AFRICAN JOURNAL OF MEDICINE and medical sciences

VOLUME 34 NUMBER 2 JUNE 2005

ISSN 1116—4077

Afr J Med med Sci (2005) 34, 115-118

Computed tomographic pattern of stroke in children with sickle cell anaemia in Ibadan

AO Ogunseyinde[1], MO Obajimi[1] and OJ Fatunde[2]
Departments of Radiology[1] and Paediatrics[2], University College Hospital, Ibadan, Nigeria

Summary
We present the findings in 14 paediatrics patients with SCA who had CT examination at the University College Hospital Ibadan on account of stroke between 1993 and 2000. There were 8 female and 6 male with a mean age of 11.25 years and SD of 3.66. Cerebral infarction was the most common finding occurring in 57% of the patients while intracerebral bleed was seen in 21%. Two patients had a mixed lesion and atrophy was seen in one patient. Five patients (36%) had their lesion on the right hemisphere while eight (57%) had their lesions on the left side. The frontal and parietal lobes were mainly affected. Prompt CT screening of the brain in a child with sickle cell anaemia who presents with symptoms and signs suggestive of stroke can help identify the particular type of lesion and this may influence mode of therapy given as well as prognosis

Keywords: *Sickle cell anaemia, stroke, cerebral infarction, intracerebral bleed, computed tomography*

Résumé
Nous presentons les données de quatorze enfants drepanocytaire anemiés avec l'analyse du CT au Centre Universitaire Hospitalier de l'Universited'Ibadan entre 1993 a 2000. Ils étaient 8 fille et 6 garcon avec la moyenne d'age de 11.25 +- 3.66 ans. L'infarction cérebrale était la donnée plus commune chez 57% des patients , alors que l'hémorrahgie intracérebrale était observée chez 21%. Deux patients avaient une lésion mixe et l'atrophie était observée chez l'un des patients. 36% des patients avaient leur lésion sur l'hémmisphere droit et 56% (8) sur l'hémisphere gauche. Les lobes frontal et pariétal étaient plus affectés. Le depistage précis du CT du cerveau aux enfants drepanocyatire anemiés et ayant les symptomes et signes suggestive du shock peut aider à l'identifier le type de lésion et ceci pourrait influencer le mode de la thérapie bien que le prognostie.

Introduction

Stroke is defined as an acute focal neurologic dysfunction lasting more than twenty-four hours. It is one of the most devastating complications of sickle cell anaemia (SCA) and approximately 7% of children with SCA experience stroke by the time they are 14years of age [1,2].

Correspondence: Dr A O Ogunseyinde, Department of Radiology, University College Hospital, Ibadan, Nigeria.

Stroke can occur during an acute vaso-occlusive pain episode or with acute anaemic crisis. Affected patients usually have clinically recognized symptoms most commonly hemiparesis or monoparesis, aphasia and seizures; coma is rarely a presenting feature [3,4]. It is important to identify patients with cerebrovascular abnormalities, so that appropriate therapy can be instituted early thereby averting devastating complications. In developing countries like Nigeria, Computed Tomography (CT) is the imaging modality in patients with neurologic deficit and a working diagnosis of cerebrovascular disease when it is affordable, as it allows prompt, precise and accurate determination of the cerebral insults [5]. Magnetic resonance imaging and magnetic angiography are now preferred modalities for continued evaluation of sickle cell patients in developed countries because of their higher sensitivity in detecting cerebral injury in SCA [6] but these are not yet available in our hospital.

We present the CT findings in 14 paediatric patients with sickle cell anaemia who presented with stroke at the University College Hospital, Ibadan.

Material and method

We reviewed the CT scan of 19 patients with sickle cell anaemia referred for CT examination on account of neurologic deficit from a possible cerebrovascular disease (stroke). The CT examination was done on a GE9000 machine, using a 5mm contiguous cut in the posterior fossa and 10mm contiguous cuts through the rest of the brain. Unenhanced examination was done when intra-cerebral haemorrhage was demonstrated. The clinical and biomedical information was retrieved from the case-notes. The films were analysed for the type and site of the lesions

Results

A total of 19 patients with sickle cell anaemia and stroke were referred for CT examinations within the period 1993-2000, but only 13 of them fell within the paediatric age group and they had a total of 14 examinations. One patient had had two episodes of stroke, at the age of 12years and 14 years. There were 8 male and 6 female. Their ages ranged between 4years and 16years with a mean of 11.03years and standard deviation of 3.66 (Table 1)

Table 2 shows the findings on computed tomography. Cerebral Infarct (Fig1) was most common occurring in 8 (57.1%) of the patients, while 3(21.2 %) had intracerebral bleed (Fig2).

Table 3 shows the distribution of the lesions. While five patients (36%) had their lesions in the right hemisphere,

8 (57 %) had their lesions on the left side and they were mainly in the frontal and parietal lobes. Only in one patient was the caudate nucleus involved.

Table 1: Age distribution of the sickle cell patients with stroke

Age Group Years	Frequency	Percentage
1-4	1	(7.1)
4-9	4	(28.6)
10- 15	6	(42.9)
16	3	(21.4)

Table 2: Findings on computed tomography

Lesion	Frequency
Infarct	8 (57.2%)
Haemorrhage	3 (21.4%)
Mixed Lesion	2 (14.3%)
Atrophy	1 (7.1%)
Total	14

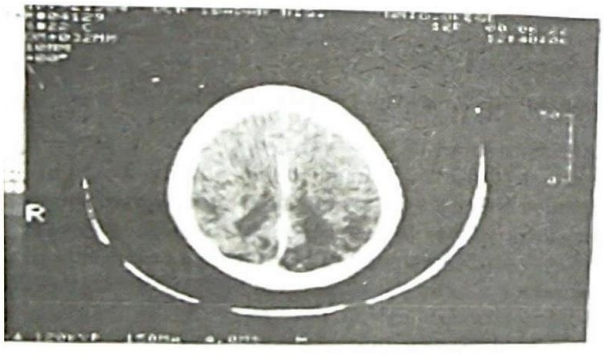

Fig. 1 (a): Computerised tomography showing hypodense area of chronic infarction in the occipital lobes (a), left fronto-occipital region (b)

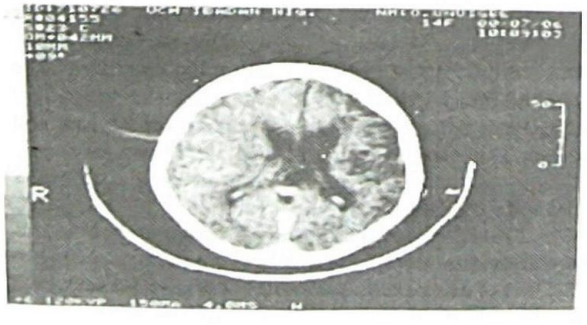

Fig. 1 (b)

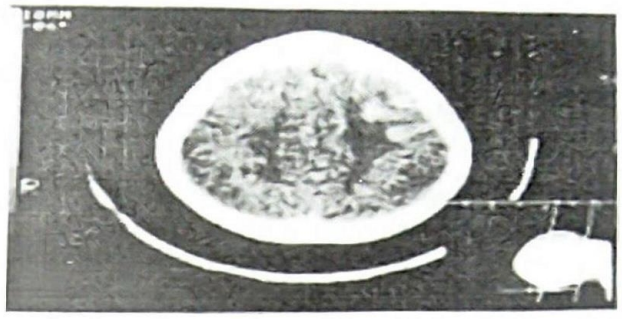

Fig. 2: Un-enahnced CT brain sowing hyper-dense area of Intra-cerebral bleed in the left fronto-parietal region.

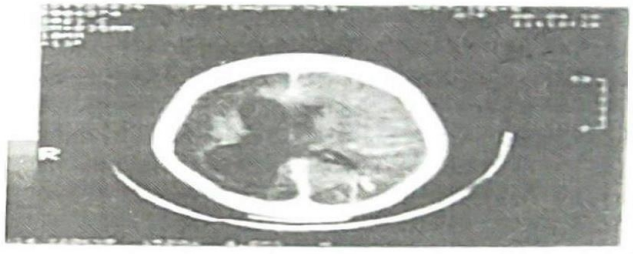

Fig. 3: CECT showing atrophy of the right cerebral hemisphere. Note the widened CSF space, reduction of brain mantle and dilation of the body of the right lateral ventricle.

Table 3: CT Findings—showing types of lesion and site of Lesion

Type of lesion	Frequency*	Site
1. Mixed lesion	2	LT Parietal lobe
2. Infarct	2	LT Frontal lobe
3. Infarct	1	RT Fronto-parietal
4. Infarct	1	RT Parieto-occcipital
5. Infarct	1	RT Frontal lobe
6. Infarct	1	RT Parietal lobe
7. Infarct	1	LT Parieto-occipital
8. Infarct	1	LT Fronto-temporal
9. Infarct	1	LT Occipital
10. Infarct	1	LT Fronto-parietal
11. Haemorrhage	1	RT Frontal
12. Haemmorrhage	1	LT Caudate nucleus
Total	14	

**One patient had 2 separate studies*

Discussion

Cerebro-vascular accident is a major complication of sickle cell disease during childhood and up to 15% of all sickle cell disease patients suffer cerebral events [2,6,8,9]. It is suggested that age is one of the risk factors for the development of stroke in patients with SCA and about 80% of patients with cerebral complications were 15years of age or less[4,8] and about 70% will have a second stroke within 36 months of the first episode [8]. The mean age in this

series is 11.25 years and 11of the14 patients (78.6 %) are below 16years; but repeat episode was rare, occurring in only one patient, two years after the initial episode. This may not reflect the true incidence of repeat episode, even though stroke is relatively uncommon in Nigerians with SCA [10]. It may also be related to the fact that many of those with stroke are unable to afford the cost of the CT examination.

The types of cerebral vascular lesions that may occur with SCA include ischaemic or haemorrhagic infarction, intracerebral bleed and subdural haematoma [8,11,12] and ischaemic infarction is the most common cerebrovascular episode. Cortical venous and sinus thrombosis have also been reported [13,14]. These are well demonstrated on CT examination because of its high sensitivity (100%) in detecting haemorrhage and 90% for subarachnoid haemorrhage.

Stroke in children with SCA has been attributed to occlusion of large and small vessels, cell sludging, and distal field insufficiency which may involve the circle of Willis primarily or major bifurcation of both internal carotid artery [6,15], and to intimal hyperplasia within large cerebral arteries and small vessels proliferation in the basal ganglia [7,12].

Genetic factors such as β-thalaseamia, high foetal haemoglobin may also influence the risk of stroke in children with SCA [4,16] and patients with sickle cell anaemia have a higher prevalence of stroke and there is no significant sex difference. All our patients had haemoglobin SS.

Cerebral infarction was the most common lesion in our study occurring in 57 %. This is similar to the findings of other workers [6-9,11,17]. Both cerebral hemispheres can be involved and the most common areas are the frontal, parietal and temporal lobes; lesions are rare in the occipital and cerebellum [17,18]. In our series, the frontal and parietal lobes were mostly involved and most lesions were on the left side. This is not surprising, as most people have left cerebral dominance.

Infarction in SCA, involve both the cortex and deep white matter. The location and the volume of cerebral infarction reflect the clinical severity of the disease and are important in defining the type and magnitude of cognitive sequelae in childhood stroke [2,17,18]. Silent infarcts have also been reported occurring in about 15% of patients with SCA [4,6,18]. This usually results from either vaso-occlusion within cerebral vessels or transient reduction in the perfusion pressure known to accompany a hypoxic or hypovolaemic episode, which occurs in aplastic or sequestration crisis or seizures. Magnetic Resonance Imaging (MRI) is more sensitive in demonstrating silent infarcts in patients with SCA. All the scanned patients had overt signs of stroke.

Intracerebral haemorrhage as the cause of stroke in patients with SCA is less common than infarct and occurs in 25% of patients, while subarachoid haemorhage occurs in less than 2% of patients [2,7,8]. These are usually due to rupture of cerebral artery aneurysms and it is more common in adult patients. Intracerebral haemorrhage was found in 21.4% of our patients and there was no case of subarachoid haemorrhage.

Chronic infarcts are characterized by cystic cavitations, which are better demonstrated on MRI [6,19]. These infarcts latter regress and become less obvious or lead to atrophy as a result of neuronal death that accompanies an infarct. Focal atrophy manifests by dilation of adjacent sulci and ventricles. Cerebral atrophy was the only finding in one of our patients.

Prompt CT screening of the brain in children with SCA who present with symptoms and signs suggestive of stroke can help identify the particular type of lesion and this may influence mode of therapy given as well as prognosis.

The ultimate advantage of brain imaging in patients with SCA will be to identify patients with cerebrovascular disease prior to development of the first stroke. This may be done with the use of Magnetic Resonance Angiography (MRA) or intracranial Doppler sonography [20]. Such at risk children may then be offered intervention regimens such as blood transfusion or hydroxyurea therapy or even bone marrow transplant, all of which may prevent development of stroke [21-22].

References

1. Armstrong FD, Thompson RJ, Wang W *et al.* Cognitive function and brain resonance imaging in children with sickle cell disease. Paediatrics 1996; 97: 864-870
2. Balkaran B, Char G, Morris JS *et al.* Stroke in a cohort of patients with homozygous sickle cell disease. J. Paed 1992; 20:360-366
3. Portnoy BA, Heroin JC. Neurological manifestation in Sickle Cell Disease and a review of the literature and emphasis on the prevalence of hemiplegia. Ann. Int. Med. 1992; 72: 643-652
4. Driscoll MC, Hurler A, Styles L, McKie V, Files B *et al.* Stroke in siblings with sickle cell trait. Blood 2003; 101: 2401-2404
5. Obajimi MO, Nyame PK, Jumah KB, and Weredu EK. Spontaneous intracranial haemorrhage: Computer Tomographic pattern in Accra. W.A.J.M 2002; 21: 60-62
6. Steen RG, Emudianughe T, Hankins GM, Wynn LW, Wang WK *et al.* Brain Imaging findings in pediatric patients with sickle cell disease. Radiol.2003; 228: 216-25
7. Dickerhoff R, Pongratz E, Scheel-Water HG. Cerebral Infarcts and haemorrhage in patients with sickle cell disease. Klin Pediatr 1994; 206: 381-384
8. Zakara M, Al Hawsawi. Stroke among sickle cell disease patients in Saudi Arabia. Annals of Saudi Med. 1998; 18: 472-474

9. Obama MT, Dongmo L, Nkemayin L, Mbede J, Hagbe P. Stroke in Children in Yaounde, Cameroon Indian Pediatr 1994; 31: 791-795
10. Adeloye A, Odeku EL. The Nervous System in Sickle cell disease. J. Med. Med.Sci. 1970; 1: 33-48
11. CJ Moran, MJ Siegel, MR DeBaun. Sickle Cell Disease: Imaging of cerebrovascular accident complication. Radiology 1998; 206: 311-321
12. Merkel KN, Ginsberg PL, Parker JC Jr, Post MJ, Cerebrovascular disease in sickle anaemia: a clinical , pathological and radiological correlation. Stroke 1989; 45-52
13. Feldenzer JA, Bueche MJ, Venes JL. Superior Saggital Sinus thrombosis with infarction in sickle cell trait. Stroke 1987; 18: 656-80
14. Oguz M, Aksurgur EH, Soyupak SK, Yidirim AU. Vein of Galen and sinus thrombosis with Infarction in sickle cell anaemia. CT Follow up and angiographic demonstration. Neuroradiol. 1994; 36: 155-156
15. Adams RJ, Nichols FT, McKie V, McKie K Milner P, Gammal TE Cerebral infarction in sickle cell anaemia, mechanism based on CT and MRI. Neurology 1988; 38: 1012-7
16. Styles LA, Hoppe C, Klitz W, Vichinsky E, Lubin B, Trachtenberg E. Evidence for HLA-related susceptibility for stroke in children with sickle cell disease. Blood 2000; 95: 3562-3567
17. Moser FG, Miller ST, Bello JA et al .The spectrum of brain MR abnormalities in sickle cell disease: a report from co-operative study of sickle cell disease. AJNR 1996; 17: 965-72
18. Craft S, Schatz J, Glauser, LeeB, DeBaun M, Effects of bifrontal stroke during childhood on attention. Evidence from children with sickle cell anaemia. Dev. Neuropsychiatry 1994; 110: 285-297
19. Pegelow CH. Stroke in children with sickle cell anaemia: aetiology and treatment Paediatr Drugs 2001; 3: 421-32
20. Palhalakis SG, Bello J, Prohovnik et al. Brain infarction in Sickle Cell anaemia; magnetic resonance correlates. Ann. Neurol. 1988; 23:125-130
21. Adams RJ, McKie VC, Hsu L, et al Prevention of first stroke by transfusion in children with sickle cell anaemia and abnormal results on transcranial Doppler Ultrasonography. N. Engl. J. Med 1998; 399: 5-11
22. Lusher JM, Haghighat H, Khalifa AS. A prophylactic transfusion programe for children with sickle cell anaemia complicated by CNS infarction. Am. J Haematol.1976; 1: 265-273

Received: 19/03/04
Accepted: 27/01/05