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Bilateral Cavernous Sinus Thrombosis in Sickle Cell Anaemia

Report of a case and its successful management

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Summary

Nottidge, V. A. and Familusi, J. B. (1976). *Nigerian Journal of Paediatrics*, 3 (1), 29. **Bilateral Cavernous Sinus Thrombosis in Sickle Cell Anaemia**—*Report of a case and its successful management.* A case of bilateral cavernous sinus thrombosis is described in a child who has sickle cell anaemia. Treatment with antibiotics and tolazoline resulted in full recovery. Previous reported instances of dural sinus thrombosis complicating sickle cell disease have been fatal. It is suggested that administration of tolazoline may have contributed to this favourable outcome.

Several cerebrovascular complications of sickle cell disease have been reported (Greer and Schotland, 1962; Mabayoje, 1956; Adeloye and Odeku, 1970), but those of the dural sinuses appear to be rare. Ford (1960), however, reported the autopsy findings of longitudinal sinus thrombosis in an eighteen-month old negro boy with sickle cell anaemia (HbS) and Schenk (1964) also reported thrombosis of the same sinus in a 12-year old boy with sickle cell trait (HbAS). Adeloye and Odeku (1970) reviewing the neurological complications in 257 children with homozygous sickle cell disease (HbS) found only one case of left lateral sinus thrombosis. As far as can be ascertained from the literature, there has been no previous report of cavernous sinus thrombosis in association with sickle cell disease. It is noteworthy that previous reported cases of dural sinus thrombosis in sickle cell disease were all discovered at autopsy. The present communication concerning a child with sickle cell anaemia complicated by bilateral cavernous sinus thrombosis is therefore of special interest because: (1) the diagnosis was made during life and (2) the patient recovered fully from the complication.

Case Report

A. O. (Hosp. No. 244732) was referred to the University College Hospital (U.C.H.), Ibadan, on July 10, 1970 at the age of $2\frac{1}{2}$ years with a 2-day history of generalised body pains, bilateral proptosis, swelling of the left side of the head, and swelling of both hands. It was further stated that from the age of six months, he had suffered from intermittent swellings of ankles, wrists and fingers, the attacks recurring at intervals of 4 to 6 months.

Physical examination revealed a febrile (temp. 40.3°C), irritable child with generalized bone tenderness. He was small for his stated age, weighing only 10kg on admission. In addition to marked bilateral proptosis and chemosis

(Fig. 1), there was complete ophthalmoplegia and marked visual loss, he was unable to perceive finger movements. There was severe mucosal pallor. He had cellulitis of the scalp, more marked over the parietal eminences, and more severe on the left than the right side. The spleen was not clinically enlarged, but the liver was palpable to 3 cm below the costal margin.

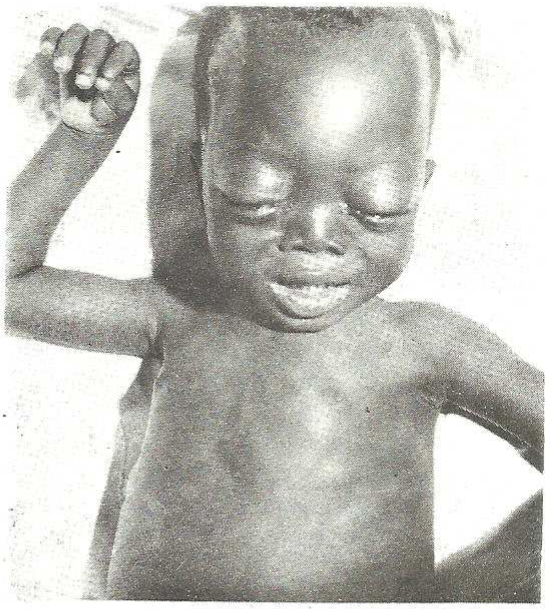

Fig. 1. Patient on admission, aged 2½ years. Note proptosis and chemosis.

Laboratory investigations showed the following: haematocrit 13 per cent; wbc 18,800 per cmm, (57 per cent neutrophils, 40 per cent lymphocytes and 3 per cent monocytes). Haemoglobin electrophoresis revealed Haemoglobin S. Rings of P. malariae were found in the blood film. An aspirate of the left parietal scalp revealed sickled erythrocytes and pus cells on microscopy. Lumbar puncture produced clear, colourless cerebrospinal fluid (CSF) which, apart from a few red cells, was normal on microscopic and microbiological examination. Skull radiographs showed widening of the frontal diploic space, a frequent finding in children with abnormal haemoglobins. The clinical diagnosis was bilateral cavernous sinus thrombosis complicating sickle cell crisis. Neuroblastoma, retinoblastoma and Burkitt's lymphoma which are more frequent causes of proptosis in children in the local environment, were considered in the differential diagnosis but later excluded.

The patient was treated with antibiotics (penicillin and streptomycin; later chloramphenicol), antimalarial (chloroquine sulphate), analgesics and anticonvulsants. He was also given tolazoline (2.5mg/Kg/day) orally on the basis that this might improve his cerebral blood flow. Because the chemosis was rapidly followed by exposure keratitis, bilateral tarsorrhaphy was performed.

The response to therapy was slow but generally satisfactory. The proptosis and chemosis gradually regressed over a period of one month, after which the tarsorrhaphy stitches were removed to reveal good vision. He was discharged from hospital on August 20, 1970 quite well, and with normal visual acuity. When last seen as an outpatient in March 1975, at the age of seven years (Fig. 2) he was physically well. His academic performance at school was also reported to be satisfactory. Psychometric assessment at this time showed an overall score on the McCarthy Scale of children's abilities within the average range. His performance on the perceptual performance sub-test, was poor. He does however, continue to have occasional bone pains and jaundice.

Comments

The increased susceptibility of children with sickle cell anaemia to various types of infection is well known (Barret-Connor, 1971). In the nervous system, the infection commonly presents as bacterial meningitis (Robinson and Watson, 1966; Adeloye and Odeku, 1970; Barret-Conner, 1971), and very infrequently as dural sinus thrombosis. Infection and the sickling diathesis appear to have been important factors in the pathogenesis of cavernous sinus thrombosis in this child. The tendency for cavernous sinus thrombosis to complicate sepsis of the face and

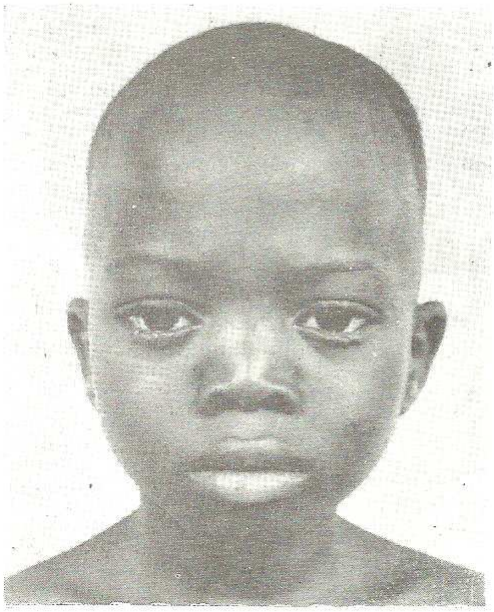

Fig. 2. Patient at age 7 years.
Note tarsorrhaphy scars but otherwise normal eyes.

the underlying soft tissues is well documented (Ford, 1960; Blackwood, 1963; Bassey and Elebute, 1968). That the thrombosis was associated with scalp infection in our patient is suggested by the high and prolonged pyrexia, the leucocytosis, and the presence of pus cells in the scalp aspirate. The susceptibility of children with sickle cell disease to cerebral vasculo-occlusive complications is also well recognised (Greer and Schotland, 1962; Gold, Hammill and Carter, 1964; Adeloye and Odeku, 1970). On the whole, cavernous sinus thrombosis is very rare in our experience in spite of the high prevalence of both the sickle cell gene and infections in the environment. It is suggested that the coexistence of infection and sickle cell crisis in our patient was partly responsible for the development of cavernous sinus thrombosis.

The established management of cases of dural sinus thrombosis consists of antibiotic treatment of coexisting infection and anti-convulsants for treatment or prevention of seizures; these are usually supplemented with other supportive measures, such as bladder care and respiratory assistance if the patient is comatose. The decision to use Tolazoline (priscol) in our patient was prompted by the observations of Smith and Turton (1951) and Engel (1952) that appreciable increase in cerebral blood flow may follow the administration of this drug. Reduced cerebral blood flow was likely to have occurred in our patient, both as a result of sludging associated with sickle cell crisis, (Lin-fu, 1972) and also because of cerebral oedema consequent on dural sinus thrombosis (Blackwood, 1963). The administration of Tolazoline may have contributed to our patient's recovery and we therefore suggest that further trials of the drug in cerebrovascular occlusive conditions are worthwhile.

The psychometric findings in our patient are in agreement with previous observations on children with sickle cell disease. Adeloye and Odeku (1970) using verbal skills and school grade placements, found intelligence in general to be normal in sickle cell disease compared with normal controls. Chodorkoff and Whitten (1963), in a more detailed study, using the Stanford-Binet and the Wechsler Intelligence Scale, also concluded that sickle cell anaemia, in itself, does not affect intellectual or psychological functioning. That our patient had average performance on the McCarthy Scale is consistent with these previous reports. Whether his poor rating on the perceptual performance subtest is aetiologically related to the sickle cell disease or the cavernous sinus thrombosis is, however, difficult to determine.

Acknowledgement

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