Multimodality Neuroimaging in Sickle Cell Anaemia - Disease Features in Children and Adults

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Learning objectives

To illustrate neuroimaging findings in sickle cell anaemia in children and adults using multiple imaging techniques including transcranial Doppler (TCD), CT/CTA, MRI/MRA, conventional angiography and to outline the role of advanced techniques such as perfusion MRI including continuous arterial spin labelling (CASL).

In certain geographic areas, radiologists may infrequently encounter sickle cell patients. This exhibit aims to provide an introduction for those less familiar with the disease and its typical neuroimaging findings. Beyond that, a more comprehensive overview will be given featuring a wider range of appearances and how these can be assessed with different imaging modalities. Possible pitfalls and diagnostic problem situations will also feature.
Background

Sickle cell anaemia (SCA) is an inherited disorder affecting haemoglobin molecules within red blood cells. In sickle cell individuals, formation of abnormal haemoglobin leads to rigid deformation of erythrocytes causing impairment of their ability to pass through small blood vessels and eventually red cell death resulting in haemolytic anaemia.

On a positive note, sickle cell disease is associated with increased resistance against malaria (plasmodium falciparum infection).

Sickle cell is the most common serious genetic disorder in England occurring in over 1 in 2000 live births [1]. The disease is most common amongst descendants of African and Afro-Caribbean heritage and in the rest of Europe is prevalent amongst ethnic groups such as those from Mediterranean countries, Turkey, and the Arabian Peninsula. Furthermore, SCA may be encountered in the population of central and South America.

Pathophysiology

In the normal individual, each haemoglobin molecule consists of 4 chains, namely 2 alpha and 2 beta chains. In sickle cell, a point mutation on the short arm of chromosome 11 causes thymine to be substituted by adenine on position 6. This results in the production of an abnormal beta haemoglobin chain (HbS), in which glutamic acid is replaced by valine.

In the homozygous patient, both sickle cell genes code for an abnormal beta haemoglobin chain (HbSS), per definition leading to classic sickle cell anaemia (SCA) and a more severe course of disease. Rarer disease variants, in which one HbS beta chain combines with another abnormal beta chain of a different mutation, e.g. HbSC, HbS-thal are termed sickle cell disease (SCD). For the purpose of this poster, the terms sickle cell anaemia and sickle cell disease may be synonymously used to describe clinically symptomatic individuals with two abnormal beta chains.

The heterozygous variant of the disease is termed sickle cell trait (HbSA), whereby one abnormal and one normal beta chain are produced, resulting in a more benign variant of the disease in the vast majority of affected individuals. Nevertheless, identification of even asymptomatic sickle cell trait carriers has implications for genetic counselling [2].

Under certain conditions, particularly deoxygenation, HbS forms polymers with other haemoglobin molecules leading to aggregates within the red blood cell. As a result, the erythrocyte loses its normal elasticity and deforms into its characteristic sickle shape.
Upon reoxygenation, polymerisation resolves and erythrocytes readopt their normal shape. However, prolonged or repeated sickling may lead to irreversible deformity and damage of the cell including increased permeability of the cell membrane. Finally, abnormal red blood cells are removed from the circulation resulting in haemolytic anaemia. It is well known that in sickle cell patients the anaemia tends to be better tolerated than vascular complications of the disease.

In SCA the rigidly distorted erythrocyte can no longer pass normally through blood vessels and in addition adheres more easily to vascular endothelium resulting in vascular congestion, occlusion and ischaemia. Sickle cell anaemia may affect any organ system, whereby recurrent episodes of vaso-occlusion and inflammation result in progressive organ damage including infarction. Amongst other sites of involvement, neurological manifestations of sickle cell anaemia are common.

**Neurovascular sickle cell anaemia**

Approximately 25% of patients with SCA will have a neurologic complication over their lifetime, and these may occur even in early childhood [2]. Neuroimaging abnormalities of sickle cell disease are not infrequently seen in a multicultural population and they can pose diagnostic challenges.

Presentations include silent abnormalities with an objective imaging correlate, for example asymptomatic arterial stenosis or previous silent infarction, which may occur in children and adults. Ischaemic strokes in sickle cell patients are common and typically present with a focal neurological deficit, especially hemiparesis, hemisensory deficits, focal seizures or visual disturbance [3]. These strokes are usually, but not always, associated with intracranial vasculopathy [4].

Neurological involvement in sickle cell disease may present both in subtle and overt neuropsychological deterioration [1]. There is evidence that psychological screening may identify patients those who have suffered silent infarcts [6].

Over time and with progressive occlusion of the main intracranial arteries a so called "Moya Moya" (Japanense : puff of smoke) appearance may be observed, which describes the formation of numerous tiny collateral vessels throughout the brain.

Severe headache, generalised seizures and reduced consciousness are more common in intracranial haemorrhage, which may occur due to haemorrhagic transformation of ischaemic brain parenchyma [5].

Furthermore, sickle cell vasculopathy is associated with an increased risk of developing intracranial aneurysms, and thus secondary haemorrhage. Aneurysms are multiple in the majority (57 %) of sickle cell patients and more often originate from the posterior
circulation (30%) than in the normal population (5-14%) [2]. The exact cause of large vessel narrowing is unclear but is thought to be mediated by endothelial injury/activation by adhesion of abnormal cellular elements which eventually results in damage of the muscularis resulting in narrowing or aneurysm formation.

Although the vast majority of patients present with ischaemic complications, other differential diagnoses for intracranial pathologies should be born in mind, particularly in view of the increased susceptibility of sickle cell patients to infection.

Bony pathology such as infarction, osteomyelitis and extramedullary haematopoiesis may also be observed on cranial imaging.
Imaging findings OR Procedure details

Our patients

Guy's and St. Thomas Hospitals NHS Foundation Trust is a central London teaching hospital caring for a large multiethnic community with over 900,000 patients contacts per year. Sickle cell tertiary services including diagnostic radiology are provided on a 24-hour basis.

In our practice, we assess neuroimaging in sickle cell patients of all ages. With routine sickle screening in place, children may be referred at a very early age for emergency or routine imaging.

In accordance with the National Standards and guidelines for the management of sickle cell disease in childhood, annual routine TCD studies are offered to sickle cell disease patients from the age of 2 years to identify children at risk of stroke. We are one of two nationally commissioned trusts in the country providing transcranial Doppler ultrasound (TCD) training to healthcare professionals [1].

The modalities

Computed tomography

CT imaging of the brain nowadays is readily available throughout the UK and Europe making it the most frequently requested modality for patients presenting with acute neurological conditions. It permits rapid assessment, although movement artefact can still be a problem in agitated patients.

Bearing radiation protection and resources in mind, adults who present with acute neurological symptoms will more frequently undergo CT imaging of the brain as the initial modality, although diagnostic cranial CT may also be performed in children in an emergency or if detailed bony evaluation is required.

Non-contrast CT is often the first investigation to exclude major intracranial pathology including CVA, but may miss early ischaemic strokes and smaller lesions. It is excellent at detecting acute blood, and as such remains the imaging modality of choice to assess for acute intracranial haemorrhage.
Next to limited sensitivity and specificity, one drawback of this modality is its use of radiation. Whilst a single study using a multi-slice spiral scanner does not result in a very large effective dose, repeat scanning of patients with chronic or recurrent conditions and imaging of children due to their increased radiosensitivity should be thought over.

Contrast enhanced CT may provide a more accurate assessment of certain pathologies including neoplastic and inflammatory conditions, but often has less diagnostic power than MRI.

**CT Arteriography (CTA)**

CT angiography (CTA) can achieve high quality assessment of intra and extracranial arterial vessels. Typically, this involves scanning the extracranial arteries of the neck and intracranial arteries including the Circle of Willis. Following contrast injection and using a contrast-tracking technique, a block of images is acquired within seconds and allows evaluation of the scanned arteries for stenosis, occlusion and dissection. CTA may also identify intracranial aneurysms depending on their size.

**Dynamic Perfusion CT (PCT)**

In some centres, dynamic CT perfusion imaging has been introduced for the assessment of suspected early stroke. Its predominant role lies in the rapid detection of acute thromboembolic ischaemic stroke aiming at reversibility via thrombolysis [7]. There is currently limited evidence to support the use of thrombolysis in sickle cell patients with acute stroke, possibly because of the chronic vasculopathic character of the disease. However, performing perfusion imaging may nevertheless be of value in the early evaluation of the extent of ischaemia.

**MRI**

Magnetic resonance imaging provides images with high inherent soft tissue contrast. A typical stroke protocol includes multiple sequences (T1, T2, FLAIR, DWI, ADC) to detect acute and chronic abnormalities.
FLAIR (fluid attenuation inversion recovery) imaging uses suppression of fluid signal to make small lesions more conspicuous and is particularly helpful to identify lesions close to the ventricles or brain surface.

DWI is a physiological MRI technique, which detects diffusion of water molecules and uses this information to form an image. Acute brain parenchymal infarction leads to cellular swelling and cytotoxic oedema with reduction of the extracellular space. The result is restricted diffusion of water molecules. Restricted diffusion leads to bright signal on DWI images. Diffusion weighted imaging is used to evaluate for acute ischaemic stroke and will demonstrate abnormalities earlier than CT.

An ADC map (apparent diffusion coefficient) can be calculated. In acute ischaemia, high DWI signal (i.e. restricted diffusion) corresponds to low signal on the ADC map.

MR imaging may be problematic in claustrophobic patients and is contraindicated under certain circumstances, e.g. if the patient has a pacemaker. In children, depending on age and the individual, MRI may require a general anaesthetic. Scanning time is longer than for CT and increases with acquisition of multiple sequences increasing the risk of movement artefact.

Magnetic resonance angiography (MRA)

MRA allows non-invasive assessment of the intra and extracranial arteries. This technique uses a "time-of-flight" (TOF) sequence, in which flowing blood is detected and used to form an image. This may be done as a 2D or 3D technique and as opposed to a traditional post contrast MRA sequence has the advantage that a contrast injection is not required. Due to reduced resolution than conventional angiogram, TOF MRA is limited in the evaluation of smaller intracranial arterial branches [7].

On TOF images, flowing blood appears bright and background tissues are suppressed.

Continuous Arterial Spin Labelling (CASL)

This is a relatively new functional MRI technique, which assesses cerebral blood perfusion without the requirement for intravenous contrast administration. As such its use is particularly suited to children or when intravenous access cannot be achieved [8].

TCD
Transcranial Doppler ultrasound (TCD) has been used since 1986 in children with sickle cell disease [3]. TCD is now an integral part of the UK national standard protocol for screening children with sickle cell disease annually from the age of 2 [1].

This type of scan is performed through the temporal bone using conventional Doppler, often in combination with colour Doppler. The colour mode assists in identification of smaller vessels, although it is not suitable to perform quantitative measurements.

TCD allows measurement of the flow velocity in the Circle of Willis branches, aiming to detect either focally increased velocity as a sign of localised stenosis or generally increased velocity indicating elevated cerebral blood flow [3]. TCD is particularly sensitive in the detection of extracranial arterial stenosis, however, the extracranial vessels tend to be less affected in sickle cell patients.

Measuring technique may slightly vary at different institutions, but common measurements may include the time averaged maximum mean velocity (TAMM) for the middle cerebral artery, anterior cerebral artery, bifurcation, distal interior carotid artery and posterior cerebral artery.

One of the main limitations of TCD, like any ultrasound, is operator dependence. Additionally, training facilities may be unavailable at some institutions.

**Conventional angiography**

This modality is a highly accurate but invasive method to visualise the intra and extracranial arterial vasculature. In sickle cell patients, it requires a rigorous protocol for hydration and reduction of HbS to < 20 % in order to avoid sickling and thus stroke.

Although in most case non-invasive methods may nowadays be preferred, invasive angiography still retains a role in problem situations, for the delineation of intracranial aneurysms, in providing a road map prior to revascularisation procedures and importantly in the context of endovascular intervention.

**Our cases**

Examples of typical and atypical neuroimaging features in sickle cell disease together with their diagnoses are given below with the corresponding images listed in the sidebar. Please note that case 1 features two movies, which may take a short while to load.
Case 1

A 2 year old with sickle cell disease was admitted with acute left sided weakness. A CT was performed at 4 hours and demonstrated no abnormality.

The patient was subsequently referred for an MRI, see Fig. 1 to 7.

Case 2

A 12 year old presented with acute left hemiplegia and facial weakness. CT imaging was performed at day 1 and day 2. MRI imaging was performed at presentation at 2 years later with MRA and MR perfusion (CASL) images also performed as part of the patient’s follow up (see Fig. 8 to 14). The diagnosis in this case is given in Fig. 14.

To revise the major vascular territories of the cerebral hemisphere, Fig. 15 provides examples of territorial infarction in different vascular territories.

Case 3

A 9 year old child presented with acute right sided weakness in 2004. An MRI was performed initially (at another institution) and another MRI was carried out 2 years later. There had been no new clinical events in the interval period. Images and the diagnosis are shown in Fig. 16 to 19.

Case 4

An 8 year presented with a gradual deterioration in school performance. No acute neurological signs were elicited. Characterstic findings an the patients diagnosis can be seen in Fig. 20 to 23.

It should be emphasized that in sickle cell disease, even advance vasculopathy can present with cognitive impairment only instead of focal neurological signs. In a child with
known sickle cell disease, any unexplained decline in school performance should prompt further assessment.

Case 5

A 7 year old with known previous infarcts developed severe headache, vomiting and drowsiness. A CT was performed as the initial investigation, see Fig. 24 to 28. Of note, the CT angiogram shown in Fig. 26 demonstrates vasculopathy, but no aneurysm.

For the same patient, repeat imaging was performed between the years 2005 and 2009 and demonstrated marked progression of vasculopathy with a Moya Moya appearance (Fig. 29 to 32).

Case 6

A 31 year old female had had previous strokes aged 4 and 26, which left her with residual right sided motor weakness. She acutely presented with sudden onset headache.

She underwent CT, MRI and MRA imaging (Fig. 33 to 36). Conventional angiography was not performed at the time.

Case 7

An 8 year old developed sudden headache, neck stiffness and vomiting. CT and CT angiography images are shown in Fig. 37 and 39 with the diagnosis given in Fig. 42.

Case 8

A 5 year old patient was being treated for osteomyelitis and developed acute left sided weakness and ataxia, see Fig. 40 to 42 for imaging findings and diagnosis.
Fig. 0: Case 1. T2/FLAIR images at the level of the corona radiata demonstrating hyperintense abnormalities involving the cortex and white matter in the right parietal lobe and in the watershed regions of both frontal lobes.

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Fig. 0: Case 1. T2/FLAIR images at the level of the centrum semiovale demonstrating hyperintense abnormalities involving the cortex and of the right parietal lobe and linear abnormalities in the watershed regions of both frontal lobes

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Fig. 0: Case 1. DWI image and ADC map corresponding to image 1 showing restricted diffusion in the right parietal lobe abnormality indicating acute infarct. The frontal lobe changes represent chronic watershed infarction.

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Fig. 0: Case 1. DWI image and ADC map corresponding to image 2 showing restricted diffusion in the right parietal lobe abnormality indicating acute infarction. The frontal lobe changes represent chronic watershed infarction, typically seen in sickle cell disease.

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Fig. 2: Case 1. 2D-TOF movie. Selected source images from a TOF MRA series showing absence of flow signal from the right ICA.

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Fig. 1: Case 1. 3D Time-of-flight MRA-MIP images showing absence of flow signal from the right ICA, with reconstitution of the ACA and MCA via the circle of Willis.

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Diagnosis

- Acute R MCA territory branch infarct
- Background old frontal watershed infarcts
- Proximal R ICA narrowing/occlusion

**Fig. 0:** Case 1. Salient features and diagnosis.

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**Fig. 0:** Case 2. Early unenhanced CT showing subtle loss of grey-white differentiation in the right MCA territory.

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**Fig. 0:** Case 2. Subsequent CT showing a clearly defined large MCA territory infarct with substantial mass effect and midline shift. Not the enlarged left lateral ventricle, due to mass effect on the foramen of Monro.

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**Fig. 0:** Case 2. Early MRI - T2-weighted image showing gyral swelling and hyperintensity in the right MCA territory.

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Fig. 0: Case 2. Follow up MRI shows evolution of the infarct into a large area of encephalomalacia. Note the silent small left basal ganglia infarct and the right-sided decompressive craniotomy.

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**Fig. 0:** Case 2. TCD showing low velocities in the right terminal ICA (no flow signal on MRA) and elevated velocities in the left terminal ICA (moderate to severe stenosis on MRA)

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Fig. 0: Case 2. FLAIR and CASL perfusion images showing reduced perfusion in the right hemisphere corresponding to the infarct but preserved perfusion in the left hemisphere.

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Diagnosis

- Large evolving acute right MCA infarct with subsequent encephalomalacia (requiring decompressive craniotomy)
- Small silent infarcts in the left anterior circulation
- Clinically some recovery, walks independently with foot orthosis
- EDAS procedure (Encephalo-dural-arterial-synangiosis) on left side recently

Fig. 0: Case 2. Summary points

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**Fig. 0:** Large vessel infarction - images showing infarcts outlining the three major vascular territories of the cerebral hemispheres.

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Fig. 0: Case 3. T2-weighted MRI showing evolution of a subacute left MCA branch infarct. Note marrow replacement and expansion that is often seen in sickle cell disease.

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Fig. 0: Case 3. T2-weighted MRI images showing evolution of left MCA infarction and cortical atrophy in the right hemisphere indicating silent infarction in the right anterior circulation.

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Fig. 0: Case 3. TOF MRA showing progressive narrowing and occlusion of the terminal internal carotid arteries bilaterally. The terminal ICAs and MCAs could not be identified on the TCD examination.

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**Fig. 0:** Case 3. Diagnosis

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Fig. 0: Case 4. Small old basal ganglia infarcts. Note the small tortuous ‘net-like’ vessels in the CSF spaces at the base of the brain and basal ganglia (marked and circled), corresponding to Moya Moya collaterals.

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Fig. 0: Case 4. FLAIR and CASL perfusion MRI images. Bilateral deep white matter old infarcts in the centrum semiovale (note the linear vertical orientation that is typically seen in deep watershed zones infarcts). Perfusion images indicate reduced cortical blood flow in the anterior and posterior watershed territories, which is not evident on the structural FLAIR images.

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**Fig. 0:** Case 4. MRA images showing occlusion of the right ICA with extensive Moya Moya collaterals, and narrowing of the left ICA with fewer collaterals.

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Fig. 0: Case 4. Diagnosis

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Fig. 0: Case 5. CT images showing acute intraventricular haemorrhage with blood in the lateral and in the fourth ventricles and old bifrontal infarcts.

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Fig. 0: Case 5. CT images showing acute intraventricular haemorrhage with blood in the lateral ventricles and old bifrontal infarcts.

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**Fig. 0:** Case 5. CTA showed severe right ICA narrowing but no aneurysms.

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Fig. 0: Case 5. Summary and salient points

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**Fig. 0:** Case 5. FLAIR images showing bilateral watershed infarcts, more on the right side. Note the hyperintensity within the sulci, slow flowing engorged pial vessels and thickened arachnoid membranes, seen in Moya Moya. These are less prominent on the 2009 study; whilst this may partly be related to improved scan quality, it is also thought to be an indicator of successful revascularisation.

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Fig. 0: Case 5. MRA showing progressive vasculopathy with narrowing of the terminal ICA and proximal MCA, severe on the right side

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Fig. 0: Case 5. TCD showing elevated right MCA velocities corresponding to severe narrowing on the MRA

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**Fig. 0:** Case 5. Follow up TCD shows progressive vasculopathy with no flow detectable in the right MCA on the TCD, corresponding to absent flow signal on the MRA.

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**Fig. 0**: Case 5. DSA showing lateral views of selective ICA injection showing progressive narrowing of the terminal ICA.

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Fig. 0: Case 6. CT showing acute intraventricular haemorrhage

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Fig. 0: Case 6. MRI showing signal characteristics of acute/early subacute intraventricular haemorrhage. Note high signal on T1, low signal on T2 and marked low signal on T2* images (T2* images should always be performed if there is a suspicion of intracranial haemorrhage because blood products become more apparent due to susceptibility effects)

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Fig. 0: Case 6. MRA shows left terminal ICA narrowing with few Moya Moya collaterals

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Fig. 0: Case 6. Diagnosis

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Fig. 0: Case 7. CT shows subarachnoid haemorrhage, mainly in the sylvian fissures and anterior interhemispheric fissure. There is also bilateral anterior cerebral artery infarction.

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Fig. 0: Case 7. CTA - MIP and 3D volume rendered images show an anterior communicating artery aneurysm.

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Fig. 0: Case 7. Diagnosis

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**Fig. 0:** Case 8. CT and MRI (DWI and ADC) showing acute left superior cerebellar artery territory infarct. Posterior circulation infarction is unusual in sickle cell disease, and other pathologies such as thrombo-embolism and dissection should be excluded.

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**Fig. 0:** Case 8. MRA - MIP showing occluded right vertebral artery

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Fig. 0: Case 8. Diagnosis

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Conclusion

Neuroimaging of sickle cell disease remains a complex topic. Knowledge of relevant imaging appearances and disease patterns is crucial, whilst the choice of appropriate imaging modalities will facilitate accurate diagnosis and management. Particularly in atypical disease or following intervention the use of multiple modalities may be helpful.
In Summary

- Three major intracranial abnormalities seen in SCD
  - Vasculopathy – Terminal ICA, proximal CoW stenosis, Moyamoya
  - Infarcts - Acute territorial infarct, Silent anterior watershed infarcts
  - Haemorrhage
- CT, fast, best for acute haemorrhage
- DWI best to detect acute infarcts
- FLAIR useful for better visualisation of small infarcts
- T2* images useful to detect haemorrhage
- MRA useful to assess large vessel vasculopathy
- CTA, DSA for selected cases (pre-surgical / bleed)
- TCD best for screening for vasculopathy
- Perfusion MRI may provide haemodynamic information

Fig. 0: Summary

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References


