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Full Length Research Paper

Stroke in Nigerian Children with Sickle Cell Anaemia

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Stroke is a leading cause of physical disability worldwide. Sickle cell anaemia (SCA) is the most common cause of stroke in children. The aim is to determine the prevalence of stroke in children with SCA at the University of Port Harcourt Teaching Hospital, Nigeria. Retrospective studies of patients with SCA (6 months to 16years) with stroke, over a 5 year period were carried out. Data was extracted from the medical records of patients. Descriptive statistics was used to analyze data. A total of 256 medical records of children with SCA were reviewed. Of these, 11 patients had stroke (4.3%). Seven (63.6%) were below 10 years. The main clinical manifestations were seizure, 6 patients (54.5%) and hemiplegia 5 patients (45.5%). Recurrent stroke occurred in 2 patients (18.2%). Computed tomography scan reports of the brain were not available in 3 patients which confirmed ischaemic stroke. Stable-state haematocrit was below 20% in 72.7% of patients. One death was documented. The prevalence of stroke in children with SCA is 4.3% and children under the age of 10 years with low haematocrit values are at greater risk of developing stroke.

Key words: Stroke, sickle cell anaemia, children, Nigeria.

INTRODUCTION

Sickle cell anaemia is one of the leading risk factors for stroke in children (Earley et al., 1998). Stroke is major cause of mortality and morbidity in children who suffer from sickle cell anaemia (Fullerton et al., 2003). SCA can cause blood cells to clump and block occlude blood vessels, thereby increasing stroke risk (Mehta and Adams, 2006). It has been argued that SCA increases the risk of ischaemic stroke because of increased heamolysis and altered rheologic properties of red blood cells (Lemogoum et al., 2005).

The incidence of stroke in children with SCA may be up to 280 times higher than in the paediatrics population (Earley et al., 1998). Annually, as many as 15 million people suffer a stroke worldwide, of these 5 million die and another 5 million are left permanently disabled, placing a heavy burden on individuals, families and communities (Fullerton et al., 2003). Recent reports have predicted that the 21st century will see a serious economic burden from non-communicable diseases in low income countries and have called for action (WHO, 2002; Connor et al., 2007).

Despite evidence of the increasing burden of stroke in Africa, data in children are rare in Nigeria. The aim of this study was to determine the prevalence of stoke in children with SCA at the University of Port Harcourt Teaching Hospital, Nigeria.

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Abbreviations: SCA, Sickle cell anaemia; **CT,** computed tomography.

MATERIALS AND METHODS

This was a retrospective study of children with SCA, who were seen in the Paediatric Department of UPTH (University of Port Harcourt Teaching Hospital) over a 5 year period (1st January, 2005 - 31st December, 2009). The UPTH is located in Port Harcourt metropolis, the Capital city of Rivers State of Nigeria. It is the main referral hospital for Rivers State and also receives referrals from neighbouring states. All children aged 6 months to 16 years of age with haemoglobin genotype SS were included. Stroke was defined as a neurological deficit lasting for more than 24 hours, excluding transient ischemic attacks, seizures without neurological deficit, meningitis and encephalitis. Children with confirmed meningitis and encephalitis were excluded. The diagnosis of SCD was based on cellulose acetate electrophoresis at alkaline pH. Data extracted from the case notes included; age, gender, haemoglobin genotype,

Table 1. Age distribution of homozygote ss with stroke.

Age (Years)	Frequency	Percentage
0.5- 5	1	9.1
>5-10	6	54.5
>10	4	36.4

Table 2. Stable state haematological data of homozygote SS with Stroke.

No. of patients	Haematocrit (%)	WBC×10^9	Platelet
1	18	22.0	256
2	31	16.0	225
3	21	8.5	420
4	19	15.3	180
5	24	18.3	194
6	16	9.4	-
7	18	20.0	230
8	16	19.4	184
9	19	16.2	174
10	17	10.8	280
11	18	16.8	412

stable-state haemoglobin and full blood count, brain CT scan, recurrence of stroke and outcome. Data collected were analysed using EPI-INFO version 2002, Microsoft word. Descriptive statistics in the form of mean, standard deviation and frequency tables was used to analyze data.

RESULTS

There were a total of 256 children with SCA, over a 5 year period made up of 149 males and 107 females. Of these, 11 patients (8 males, 3 females) had clinical manifestation of stroke. The prevalence of stroke was 4.3%. The age distribution of the patients showed that 54.5% of the patients were between 5 and 10 years (Table 1). The clinical manifestations were seizure, 6 (54.5%), hemiparesis/hemiplegia 5, (45.5%), speech disorder 1, (9.1%), facial nerve palsy 1, (9.1%) and coma 2, (18.2%). Recurrent stroke was observed in 2, (18.2%). Computed tomography (CT) scan of brain was available in 3 cases which confirmed occurrence of ischaemic stroke. The CT scan result was not available in 8 patients probably because of lack of finance. The mean stable state Packed Cell volume for patient with stroke and those without stroke is 19.73±9.2 and 18.12±3.7 respectively. Stable-state haematocrit of patients with stroke showed that 8 (72.7%) patients had level below 20%, 2, (18.2%) between 20 and 30% and 1 (9.1%) had level above 30% (Table 2).

White blood cell count was elevated in 9 patients (81.8%) as shown in Table 2. Lipid profile was not available. All the patients were managed on chronic blood transfusion. One (9.1%) death was documented.

DISCUSSION

Stroke is one of the most important neurological complications reported in many patients with SCA (Plumacher et al., 2004). The percentage of occurrence of stroke in our study is 3.2%. This is comparable to some other studies (Serjeant and Serjeant, 2001; Wood, 1978). Higher percentages had been reported (Balkaran et al., 1992; Njamnshi et al., 2005; Van-Hoff et al., 1985). The difference may be attributable to the fact that most cases of SCA were not present in the hospital but rather in spiritual homes, because some parents believe that symptoms associated with sickle cell disease are of malevolent reincarnation (Nzewi, 2001).

The majority of our patients were male, which is consistent with other studies (Balkaran et al., 1992; Annobil et al., 1990). The reason for male predominance is unknown. Furthermore, in the literatures, most of the children with SCA affected by stroke fall within 3 to 10 years of age (Balkaran et al., 1992). This is consistent with our finding where the youngest patient was four years and the oldest 10 years. The reason why this age group is at risk for stroke is not known.

In this study, seizure was the commonest clinical manifestation of stroke followed by paralysis/weakness of the limbs in 54.5% and 45.5% of cases respectively. However, a Cameroonian study (Obama et al., 1994) found paralysis of the limbs in 97.1% while in India, Chopra and Prabhakar (1979) noted motor impairment in 100% of their patients. There was recurrence of stroke in 18.2% of our patients. This is lower than 46% reported by Balkaran et al. (2001).

Low hematocrit level was found in more than two third of our patients with stroke. This agrees with previous study (Prohovnik et al., 1989). A low hematocrit level is thought to be associated with increased cerebral blood flow, which is considered a risk factor for stroke in sickle cell disease (SCD) (Herold and Brozovic, 1986; Prohovnik et al., 1989). Steady-state leukocyte count was high in more than three quarter of cases. This is consistent with previous study (Balkaran et al., 1992).

Computed tomography (CT) of the brain is deemed by most authors the most suitable imaging method by virtue of its swiftness, practicality and availability, being able to clearly reveal hemorrhagic events and differentiate them from ischemic events (Gerald et al., 1980). More so, CT is less dependent on the patient's clinical stability, a factor which often contraindicates use of magnetic resonance imaging (MRI) in initial stages of the disease (Gerald et al., 1980). In our communication, CT scan was available for 3 of our patients and showed evidence of

cerebral ischaemia. A retrospective study by Steen et al. (2003), found high rates of radiological changes in children with SCD and that cerebral infarct was found in 35% of children studied.

Our patients were managed with blood transfusion and physiotherapy. There have been no acute stroke treatment studies in SCA, but hydration and exchange transfusion are often recommended (Mehta and Adams, 2006). Transcranial Doppler screening of all children with SCA, and initiation and maintenance of chronic transfusion to maintain hemoglobin S below 30% in the high-risk group, is the only proven prevention strategy for stroke in SCA (Mehta and Adams, 2006). Hydroxyurea is being studied as secondary stroke prevention currently (Lefevre et al., 2008). No recommendation specific to SCA regarding the use of antiplatelet agents or anticoagulants in ischemic stroke can be made (Mehta and Adams, 2006). Bone marrow transplantation can be curative for SCA, and limited data support its use to prevent stroke in SCD (Mehta and Adams, 2006).

Limitation of this study includes retrospective nature of the study, and non availability of the CT scan of the brain for some cases. Also, some folders could not be traced because of poor record keeping, hence, the small number of the patients in this study.

In conclusion, this study has demonstrated that children under the age of 10 years with sickle cell disease are at greater risk of developing stroke and that majority of them had low stable state haemtocrit level. There is need to prevent its occurrence by identifying high-risk patients using noninvasive diagnostic methods such as transcranial ultrasonography and the selective use of transfusion therapy.

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