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CLINICAL QUIZ

A confused child: Question

Emer Fitzpatrick · Philip Mayne · Denis Gill

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Case summary

A 6-year-old boy presented to the casualty department at 09.00 hrs, having woken up the same morning 2 hours prior to arrival, confused, agitated and vomiting.

On arrival at the hospital, he was unresponsive to pain. He had a Glasgow coma scale score of 4. His pupils were dilated, but reactive. He was apyrexial (36.7 degrees) and remained so. Blood pressure was 160/100 mmHg, pulse rate 98/min, respiratory rate 20/min. On examination he was moderately dehydrated, had an active precordium, but no murmur. Chest was clear to auscultation; his abdomen was soft with no masses or organomegaly. He had increased tone bilaterally with extensor plantar responses. There was no rash or neck stiffness. His fundi were unremarkable on examination. There was no history of trauma or drug ingestion, and he had been last well the previous day. He was intubated, commenced on intravenous fluids and given aciclovir and antibiotics based on a clinical diagnosis of encephalopathy.

Past medical history included recurrent urinary tract infections, severe vesico-ureteric reflux and and ureteric tapering and reimplantation 2 years previously. On his last admission, his plasma urea and electrolyte levels had been normal. He was on no regular medication.

Laboratory investigations showed a plasma glucose of 11 mmol/l, haemoglobin 13.1 g/dl, white cell count 11.7×10^9/l, neutrophils 9.3×10^9/l and platelets 613×10^9/l. Plasma sodium was 144 mmol/l, potassium 4.0 mmol/l, chloride 108 mmol/l, urea 19.9 mmol/l, creatinine 101 micromol/l, calcium 2.48 mmol/l and phosphate 1.32 mmol/l. His serum ammonia concentration was 244 micromol/l (reference range<40 micromol/l). A venous blood gas showed a pH of 7.36, with a bicarbonate of 18.7 mmol/l.

Coagulation was normal with a prothrombin time of 14.9 seconds and an APTT of 29.3 seconds. Plasma albumin was 51 g/l, liver function tests were normal. C-reactive protein /ESR were not available. Serum amino acids and urinary organic acid analysis were later reported as normal. CSF analysis showed no white cells or red cells and was sterile. CSF glucose was 5.6 mmol/l (plasma glucose 11 mmol/l), CSF lactate was 2.5 mmol/l (plasma lactate 1.52 mmol/l). CSF protein was 280 mg/l (0–460). CSF amino acids showed an elevated glutamine of 2268 micromol/l (421–597). CSF & serum PCR for meningococcus, pneumococcus and Herpes simplex were negative. Urinalysis revealed a urinary pH of 8.5, with 250 white cells, no red cells and three plus organisms. A renal ultrasound demonstrated bilateral hydronephrosis and hydroureters. Computerised tomography (CT) of his brain was normal.

Questions

1. What is the likely explanation for this boy's encephalopathy?
2. What are the predisposing factors leading to and pathogenesis of this condition?
3. What is the usual culprit?
4. What is the management?

E. Fitzpatrick · D. Gill (✉)
Department of Paediatric Nephrology,
Children's University Hospital,
Temple Street,
Dublin 1, Ireland
e-mail: gilld@iol.ie

P. Mayne
Department of Clinical Biochemistry,
Children's University Hospital,
Temple Street,
Dublin 1, Ireland