tend to look alarming but surgical measures do not seem to be required unless penetration of the sagittal sinus occurs. However, lesions are occasionally very large and measure up to 10 cm in length from anterior to posterior fontanelle. As the extensive eschar on these separates during the first month of life there is more of a risk of haemorrhage, and very early excision of the membrane and covering by rotation of skin flaps has been suggested as the treatment of choice (Lynch and Kahn, 1970). Good growth of hair has been reported after this procedure.

Summary

A family with multiple congenital scalp defects, over two generations and probably genetically determined, is described. Although alarming in appearance, surgical intervention is not indicated at least for small lesions. The risk of haemorrhage and meningitis is emphasized.

We thank Dr. Gerald Neligan, University of Newcastle upon Tyne, for referring the case.

References


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Reye’s syndrome associated with acute tubular necrosis

We report the cases of 2 children with Reye’s syndrome (acute encephalopathy and fatty degeneration of viscera), both of whom developed acute renal failure, a hitherto unreported complication, and in one of whom the syndrome followed an influenza A virus infection.

Case reports

Case 1. A boy aged 6–25 years, became ill with anorexia, diarrhoea, and vomiting. 3 days later he had a grand mal convulsion and was admitted to hospital. His past history included the following. (a) Neonatal convulsions thought to be secondary to a cerebral haemorrhage; he had been left with a left hemiparesis, but his intellectual progress had been normal. (b) Two grand mal convulsions at the age of 3 years, since when he has been taking phenobarbitone.

On admission to hospital he was afebrile and had further convulsions, between which he did not regain consciousness. A lumbar puncture showed clear, cell-free cerebrospinal fluid (CSF) under normal pressure, with a protein content of 15 mg/dl. Blood glucose and calcium were normal. There was no clinical evidence of infection. He was later transferred to The Hospital for Sick Children, London. On arrival he was deeply comatose, with generally increased muscle tone and signs of his pre-existing hemiplegia. The liver was palpable 1 cm below the costal margin. Blood pressure was normal. Haemoglobin was 13·8 g/dl, white cell count 6800/mm³, blood urea 136 mg/dl, potassium 5·4 mEq, sodium 131 mEq, and chloride 99 mEq/l. The platelet count was 83 000/mm³, prothrombin time 39 s (control 14), thrombin time 12·5 s (control 9). Fibrin degradation products were not detected in the blood. Repeat CSF analysis was normal apart from a protein content of 90 mg/dl. The blood lead level was normal; calcium 8·9 mg/dl, bilirubin 2·4 mg/dl, serum aspartate aminotransferase 4300 IU/l, serum alanine aminotransferase 4075 IU/l, blood ammonia 90 μg/dl, and a C3 level of 20% of the standard reference serum. Viral studies showed no evidence of a recent infection with herpes simplex, mumps, measles, or influenza A or B viruses. An electroencephalogram showed severe generalized abnormalities with a gross excess of slow-wave activity.

He initially passed small quantities of urine containing blood and protein, but within 68 hours of admission he developed frusemide-resistant anuria. Peritoneal dialysis was instituted. A renal biopsy performed 2 weeks later showed acute tubular necrosis with tubular regeneration. Renal function recovered spontaneously after 3 weeks’ dialysis, and liver function tests reverted to normal. He has remained in coma, with frequent convulsions, and the signs of a spastic quadriplegia.

Case 2. A boy aged 10–25 years, became ill with generalized abdominal pain, nonproductive cough, and pyrexia. 24 hours later he had a prolonged generalized convulsion and was admitted to hospital. There was evidence of influenza A in the local community at this time. His history included (a) hypothyroidism diagnosed at the age of 9 months, since when he had received replacement thyroxine, (b) convulsions with fever at
the age of 5 years, after which he was maintained on phenobarbitone.

On admission to hospital he was hyperventilating, and unconscious without focal neurological signs. Blood pressure was normal. Relevant laboratory results included normal plasma electrolytes, blood urea 50 mg/dl, blood glucose 55 mg/dl, plasma salicylate zero, and plasma ammonia 76 µg/dl. CSF was entirely normal, and blood cultures were negative.

He developed diuretic-resistant oliguria and was transferred to Guy’s Hospital. On arrival he was unconscious, and oedematous with a blood pressure of 140/80 mmHg. Blood urea 182 mg/dl, plasma creatinine 9.7 mg/dl, plasma sodium 122 and potassium 6.8 mEq/l, total bilirubin 3.2 mg/dl, platelet count 73 000/mm³, prothrombin time 29 s (control 12), thrombin time 21 s (control 12), fibrin degradation products present at a serum dilution of 1:20, aspartate aminotransferase 550 IU/l. A blood screen for drugs and poisons was negative apart from the presence of barbiturates at a therapeutic level. C3 complement level was 28 mg/dl.

Nose and throat swabs, faeces and specimens of CSF were negative for viruses by routine culture methods using primary monkey kidney, Hep 2 and W.1.38 cells. The results of serological tests for influenza A virus are shown in Table.

He became anuric and was treated for 3 weeks with peritoneal dialysis. A dynamic renal scintillogram, intravenous urogram, and renal biopsy all showed changes consistent with acute tubular necrosis. Liver biopsy showed a normal lobular pattern with swelling of hepatocytes. A few cells contained fatty vacuoles. Renal function returned after 3 weeks and 2 weeks later it had become normal. However, there was a gradual deterioration in his nutritional and cerebral state. 2 months after the onset of illness he developed bronchopneumonia and sepsicaemia and died without regaining consciousness.

At necropsy there was a large pale liver (weight 1225 g) with moderate diffuse fatty changes; absent thyroid tissue; a fungal granulomatous lesion in the right frontal lobe (thought to be an aspergillosa); and slight fatty change in the renal tubular cells.

Discussion

Renal histology in fatal cases of Reye’s syndrome shows some fat deposition in the tubular cells (Mann et al., 1962). In the original cases reported by Reye, Morgan, and Baral (1963), and in Becroft’s (1966) subsequent series the blood urea was frequently raised. However, oliguria was not reported and renal histology at necropsy showed no evidence of acute tubular damage.

Both our cases showed the following features of Reye’s syndrome: a prodromal illness, a clinical picture of encephalopathy with convulsions, transitorily raised serum transaminases with minimal rise in bilirubin, abnormal coagulation tests (Schwartz, 1971), and raised blood ammonia levels. In Case 2 there was marked hyperventilation, and the liver histology was considered compatible with a diagnosis of Reye’s syndrome.

Case 2 also showed evidence of a recent influenza A virus infection. Influenza B virus has usually been associated with Reye’s syndrome, and some instances have been described where there was evidence of a dual infection with influenza A and B viruses (Linneman et al., 1974), but this is the first case described in England where influenza A alone has been implicated.

Although in neither case was there hypoglycaemia, this is said to occur infrequently in children over the age of 5 years. Thus the clinical and biochemical picture of Reye’s syndrome in both these children was associated with an episode of acute renal failure. Renal biopsy confirmed the diagnosis of acute tubular necrosis and there was recovery of renal function after a period of peritoneal dialysis.

The cause of Reye’s syndrome is obscure, but many toxic agents including viruses, drugs, and alcoholic compounds have been suggested as aetiological factors. There has been nothing to suggest that ingestion of barbiturates in therapeutic levels may be implicated. Case 2 had a cerebral aspergilloma, whose cause, relation to his illness, and duration could not be determined. In Thailand, Olson et al. (1971) have suggested that aflatoxins, which are produced by various species of aspergillus, may be responsible for causing Reye’s syndrome.

<table>
<thead>
<tr>
<th>Date of serum</th>
<th>Influenza A complement fixation test</th>
<th>Haemagglutination inhibition test against A/ENG/42/72 (current influenza A strain)</th>
<th>IgM test for influenza A antibodies (immunofluorescence)</th>
</tr>
</thead>
<tbody>
<tr>
<td>6 March 1974</td>
<td>1/10</td>
<td>1/32</td>
<td>Not tested</td>
</tr>
<tr>
<td>22 March 1974</td>
<td>1/60</td>
<td>1/256</td>
<td>Positive</td>
</tr>
<tr>
<td>1 April 1974</td>
<td>1/80</td>
<td>1/512</td>
<td>Not tested</td>
</tr>
</tbody>
</table>
Short reports

Summary

Two cases of Reye's syndrome, complicated by acute reversible renal failure, are presented. One case followed an influenza A virus infection.

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References


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Life tables for cystic fibrosis

In this study a comparison is made between the survival rates obtained from life tables of 215 children attending The Hospital for Sick Children during the 5 years from 18 January 1969 to 31 December 1973, and the two previous series reported from this hospital (Mantle and Norman, 1966; George and Norman, 1971). The life tables were derived according to accepted statistical methods (Hill, 1966), which have been described in the previous papers. Criteria for acceptance were as in previous studies. Children who were seen purely for confirmation of diagnosis and who were not followed up have not been included.

Cystic fibrosis not presenting as meconium ileus

In the present series 35% were diagnosed before the age of 6 months compared with 26% in the previous 5 years. 46% were diagnosed by one year in the present study compared with 43% in the earlier ones. There was only one death under the age of 7 years in the present (1969–73) series. Mortality was diminished at all ages (Fig. 1) and the gap between the proportion alive at the end of each period when entered at diagnosis, which tends to overestimate mortality, and when entered at birth has narrowed considerably up to the age of 15. After this age the data become less meaningful due to losses from transfer and death.

Cystic fibrosis presenting as meconium ileus

Of the 55 cases in this group the obstruction was relieved by Gastrografin enema in 5 infants, by laparotomy alone in one, and by small intestinal resection usually with a Bishop-Koop anastomosis and ileostomy, in 49. 91% were alive at one month compared with 81% in the preceding survey; 85% at one year compared with 68%; and 79% at 5 years compared with 54% (Fig. 2). However, cases presenting with meconium ileus have a poorer life expectancy than those in the other group, even excluding those dying during the first 6 months (Fig. 3).

Discussion

There has been an improvement in life expectancy in both groups, which is most marked up to 5 years in the meconium ileus group and up to 7½ years in the group with no meconium ileus. Death in the meconium ileus group is usually due to pulmonary sepsis, but this is closely associated with poor early progress and a prolonged stay in an acute hospital ward, as a result of extensive intestinal resection and consequent malabsorption and malnutrition. It is likely that those cases that escape with a Gastrografin enema or simple laparotomy only, fare very much better, but the numbers are too small for statistical analysis.

The results, though confirming that cystic fibrosis remains a serious condition with a limited life expectancy for many of the older children, give good reason to expect that life expectancy will continue to improve for the more recent cases, given earlier diagnosis and effective management with avoidance of serious respiratory infection in the early years. Unfortunately, the present data do not indicate any trend to earlier diagnosis after the first 6 months. Although there is a clear clinical impression that early diagnosis is of major importance in the prevention of serious respiratory infections, the life expectancy of those presenting during the first 12 months is no greater than the average for all cases (Fig. 4). The value of early diagnosis will only be settled when neonatal screening with effective follow-up has been carried out in a co-ordinated manner over a wide region.

Summary

The past 5 years have seen a continuing improvement in life expectancy in cystic fibrosis.