is this most likely to be a sickle
cell crisis? - **A**

1. Yes

2. No

3. Not sure



Is this most likely to be a sickle cell crisis? - A

1. Yes



2. No



3. Not sure



Exchange transfusion

Diagnosis:

sickle cell crisis

- Given 6 unit exchange transfusion, E-, K-
 - Hb ↑ from 6.7 to 9.4 g/dL
 - HbS level ↓ from 48% to 21%

24 hours post exchange

- Continuing fever, pain, dysnoea
- Hb ↓ 7.1g/dL
- Spherocytes noted on blood film

Is this most likely to be a sickle
cell crisis? - Q

1. Yes

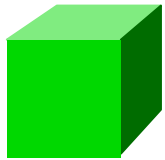
2. No

3. Not sure



Is this most likely to be a sickle cell crisis? - **A**

1. Yes



2. No



3. Not sure



Finally...

- Diagnosis: Sickle cell crisis
- Following day Hb ↓ 5.2g/dL

- Clinical review 7 days later

? Transfusion Reaction

Transfusion reaction investigation results

Pre-exchange sample

- Antibody screen confirmed negative
- DAT positive
 - IgG + C3
- Anti-Jk^a detected in an eluate

Post-exchange sample

- Antibody screen confirmed negative
- DAT positive
 - IgG + C3
- Anti-Jk^a detected in an eluate

2/2 post delivery units were Jk(a+)
4/6 units for exchange were Jk(a+)

What was the diagnosis post-exchange - Q

1. Continuing sickle cell crisis?

2. Delayed HTR?

3. Acute HTR?

4. Something else

■ 1st choice

■ 2nd choice

■ 3rd choice

■ 4th choice



What was the diagnosis post-exchange - A

1. Continuing sickle cell crisis?



2. Delayed HTR?



3. Acute HTR?



4. Something else



SHOT report and outcome

- Reported to SHOT as acute HTR
- Probably delayed HTR on readmission
- Probably also acute HTR post exchange
- Probably also sickle cell crisis



Lessons

If a patient with sickle cell disease presents with symptoms of sickle cell crisis but a disproportionate fall in Hb 3 -14 days post transfusion consider a diagnosis of DHTR

Include a DAT and eluate as part of the investigation