Short report

Cranial CT in the haemolytic uraemic syndrome

DB MENDELSON, * Y HERTZANU, * B CHAITOWITZ, * JD CARTWRIGHT†

From the Departments of Diagnostic Radiology* and Paediatrics, † Johannesburg Hospital and University of the Witwatersrand, Private Bag X39, Johannesburg 2000, Republic of South Africa.

SUMMARY A patient with the haemolytic uraemic syndrome and severe encephalopathy is described. The initial extensive brain hypodensity on CT scan was followed 10 days later by diffuse enhancement of the cerebral gray-white matter interface. A possible explanation for this phenomenon is suggested.

A wide spectrum of neurological complications has been described in the haemolytic uraemic syndrome. We present a case with major encephalopathy in whom serial computed tomogram (CT) scans were remarkably abnormal.

Case report

An eleven-month-old child with the haemolytic uraemic syndrome presented with generalised seizures and became comatose with decerebrate rigidity. The initial unenhanced CT scan showed extensive bilateral frontal and parietal hypodensity. These features were in keeping with ischaemic or infarctive oedema. The posterior fossa, basal ganglia and thalami appeared uninvolved (fig 1). An enhanced scan was not performed at this time as the patient was in acute renal failure. The enhanced scan was done 11 days later while the child was on peritoneal dialysis. This scan showed diffuse contrast enhancement at the cerebral gray-white matter interface (fig 2A, B, C). In addition the ventricular system was more dilated. Over the next ten weeks there was a gradual improvement in renal function; however, neurologically the child remained decerebrate with generalised rigidity. There was no change in her head size. A repeat CT scan showed generalised cerebral atrophy and subcortical white matter demyelination (fig 3A, & B). The child was discharged and on follow up over the past 6 months her neurological status remains unchanged. She remains in a day centre for the mentally retarded on anti-seizure medication.

Address for reprint requests: Dr DB Mendelsohn, Dept of Radiology, Johannesburg Hospital, Private Bag X39, Johannesburg 2000, South Africa

Received 8 December 1983. Accepted 30 January 1984

Discussion

The haemolytic uraemic syndrome is the commonest cause of acute renal failure in infants and young children, with a mortality of 10-20%.^{1,2} Central nervous system involvement occurs in 30% to 50% of cases and includes generalised or focal seizures, alterations of consciousness, hemiparesis, aphasia and decerebrate posturing.^{3,4} Necropsy brain studies have shown ischaemia and focal areas of infarction with surrounding oedema and necrosis. Several causes have been postulated, including vascular occlusive lesions, oedema and hypoxia and microthrombi.⁵ In an extensive review of the English literature we were able to find only a single report relating to the cranial CT findings in the haemolytic uraemic syndrome. These included initial hypodense areas, indicative of cortical infarction into which haemorrhage was shown two weeks later. These features were presumed to be due to delayed haemorrhage secondary to cerebral microangiopathy and associated microthrombi.⁶

In our patient the cerebral enhancement was consistent with an extensive blood brain barrier disturbance at the gray-white matter interface related to cerebral microangiopathy. Extensive haemorrhage alone is unlikely in view of the distribution and magnitude of the increased density on CT. We cannot explain the selective involvement of this interface and future cases with necropsy findings may clarify the mechanisms responsible for the above CT appearances.

Fig 1 Initial unenhanced scan showing diffuse bilateral and symmetrical frontal and parietal hypodense areas. The basal ganglia, thalami and posterior fossa are normal.

Fig 2A, B and C Enhanced scan performed 11 days later demonstrates extensive gray-white matter interface enhancement. The ventricular system is more dilated than the initial scan. A scalp vein infusion set is noted on the right.

Fig 3A and B Pre-and post-contrast scans 10 weeks later show diffuse leukomalacia and cerebral atrophy.

References

- ¹ Gianantonio CA, Vitacco M, Mendilaharsu F, Gallo GE, Sojo ET. The hemolytic-uremic syndrome. *Nephron* 1973;**11**:174–92.
- ² Kaplan BS, Thomson PD, de Chadarevian JP. The hemolytic uremic syndrome. *Pediatr Clin North Am* 1976;**23**:761–77.
- ³ Rooney JR, Anderson RM, Hopkins IJ. Clinical and pathological aspects of the central nervous system involvement in the haemolytic uremic syndrome. *Aust Paediatr* 1971;**7**:28–33.
- ⁴ Bale JF, Brasher C, Siegler L. CNS manifestations of the Hemolytic-Uremic Syndrome. *Am J Dis Child* 1980;**134**:869–72.
- ⁵ Upadhyaya K, Borwick K, Fishaut M, Kashgarian M, Siegel NJ. The importance of non renal involvement in hemolytic uremic syndrome. *Paediatrics* 1980;**1**:115–20.
- ⁶ Crisp DE, Siegler RL, Bale JF, Thompson JA. Hemorrhagic cerebral infarction in the hemolytic-uremic syndrome. *J Paediatr* 1981;**99**:273–6.