Cerebral atrophy and nephropathic cystinosis

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SUMMARY The management of end stage renal failure in cystinotic children is correlated with a longer survival, sometimes complicated with neurological abnormalities. Cranial computed tomography was performed in 10 patients and showed a significant atrophy; the pathogenesis of this damage remains unclear.

The full description of cystinosis is yet to be completed as until the last 15 years all those patients with cystinosis died early from end stage renal failure; central nervous system involvement was not often reported as a late complication. The patients' survival over the second or third decade may be complicated with neurological manifestations, and it has been shown that some patients with nephropathic cystinosis have abnormalities on cranial computed tomography.

Patients and methods

Several types of neurological symptoms were noted in 10 cystinotic patients (group 1; mean (SD) age 14.2 (4.0) years), such as repeated seizures, tremor,
mental retardation, and pseudobulbar or pyramidal syndrome. Ten control patients with another primary renal disease (group 2; mean (SD) age 11-8 (3-7) years) had no cerebral abnormalities.

To decide if these features were related specifically to cystinosis or to severe chronic uremia and/or its treatment, we studied the cranial computed tomograms of the patients in both groups. Both groups included dialysed or transplanted children without significant difference in length of end stage renal failure management (4-9 (SD 3-9) years in group 1 v 4-4 (2-3) years in group 2) or in parathormone and aluminum plasma concentrations. Treatment with steroids is known to induce an appearance on computed tomography of a brain that looks shrunken; such treatment had been used for some subjects in both groups (for transplantation, nephrotic syndrome, etc) without significant difference in cerebral computed tomogram data. Most of the cystinotic patients had mild hypothyroidism controlled by hormonal treatment.

Cerebral atrophy was evaluated by quantitative indexes of the ventricle:brain ratios on two distinct computed tomography sections: A ratio for the largest representation of the lateral ventricles, and B ratio for the largest representation of frontal horns, including Monro's foramen.4

Results

Figure 1 shows that the mean (SD) A ratio is 0-27 (0-09) in group 1 and 0-14 (0-04) in group 2 (p<0-01); mean (SD) B ratio is 0-15 (0-04) in group 1 and 0-09 (0-03) in group 2 (p<0-01). The brain atrophy is confirmed in cystinotic patients by the enlargement of cortical sulci, as only two out of 10 in group 1 versus eight out of 10 in group 2 had no significant measured widening (Figures 2 and 3).

Discussion

The pathogenesis of this progressive cerebral damage remains unclear. An imbalance in cerebrospinal fluid is a possibility, as cystine crystal deposits have been found in choroidal plexi and meninges at
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autopsy,3,5 cerebral cell atrophy in relation to the metabolic disorder is also a possibility.

We conclude that children with nephropathic cystinosis have cortical and subcortical atrophy after 10 to 20 years of age, and this feature suggests specific central nervous system complications, which are not yet fully understood.

References

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Myocarditis after triple immunisation

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SUMMARY We describe a 3 month old infant who developed myocarditis several hours after diphtheria, tetanus, and pertussis vaccination. The time of occurrence of symptoms, the clinical course, and the negative virological studies suggest a possible cardiogenic adverse reaction to the vaccine.

Severe systemic adverse reactions after diphtheria, tetanus, and pertussis vaccination are uncommon and include extreme irritability, collapse or shock like episodes, convulsions, and encephalopathy.1,2 Cardiac side effects associated with this vaccination have been reported rarely and are most probably related to the pertussis component of the vaccine.3,4 To the best of our knowledge, myocarditis after the vaccination has not been reported. We describe a case of acute myocarditis that developed a few hours after diphtheria, tetanus, and pertussis vaccination.

Case report

A 3 month old infant was admitted to our hospital 24 hours after receiving his second diphtheria, tetanus, and pertussis and oral polio vaccination because of severe respiratory distress and cyanosis. The infant was delivered at term, weighing 3250 g, and had had an uneventful neonatal period. The first diphtheria, tetanus, and pertussis and trivalent oral polio virus immunisation was administered at 6 weeks without any adverse reaction. The parents stated that the child had been normal and playful until 12 hours after the second administration of the vaccines, when irritability and mild respiratory difficulties appeared.

On admission, physical examination revealed a 3 month old, well nourished, well developed, acutely ill infant. The temperature was 38.5°C, pulse rate 200 beats/minute, and respiration rate 150/minute. His colour was ashen, and a mild oedema of the extremities was evident. On auscultation the heart sounds were of poor quality, occasionally a gallop rhythm was heard, and no murmurs were audible. The lungs were clear, and the liver edge was palpated 5 cm below the right costal margin. All the peripheral pulses were weak. The capillary blood gases showed metabolic acidosis with pH 6.7, carbon dioxide tension 28 mm Hg, oxygen tension 67 mm Hg, base excess -28 mM/l, and bicarbonate 5 mM/l. White blood count was 20.6×109/l, with 70 per cent lymphocytes; haemoglobin 93 g/l; serum urea nitrogen and creatinine 14 mmol/l and 90 mmol/l, respectively. The sodium, potassium, and calcium concentrations were normal.

The serum creatine phosphokinase activity rose abruptly to 348 IU/l (normal is 5-80 IU/l) on the second day after admission, reflecting myocardial damage. An electrocardiogram disclosed low voltage QRS complexes on limb leads and non-specific ST changes and T wave flattening on precordial leads. X ray film showed generalised cardiac en-