Pediatric Stroke In A Nigerian Child With Sickle Cell Disease: A Case Report

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Citation

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Abstract

A nine and half year old girl with sickle cell disease (HbSC) developed stroke following a febrile illness. The diagnosis was clinical, as CT scan was not done. She was managed with a single volume exchange blood transfusion and she made remarkable neurological recovery. Few months later, she developed avascular necrosis of the head of femoral bone. No previous report of pediatric stroke has come from this locality. This report supports the role of exchange blood transfusion in the management of cerebrovascular disease complicating sickle cell enemia.

INTRODUCTION

Stroke is a neurologic disorder resulting from inadequate supply of blood to a part of the brain. Of recent, pediatric stroke is being increasingly recognized and researched into as a cause of significant morbidity and mortality. The incidence of pediatric stroke has been estimated to be 1.29-2.3/100,000 American children per year [1, 2], 2.1 in Hong Kong [3], and 1.85 [4] in Cameroon[4] with sickle cell disease (SCD) [1,4,5,6] being a leading etiological factor especially in populations with high representation of black Africans. A look at the records over the last ten years in our 500-bed university teaching hospital serving about 10 million people in south western Nigeria showed 595 cases of stroke recorded, the youngest being a 26 year-old man.

Stroke may be ischemic, hemorrhagic or cryptogenic, the ischemic type being the commonest either generally or among children with SCD. Stroke is a differential diagnosis of depressed consciousness especially when associated with lateralising signs. However, the rarity of severe essential hypertension and age-related acquired heart diseases relegates this disabling condition from consideration by the primary physician in non-specialized units. In sub-Saharan Africa with dearth of skilled personnel and equipments for modern investigation, nervous system infections will rather take priority consideration and delay early diagnosis and appropriate treatment.

There is paucity of data on pediatric stroke in Nigerians despite the high prevalence of the HbS gene. The report is a contribution in this direction.

CASE REPORT

A 9½ years old girl was admitted in August 2004 into the children's emergency ward of Wesley Guild Hospital, Ilesa, Nigeria, with one day history of fever, vomiting and lower abdominal pain for which she had commenced antimalarial (Amodiaquine) therapy. The abdominal pain was of sudden onset, non-colicky, non-radiating and located in the lower region. Urine was dark coloured urine but there was no change in bowel habit. She had been diagnosed five years earlier as SCD (HbSC) patient and had received blood transfusion for severe anemia. She however defaulted from follow-up.

On arrival, she was conscious, ill looking, groaning in pain, pale (hematocrit 20%) but afebrile (T36°C). She weighted 27kg. Pulse and respiratory rates were 100 and 50 per minute respectively. There was suprapubic tenderness with hyperactive bowel sounds while the liver was palpable to 7cm below the coastal margin. With the impression of abdominal pain crises in a sickle cell patient, she was commenced on 4.3% dextrose in fifth saline infusion (100ml/kg) and pentazocin injection.

Two days later, she became febrile (T38.2°C) and her hematocrit had dropped to 16%. She was given blood transfusion. Ciprofloxacin was added to her treatment because Klebsiella species sensitive to this was isolated from her urine culture. A day later, she became drowsy, non-communicating and was barely able to localize pain. There

were neither signs of meningeal irritation, seizures, cerebro-spinal fluid abnormalities or significant counts of malaria parasite on blood film. With the presumptive impression of possible cerebral malaria, she was commenced on Artemether (3.2mg/kg/d for three days).

Further clinical review revealed global hypotonia, hyporeflexia and right lower limb weakness (muscle power grade 2) and right facial nerve palsy. An impression of left frontal lobe deficit, with thrombotic stroke was made. CT scan was not done for logistic reasons. The patient had single volume exchange blood transfusion (EBT) with HbAA blood as well as low dose Aspirin (75mg/day). Over the next 24 hours, there was a dramatic improvement in the level of consciousness and verbal communication. Physiotherapy was commenced and she was discharged on the 11th day of admission but she defaulted follow-up.

Four months later, the patient developed a left hip pain with limping gait following a stressful walk. The hip X-ray revealed avascular necrosis and flattening of a portion of the left femoral head. She was confined to bed to aid healing of the infarcted bone but her gait remained wobbly for months thereafter.

DISCUSSION

Sickle cell disease is an autosomal recessive inherited disorder with cerebrovascular accident (CVA) as one of its major complications [7]. The age at first accident is usually within the first decade of life [3,8]. Most SCD patients are admitted for severe anemia or pain crises (either of the bone or abdomen). Management with blood transfusion, generous fluid intake and analgesics with or without antibiotics is almost a routine.

The diagnosis of stroke in a child with SCD disease is not an easy one in our setting. In sub-Saharan African countries like Nigeria, that are holoendemic for malaria, cerebral malaria is given top consideration in any febrile unconscious child with or without seizures. Addition of lateralising signs makes intracranial infections like meningitis or encephalitis likely first choice diagnoses. The management of this case was along these lines. However, the stuttering level of consciousness, aphasia and motor deficit ensuing after two days of admission in a known SCD patient with urinary tract infection and ongoing antimalarial and antibiotic therapy were suggestive of incipient stroke.

Apart from congenital heart malformations and vasculopathies, SCD and infections are well known risk factors for arterial ischemic stroke [6] in children while low steady state hemoglobin concentration is an added risk factor for those with SCD [9]. Severe anemia was the diagnosis on the two hospital admissions in this patient. However; anemia and urinary tract infection were trigger factors here, as the child was conscious and alert in the first two days of admission. In assessing the risk for first stroke in SCD patients transcranial doppler ultrasound (TCD) measurement of cerebral blood flow rates in excess of 200cm/sec has been found to be very efficient [10]. Furthermore, cranial computed tomography, magnetic resonance imaging and magnetic resonance angiography are important in making a diagnosis while contrast cerebral angiogram [11,12] is confirmatory at the acute phase. Lack of these radiologic facilities made our reliance on clinical features with high index of suspicion [13] imperative in making the diagnosis of stroke in our patient. The remarkable improvement in mental state and motor function in our patient gives credence to the efficacy of exchange blood transfusion although simple blood transfusion is a recognised alternative. Following the first stroke event, recurrence is a major issue and it is known to affect about 25% of arterial ischemic stroke victims after the newborn period. Known risk factors for recurrent stroke include abnormal TCD values[14]. In preventing stroke recurrence in SCD patients, chronic blood transfusion program (CBT) [10,15,16] either by simple partial or exchange blood transfusion, anticoagulation [17], thrombolysis [18], hydroxy urea [19] or bone marrow transplant [20] have been reported to be effective though CBT is more widely employed. CBT has the purpose of diluting the sickle cell hemoglobin to less than 30% [13]while bone marrow transplant eliminates the occurrence of sickled cells. The other adjunctive therapies reduce the formation or propagation of thrombi. Our patient was offered CBT but mother declined. The avascular necrosis of the femoral head, which puts her at great risk for recurrent stroke, might have been prevented although there has been no study on other vasoocclusive problems as risk factors for recurrent stroke in these patients. The Aspirin was to act as antithrombotic against hyperviscousity. Patient might have defaulted its use or the drug was ineffective. CBT is practically difficult in most developing countries for infrastructural and socio-economic reasons including sourcing donor blood and preventing or tackling hemosiderosis if and when it occurs.

This case highlights the problems of diagnosis and prevention of pediatric stokes in this environment and alerts physicians to its possibility here in Nigeria and other resource-poor areas of the world.

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