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An Unfortunate Outcome in a Child with an Encephalocoele from a Rural Area

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Summary

Oseni SBA, Oyelami OA, Owa JA. An Unfortunate Outcome in a Child with an Encephalocoele from a Rural Area. *Nigerian Journal of Paediatrics* **2004;31:93.** A case of a three-and-a-half month old infant from a rural area, who presented with an encephalocoele, is reported. The case was complicated by acute rupture of the encephalocoele. Due to financial difficulties, there was late presentation at an appropriate health facility, and this delayed adequate management. The problem was further compounded by an industrial unrest in the public health sector at the time. There is a need to site investigational and surgical services for minor neurosurgical problems in the rural areas of the country.

Keywords: encephalocoele, rural, infant

Introduction

AN encephalocoele is one of the major craniospinal dysraphisms otherwise called, neural tube defects. These defects are due to failure of closure of neuropores, which in the case of an encephlocoele, is the rostral neuropore, between the 23rd and 25th days of gestation.[1,2] This leads to herniation of cerebrospinal fluid-filled meninges with or without brain tissue through the defect in the skull. This neurosurgical disorder requires the type of care that is rare in most cities in the country and are non-existent in the rural areas. The obvious disfiguring lump on the head may have prompted the quest for orthodox or traditional medical intervention. The following case is presented to illustrate the case of an unfortunate infant from a rural community who had an encephalocele.

Case Report

WT (144033) was a three-and-half-months old male infant, the first of a set of twins brought from a nearby village to the Wesley Guild Hospital, Ilesa, on account of dripping of "clear water" from a swelling at the back of his head, for three days. The swelling was noticed at birth and had not changed in size until the dripping started and the swelling gradually reduced in size. There had apparently been no interference with the swelling prior to the presentation. A day before presentation, the patient had brief episodes of twitching of both legs and the right hand. The mother could not ascertain if consciousness was maintained during the twitching. The child also refused to suck at the breast but there was no associated fever.

The patient was a product of a full term pregnancy and normal delivery at home. The pregnancy felt heavier than the previous ones but it was not known if the liquor was excessive. The baby cried soon after birth and had been sucking well since then. The neonatal life was uneventful. The patient was yet to achieve neck control at presentation but could smile. He had not received any immunization. Both parents were primary six school certificate holders. The mother was a petty trader while the father was a peasant farmer and part-time musician. The second twin and two older siblings were well.

The patient weighed 4.65kg with a supine length of 56cm and could visually follow the examiner. Rectal temperature was 37.5°C with pulse and respiratory rates of 120/min and 32/min, respectively. Examination of the head revealed a head circumference of 43cm with gaping saggital suture. The anterior fontanelle measured 6cm longitudinally and 4cm across while the posterior one measured 8cm vertically and 5cm at the base. A

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scaly lump of redundant skin was in its depression (Figs 1 and 2). The lump did not trans-illuminate but clear and colourless fluid was dripping from a sinus in its posterior part. No scarification mark was noticed on the head or other parts of the body. The patient was jittery with increase in muscle tone in all the limbs. The deep tendon reflexes at the knees and ankles were equivocal while ankle clonus was absent. Superficial sensation was preserved. There was no spina bifida or any other abnormality observed on examination of the eyes, chest, abdomen and appendicular skeleton.

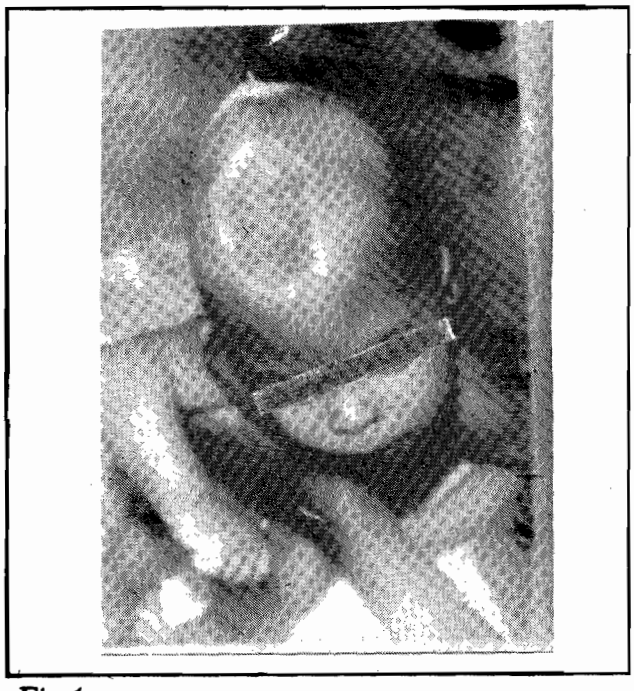

Fig. 1

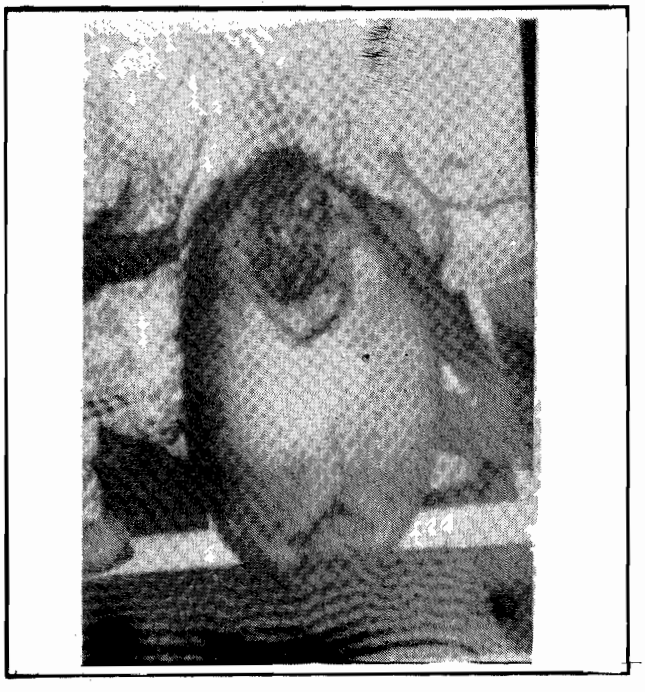

Fig. 2

A diagnosis of ruptured occipital encephalocoele with leaking sinus and possible meningitis was made. The patient was admitted and started on intravenous crystalline penicillin 400,000 units/kg/day and chloramphenicol 75mg/kg/day. Sterile saline dressing was applied daily to the leaking sinus. Further investigations or referral to a neurosurgeon could not be made, as the patient presented at a time when there was an industrial crisis that paralysed health services in the public health sector of the country. Breastfeeding was advised as the patient was observed to be sucking well.

Four days later, the patient developed cold extremities but was being kept warm with a blanket, without good effect. The following day he became weak, refused to suck at the breast and soon developed gasping respiration. Resuscitation failed and the patient eventually died on the fifth day of admission. The parents refused autopsy. The highest temperature recorded during the course of admission was 37.8°C.

Discussion

Encephalocoele is a herniation of the brain or its covering meninges through a defect in the skull. It is a major neural tube defect occasionally encountered in paediatric practice. The incidence is 1-4/3,000-10,000 live births.[3-5] A previous review of cases in this environment reported an incidence of 0.5/1,000 live births.[6] The cranial type of dysraphism is thought to occur as a result of faulty closure of the rostral neuropore between the twenty-third and twenty-fifth day of gestation.[1,2] It presents as a mass of variable size on the head. While about 75 percent of encephalocoeles occur in the occipital area, less common sites include fronto-nasal, parieto-occiptal, nasopharyngeal and orbital regions.

Depending on the contents of the sac,[7] it can be a cranial meningocoele (encephalo-meningocoele) i.e. a cerebrospinal fluid (CSF)-filled sac with no neural elements; a meningoencephalocoele i.e. sac containing appreciable amounts of normal or malformed brain tissue; an encephalocystocele – where a portion of the cerebral ventricle with its overlying cerebral mantle herniates into the sac or encephalocystomeningocoele, where large amounts of CSF surrounds the herniating brain substance. Simple cranium bifidum is the existence of the large gap without protruding sac. In this patient, the sac had collapsed, leaving redundant scaly skin in the depression of the gaping posterior fontanelle (Fig 3). This suggests a case of cranial meningocoele, which was expected to have good prognosis as in cases of meningocoele of a spinal bifida.[8]

An encephalocoele may be diagnosed antenatally by an abdominal ultrasound where the lesion gives the

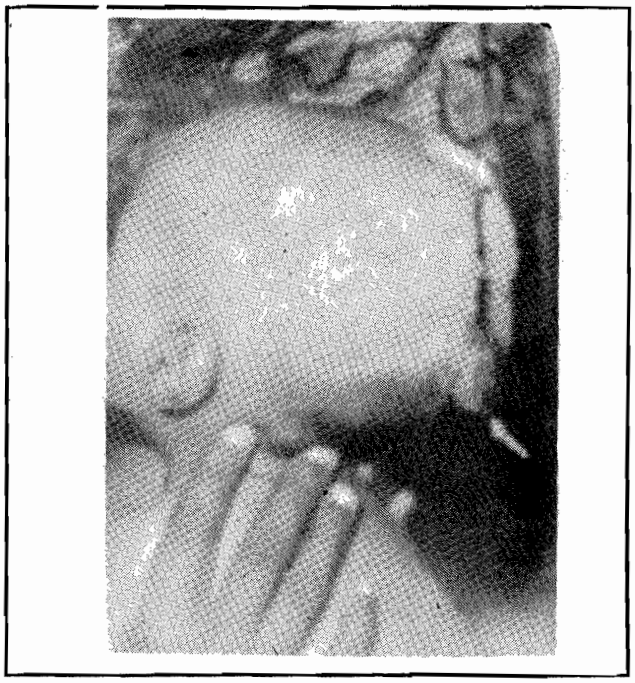

Fig. 3

appearance of a sac-like projection from the head and a tongue-like ecogenic protrusion (the "lemon sign") indicating herniating brain tissue into the sac.[9,10] Its diagnosis at birth is easy except when it is located in the nasopharyngeal or retro-orbital sites. Skull roentgenogram will often identify the gaping suture line at the site of the lesion while skull ultrasound scan may help to clarify structures in the hernial sac as well as possible association with hydrocephalus as in cases of autosomal recessively inherited Chemike syndrome. CT scan and MRI are important investigations that were out of the reach of this poor family. Ophthalmologic examination should be carried out to exclude Knoblock syndrome in which occipital encephalocele is associated with ocular abnormalities like myopia, vitreo-retinal degeneration and retinal detachment. Our patient did not have microcephaly, polydactyly, cleft lip or palate, microphthalmia or abnormal genitalia to suggest Meckel-Gruber syndrome – another autosomal recessive disorder associated with encephalocele.[9] Spinal x-ray could not be done to rule out associated spina bifida occulta although a thorough search did not reveal any tell-tale sign like a tuft of hair, lipoma, dermal dimple, sinus or fistula along the spine that would suggest such.

Most encephalocoeles trans-illuminate regardless of the content while the head circumference is often normal except when there is associated significant hydrocephalus. In our patient, transillumination was negative raising the hope of a good prognosis if surgery could be done. However, the CSF fistula posed the danger of meningitis. This was assumed, hence our use of antibiotics pending definitive surgery that was not possible in the circumstance. Another life-threatening danger in patients with encephalocele as in spina bifida is rupture of the sac leading to loss of CSF.[12] This possibility is an indication for urgent surgical intervention in the neonatal period.[13] Rupture is usually due to birth trauma;[12] our patient was three and half months old at presentation. Healing would have occurred if there was any abrasion at birth and the integrity of the covering skin at this age would require significant trauma to rupture it.

Ours is a culture where swellings of any part of the body will not be left alone but interfered with in the form of hot fomentation and application of all kinds of ointments and traditional remedies. Sometimes scarification marks or outright incision are made in the hope of draining the supposed fluid content sometimes regarded as "black blood". This may amount to "fistulotomy." These traditional interventions are common, for example, in sickle cell anemia patients with persistent splenomegaly, kwashiorkor and nephrotic syndrome patients with pedal oedema or in people with arthritis. Although this was denied, we think this was probably the cause the leakage of CSF fluid from the encephalocoele at presentation.

The embarrassment caused by the disfiguring encephalocoele probably prompted the parents to seek some form of intervention to get rid of the lump. The poor educational and low economic power of the family might have made traditional options more attractive than orthodox health care. The universal basic education scheme currently being canvassed by the Federal Government, if faithfully implemented, will hopefully increase the perception and preference for orthodox health care by the rural communities. Accessibility will improve if investigational and surgical services for minor neurosurgical problems are located at the secondary and sub-tertiary health care delivery levels while preconceptional folic acid supplementation may prevent re-occurrence of the condition.[14,15] Furthermore, improved referral system within the health care delivery system will enhance the care of such patients. Perhaps, the National Health Insurance Scheme that has been on the drawing board for so many years holds some promise for indigent patients.

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