A Nigerian Child With Spina Bifida Cystica And A Ruinous Aftermath

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Citation

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Abstract

We report a 3-day old term male Nigerian baby with an uncomplicated unruptured spina bifida cystica who defaulted from neurosurgical care and had incision and drainage done by a quack. He presented 3 months later with hydrocephalus and other neurologic impairments. The socio-economic issues that led to this grave outcome are discussed.

INTRODUCTION

Spina bifida cystica, a congenital neural tube defect, may be associated with hydrocephalous and recurrent meningitis. [1] It occurs in about 0.1 – 3.5 in 1,000 live births. [2] The aetiology is uncertain although folic acid supplements given to mothers in pregnancy reduce the risks. [1, 2] Myelomeningocoele and meningocoele are the two common forms of spinal bifida cystica. Meningocoele is less common and less severe being often devoid of neurologic deficits. [2,3,4]

In Nigeria, effective surgical therapy is available [5] but usually fraught with many socio-economic issues some of which are highlighted in this report.

CASE REPORT

A 3-day-old boy was brought to the Wesley Guild Hospital (WGH), Ilesa, Nigeria on December 27, 2004 because of a cystic lower back swelling noticed at birth. The 28-year-old Para 3^{+1} mother had antenatal care and delivery at a clinic operated by a Community Health extension Worker (CHEW). She refused folate supplements during pregnancy for fear of inducing neonatal jaundice. She had previously experienced a first trimester spontaneous abortion. She earns about Three Thousand Naira (Twenty-two US Dollars) monthly as a seamstress. The 45-year-old father logs timber. Both parents were educated up to the primary level. The siblings, aged 7 and 4 years were apparently well.

The term infant weighed 3.0 kg with a normal head circumference (34.0 cm) and full length (46.3 cm). The cystic mid-line swelling at his lower lumbar region measured 2 by 1 cm with normal overlying skin and no discharges. No other abnormality or neurologic deficits were observed. Spina bifida cystica was diagnosed. Following counsel, our offer of investigations and possible neurosurgical intervention was declined.

He was re-presented when aged 3 months, with vomiting and fever of 4 days. The very day they declined our medical advice, the lesion was reportedly incised by the same CHEW and thereafter, dressed daily with methylated spirit and given intramuscular Penicillin injections for 5 days. There was an initial clear fluid discharge which later turned purulent and dried up prior to the healing of the overlying skin.

The infant weighed 5.0 kg, was severely pale, febrile (38.5 °C) and drowsy. Head circumference was 60.0 cm with cranio-facial disproportion, shiny scalp with distended veins, ‘sun-setting' eyes and cracked-pot sign. Both fontanelles were wide and tense. The upper limbs were spastic with both hands persistent fisted. The left lower limb was flaccid but sensations were preserved. The right lower limb was normal. The fleshy lower lumbar swelling measured 3 by 4 cm with a linear scar about 2.5 cm but no discharge. There were peri-anal excoriations, reduced anal tone, continuous faeco-urinary dribbling. He was in congestive cardiac failure. Diagnoses were iatrogenic ruptured spina bifida cystica with hydrocephalus, anaemic heart failure and probable septicaemia to exclude meningitis. Figure 1 shows the baby and Figure 2, the lumbar lesion.

Figure 1

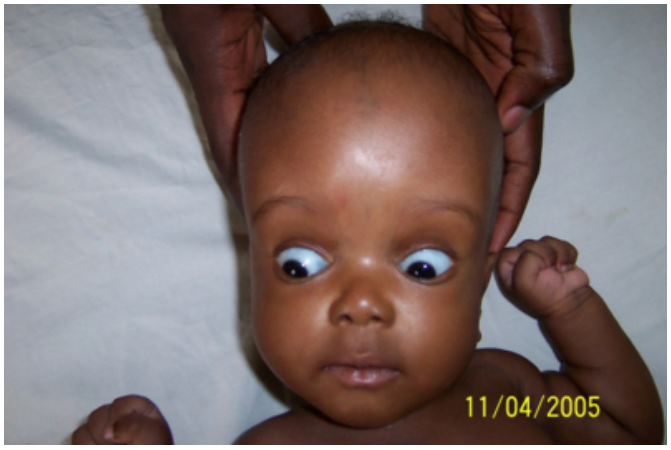
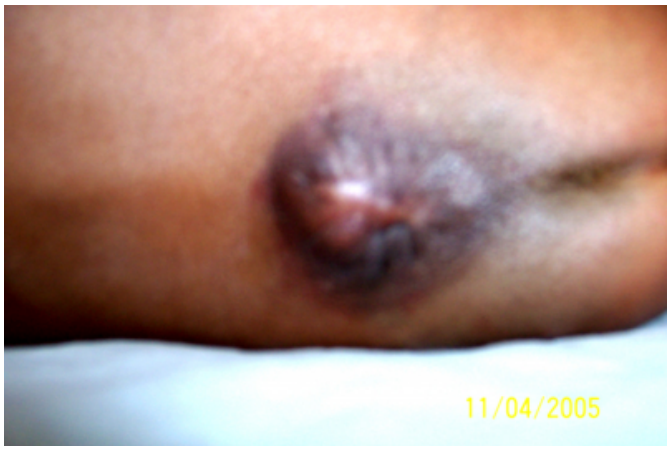


Figure 2



Admission haematocrit was low (11%) but blood sugar (3.7 µmol/ L) normal. The total leucocytes count was 7,000 cells/ mm^3. Blood film showed moderate asexual forms of Plasmodium falciparum. Blood and urine cultures were sterile. Cerebrospinal fluid could not be obtained for analysis because of the mass. Neuro-diagnostic imaging was unavailable. The treatments given included blood transfusion, intramuscular Artemether and intravenous Ampicillin. After 7 days, he made remarkable clinical improvement, parents were counselled and child was referred for neurosurgical review. Again, they defaulted from further follow-up.

DISCUSSION

People with spina bifida may have physical challenges,[1, 2] but good treatment benefits them and the society as some may lead productive lives.[2, 3] Prognosis of unruptured spina bifida cystica with meningocoele is good as surgical advances and the adjuvant therapies needed for rehabilitation have dramatically improved the prospects. [2,3, 4] Such surgical care is available in few tertiary centres in Nigeria although very expensive.[5] However, as typified by our patient, they are poorly utilised because of poverty and adverse socio-cultural influences. The latter and ignorance probably influenced the preference of a seemingly cheaper alternative in a CHEW, who unfortunately operated regardless of ethics, law or knowledge. CHEWs have important contributions in the primary care programme implementation but this does not include highly specialised surgeries. This highlights the laxity in the regulation and control of medical care in the country.

The severe disabilities associated with neurosurgical care of patients with open spina bifida are enormous [2,3,4] and a society like ours lacks the requisite social backup for them.[5] It is unfortunate that an infant, who presented early enough with an apparently uncomplicated Spinal bifida cystica, defaulted from standard care only to re-present with gross neurological impairments. With good social support, an early skilled neurosurgical repair could have prevented this calamity, despite his parental financial inadequacies.

Had our patient's lesion not been operated by a quack, would he have developed hydrocephalus? Contrariwise, hydrocephalus arising de novo with Spinal Bifida Cystica may be part of Arnold Chiari Malformation.[6] However, the hydrocephalus in this case was not obvious at birth. The fact that the mother had a preceding spontaneous abortion may suggest a pre-existing chromosomal disorder in the family. Most first trimester abortions are known to have chromosomal defects.[6]

Unfortunately, an unfounded fear of inducing a disease in the oncoming child cost the mother the protective effects of antenatal folate supplementation. Ironically, the baby she tried to protect from treatable neonatal jaundice developed a disorder as serious as a neural tube defect! This family could have benefited from well-equipped prenatal care particularly, genetic counselling.[1, 2, 6] This calls for improved health education drive to improve the rate of utilisation of available health services. The social welfare sector of the country's healthcare system needs to be strengthened in order to minimise such misadventure into the hands of poorly skilled attendants.

This case underscores the need for the effective regulation and control of the functions of different cadres of healthcare practitioners. This has benefits including protecting patients from iatrogenic insults. Also, we wish to emphasis the need for an improved social welfare system and better funding of

the health sector in the country, particularly with respect to dysmorphic infants.

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References

1. Association for Spina Bifida and Hydrocephalus. Information on Spina Bifida. http://www.fortunecity.com/millenium/plumpton/268/sb.htm Date assessed: April 25, 2005.
2. Foster MR. Spina Bifida. eMedicine. http://www.emedicine.com/orthoped/topic557.htm Date last updated: November 11, 2004, Date assessed: April 25, 2005.
3. Oakeshott P, Hunt GM. Long-term outcome in open spina bifida. Br J Gen Pract. 2003; 53 (493): 632 - 6.
4. Salomao JF, Pinheiro JA, Carvalho JG, Leibinger RD, Lucchesi G, Bomfim V. Myelomeningocele: surgical treatment and results. J Pediatr (Rio J) 1995; 71(6): 317 - 21.
5. Olumide AA, Adeloye A. Management of Spina Bifida Cystica at Ibadan. Nig J Paediatr 1980; 7 (2): 46 - 50.
6. Klaus MH and Fanaroff AA (eds). Care of the High-risk neonate 5th Edition. Philadelphia, WB Saunders, 2001.

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