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Case Report

Congenital Hydrocephalus in Identical Twins: Case Report

Oluwamayowa O OPARA*, Lasseini ALI, Nasiru J ISMAIL, Olugbenga O OGUNLEYE and Mahmud M RAJI

Department of Neurosurgery, Usmanu Dan Fodiyo University Teaching Hospital, Sokoto, Sokoto State, Nigeria

***Corresponding Author:** Oluwamayowa O OPARA, Department of Neurosurgery, Usmanu Dan Fodiyo University Teaching Hospital, Sokoto, Sokoto State, Nigeria.

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Abstract

Congenital hydrocephalus in identical twins is rare. Despite the fact that twinning and congenital hydrocephalus are common in Nigeria, congenital hydrocephalus in identical twins is still very rare in Nigeria with only one case reported so far. This case report highlights the second case from Nigeria, to add to the literature.

A set of male identical twins presented to us at 6 months of age with progressive head enlargement after birth. They had obvious craniofacial disproportion, distended scalp veins, sun setting eyes, and brisk knee jerk reflexes bilaterally. There was no thumb adduction. One had ventriculoperitoneal shunting and was discharged home. The other unfortunately, died from other causes.

Keywords: Congenital, Hydrocephalus, Identical Twins

Introduction

Congenital hydrocephalus is a common aetiology of hydrocephalus in Nigeria [1]. Twinning rates in Nigeria are also one of the highest in the world. However congenital hydrocephalus in identical twins remains a rarity even in Nigeria. The following is a description of the first case reported from our centre in Northern Nigeria.

Case Report

A set of male twins at 6 months of age presented to our outpatient clinic with history of progressively increasing head size noticed about a week after delivery. They were born at term to a 20 year old mother who had no scheduled antenatal care and no history of febrile illness in pregnancy. Delivery was at home in a rural area, and there was no history of jaundice or fever in the neonatal period. The mother was a full-time housewife with no income and father was a commercial motorcycle rider.

Physical examination showed a set of identical twins. They both had obvious craniofacial disproportion with distended scalp veins, a bulging and tense anterior fontanelle, with sutural diastasis, sun-setting eyes and brisk knee jerk reflexes bilaterally. There was no back swelling, no limb deformity, and no adducted thumbs. One of the twins (twin 1) had severe sepsis, focus on the chest. They both underwent a transfontanelle ultrasound scan with findings of panventriculomegaly. A CT scan could not be done for logistic reasons.

Twin 2 had an emergency ventriculoperitoneal shunting with intra-operative findings of clear cerebrospinal fluid under high pressure. Ventricular access was gained via Keen's point and a medium pressure Chhabra shunt was used. He had an uneventful post-operative course and was discharged home on the seventh post-operative day after removal of stitches. Twin 1 was admitted in the intensive care unit where he died while being resuscitated.

Discussion

There are very few reports of congenital hydrocephalus in identical twins found in literature. Borle was said to have in 1953 reviewed cases reported since 1899, a period of 54 years and only 7 pairs were reported [2]. Gellman in 1959 critically assessed Borle's report and noted that at least 1 of the 7 she reported was likely post meningitic albeit in identical twins. He also noted a 1956 report by

Conn et al describing congenital hydrocephalus in a pair of identical achondroplastic twins. He went on to give a case report of a set of identical male twins with congenital hydrocephalus who did not present in hospital until about 5 years of age for an entirely different medical reason. Even though the pictures in the article do not show a markedly enlarged head, the ventricular system was said to be moderately distended on pneumoencephalogram [2]. Ahmet et al in 2004 reported a set of female identical twins in Turkey who presented at birth [3]. Wachi., *et al.* "report was of external hydrocephalus in identical twins. Idowu et al in Lagos, Nigeria also reported one case in identical male twins [4]. Other studies have noted congenital hydrocephalus in one of the twins [5], but in both simultaneously is very uncommon.

Congenital hydrocephalus is thought to have varied aetiologic factors including genetic and environmental factors. It could also be syndromic or non-syndromic. Syndromic types of congenital hydrocephalus may be associated with Dandy walker malformation, Chiari malformation, trisomy 13, 18, etc. The gene that has been linked to isolated congenital hydrocephalus is the x-linked HSAS1 gene which has been found to occur in males with associated congenital aqueductal stenosis, and the characteristic findings of adducted thumbs in these patients. This is also known as the Bicker Adams syndrome.

In the twins presented in this article, no other congenital lesion was detected grossly, and they did not have adducted thumbs. Unfortunately genetic studies and a computerized tomography scans could not be done in these patients for logistic reasons.

Conclusion

In conclusion, congenital hydrocephalus in identical twins remains a rarity and seems to be more common in males. Further studies would help in elucidating the specific aetiology of this condition.

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