

#### 36 year old female

- Sickle cell disease, HbSS
- Previously well controlled on hydroxycarbamide since 25.01.12, except for interruption to treatment for 12 months Nov 2015-2016 for unsuccessful attempt at pregnancy
- Right hip avascular necrosis awaiting total hip replacement under Mr Li at King's College Hospital
- Right shoulder AVN, left knee pain waiting MRI

#### **Annual review February 2017**

- Observations: Weight 58.9 kg, height 155 cm, BP 116/58 mmHg,
   O<sub>2</sub> sats 98%, pulse 77 bpm
- Medications: Co-codamol 30/500 2 tablets qds, Penicillin V 250mg bd, Folic Acid 5mg od oral, Hydroxycarbamide 1g daily, Morphine sulphate oral solution 5mg 2-4 hourly PRN, Paracetamol 1g QDS PRN, Desunin 800iu od
- Laboratory results: satisfactory, ferritin 405, normal urine albumin:creatinine ratio
- Normal echocardiogram August 2013
- No acute admissions or transfusions for >3 years
- Stable hip pain, referred to Orthopaedic surgeon at KCH

## 04.03.16 Right AVN, on regular analgesia, referred for consideration of hip replacement surgery



# Emergency admission Elswhere March 2017

- Sudden onset headache while watching television 23.03.17
- Called LAS initial reluctance to transfer patient to hospital 'she is just hysterical'
- Emergency CT revealed subarachnoid haemorrhage
- CT Arteriogram: left distal internal carotid artery (ICA) aneurysm
- · Treatment: Cerebral fluoroscopic guided aneurysm wiring
- Continuing residual headache and nausea
  - Lumbar puncture: normal opening pressure, normal CSF results
- Red cell exchange 11.04.17 prior to discharge

#### MRA 27.03.17



#### MRA Head KCH 14.07.17



#### MRA Head 14.07.17

- No acute or established cortical infarct.
- Established left thalamic and bilateral peritrigonal white matter lacunes. Foci of right caudate head, right frontal deep white matter and left vermian microhaemorrhages. Focal irregularity at right A1/A2 junction may represent a tiny aneurysmal bleb. Generalised vascular tortuosity but proximal branches at Circle of Willis are patent with no focal stenosis, occlusion or acute dissection. No moyamoya collaterals.
- <u>Summary</u>: Established central ischaemic foci.
   No definite features of sickle maco vasculopathy

## Department of Neurosurgery Joint Neurovascular MDM – 18.07.17

- DIAGNOSIS: Ruptured left distal internal carotid artery aneurysm
- HISTORY: Subarachnoid haemorrhage (March 2017)
- PAST MEDICAL HISTORY: Sickle cell anaemia
- <u>PROCEDURE</u>: Coil embolization of left internal carotid artery aneurysm (March 2017, Queen's Hospital, Romford)
- IMAGING REVIEW: MRA head 14.07.17
- MDT DECISION: Stable treated carotid termination aneurysm. A1/A2 looks irregular but no saccular aneurysm. No other significant intracranial aneurysms. MRA imaging follow up in one year.

Laboratory results				
	Pre exchar	nge 16.10.17	Post exchange 17.10.17	
Haemoglobin g/l	87		105	
Haemoglobin S%	31.2		9.5	
PCV	0.280		0.321	
		16.10.17		
Platelets		494 x 10 <sup>9</sup> /L		
White cell count		7.88 x 10 <sup>9</sup> /L		
Neutrophils		3.85 x 10 <sup>9</sup> /L		
Ferritin		617 ug/L		
Creatinine		48 umol/L		
bilirubin		35 umol/L		
AST		35 IU/L		
GGT		54 IU/L		
ALP		59 IU/L		
Urine albumin : creatinine ratio		3.8	(normal <3.5)	

#### Currently

- 6-weekly 8 unit red cell exchange transfusion programme via peripheral access since 11.04.17
- Blood results stable
- No complaint of headache
- On waiting list for right total hips replacement surgery

#### Questions about this case:

- Should she be for indefinite exchange transfusion?
- Or be offered a switch back to hydroxycarbamide treatment?
- Was event related to sickle vasculopathy?
- Or incidental?

#### 54 year old male

- Previously known elswhere
- · Past medical History:
  - Sickle cell disease, HbSC
  - Previous stroke with right sided weakness
  - Epilepsy
  - Schizophrenia
  - Long-standing learning disability
  - Hypertension
  - Severe hearing impairment, and sickle retinopathy
  - Asthmas and heavy smoker
  - Pulmonary embolism 2015, on long-term anticoagulation

### Social history pre-admission

- Supported housing in own flat
- 3 x a day carer for all ADLs (meal prep, medication prompt and all personal care)
- Sister reported regular falls, loss of balance and impulsive due to long standing cognitive impairment
- Mobile with stick independently indoors, deteriorating for 2 -3 months, rarely leaving home, awaiting wheelchair for outdoor use

## Medications (May 2017)

- Amisulpride 100mg QDS
- Folic Acid 5mg once a day
- Penicillin V 250mg BD
- Ranitidine 300mg once a day
- Sodium Valproate 500mg BD
- Baclofen 10mg TDS
- Ibuprofen 400mg TDS
- Dihydrocodeine 60mg QDS
- Prochlorperazine 5mgs BD
- Salbutamol inhaler
- · Rivaroxaban 20mg once a day

### **Laboratory Results**

31.03.17			
Haemoglobin	135 g/L		
White cell count	12.6 x10 <sup>9</sup>		
Platelets	437 x10 <sup>9</sup>		
Potassium	4.5 mmol/L		
Creatinine	69 umol/L		
Bilirubin	10 umol/L		
Aspartate Transaminase	22 IU/L		
Albumin	39 g/L		
Ferritin	66 ug/l		
Urine protein:creatinine ratio	42		

### Admitted to DGH 05.06.17

- Found unresponsive by carers
  - Agitated and possible abdominal pain on admission
  - Initially treated as sickle VOC
  - CT and subsequently MRI head: new infarct

CT 06.06.17



#### MRI 15.06.17



## Transferred to KCH Friends Stroke Unit for rehabilitation 21.06.17

- Dense left hemiplegia, severe muscle wasting, soft tissue contractures
- Limited passive range of motion fingers/shoulder/elbow
- Unable to tolerate passive stretching, palmar protector, or bespoke fabricated hand splint
- Unable to follow verbal instructions including gestural and copying, no reliable Y/N answers, unable to read short sentences, or write beyond copying a few letters
- New left sided weakness of face and palate with swallowing problems (soft diet) and gradually improving dysarthria
- Old right sided weakness, learning disabilities, difficulty engaging in therapy, agitated

#### Laboratory results at KCH

	Admission 22.06.2017	Discharge 29.08.2017
Haemoglobin	117 g/l	124 g/l
Platelets	607 x 10 <sup>9</sup> /L	591 x 10 <sup>9</sup> /L
White cell count	13.68 x 10 <sup>9</sup> /L	11.30 x 10 <sup>9</sup> /L
Neutrophils	8.07 x 10 <sup>9</sup> /L	6.77 x 10 <sup>9</sup> /L
Creatinine	82 umol/L	58 umol/L
Bilirubin	13 umol/L	11 umol/L
AST	26 IU/L	25 IU/L
GGT	153 IU/L	116 IU/L
ALP	95 IU/L	116 IU/L

#### Investigations at KCH

CT Angiogram intracranial and neck vessels 23.06.17: Normal appearances of internal carotid arteries with no stenosis, dissection or aneurysm seen. Vertebral arteries co-dominant, with no focal stenosis identified. Normal configuration of the Circle of Willis. Established right MCA territory infarct and bilateral lacunar infarcts.

No imaging features of sickle vasculopathy within cervical and proximal intracranial arteries

#### Further investigations at KCH

- Echocardiogram 23.06.17: Normal LV size with mild concentric LVH. Normal LV systolic function. No significant valvular abnormalities. No obvious intracardiac mass/shunt seen. LVEF 57 %.
- 24 hr ECG 23.06.17 Sinus rhythm with occasional ectopics
- Carotid Doppler 22.06.17: Carotid and vertebral arteries widely patent with no significant stenoses
- Thrombophilia screen 23.06.17: normal

#### Management at KCH

- Intensive stroke rehabilitation at King's Friends Stroke Unit 21.06.17-09.09.17
- 14.07.17 Botox injections to treat spastic contractures
- Not exchange transfused as not on regular transfusion program despite previous CVA, acute CVA
   3 weeks ago, unable to consent, unclear clinical benefit
- Apixaban re-started in view of previous PE; history of ischaemic infarcts of probable embolic origin

#### Clinical course

- Able to make basic needs known verbally, orientated to place and person
- Ongoing need for assistance for all activities of daily living, including personal care, toileting, medication
- Marked improvement in mobilisation and speech but unstable gait, long-standing learning disabilities, poor compliance and tendency to self-neglect
- Discharged to nursing home near sister's address

#### Medications on discharge:

- Cholecalciferol (Fultium-D3) 800 units od
- Folic Acid 5 mg od
- Amisulpride 100 mg qds
- Ranitidine 300 mg od
- · Penicillin V 250 mg od
- · Sodium Valproate EC 700 mg bd
- Salbutamol Aerosol Inhaler 100mcg 1 to 2 Puffs PRN
- Nicotine Inhalator 15mg (Nicorette) PRN
- · Hyoscine Butylbromide 20 mg qds
- · Lidocaine 5% Medicated Plaster every 24 hours
- · Paracetamol 1000 mg qds
- · Baclofen 10 mg tds
- · Apixaban 5 mg bd

#### Questions about this case:

- Was he a candidate for acute and ongoing red cell exchange?
  - How would this be facilitated (poor compliance, limited understanding, dislike of needles)?
- Is there a place for hydroxycarbamide therapy?
  - (but poor compliance and erratic clinic attendance, need for regular poorly tolerated blood tests, limited evidence for benefit)?
- Now cared for in stable nursing home environment
  - perhaps compliance will improve

