

Transient Hyper-Ammonemia of Newborn; a case report

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Citation

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Abstract

CASE

A 5 days old neonate, born in hospital, by vaginal route, at 35 weeks of gestation, to a primi-gravida mother, with regularly supervised antenatal period. There was no history of consanguinity, maternal or obstetrical illness. Chief complaints on day of admission were refusal to feed, poor sucking, and lethargy for 2 days. Fever, decreased urine output, icterus, vomiting, unconsciousness and seizures were absent. APGAR score was 8, 9 at 1 and 5 minutes. The child weighed 2030 grams at birth and was exclusively breastfed till admission. On examination

his vital parameters, head to toe examination, anthropometric parameters and systemic examination was normal. Neuro-cutaneous markers were absent. Primitive reflexes: Rooting & Sucking, Moro's, Grasp and cry were weak. A working diagnosis of near term neonate with late onset sepsis was made but all the investigations for sepsis, urinary infection and meningitis were negative. Treatment in form of antibiotics was started while continuing oro-gastric feeds.

In next 24 hours there was episode of generalized clonic seizure of 10-15 seconds duration followed by post seizure shallow respiration and subsequent respiratory failure needing mechanical ventilation. The child was lethargic with hypotonic, shock was absent. Inborn error of metabolism was thought of. Investigations showed serum Ammonia-3690 µg/dl, respiratory alkalosis and anion gap of 22 meq/l. Urine for ketones and reducing substance was negative, liver function and kidney function test were normal. Management of urea cycle defect was started in form of Sodium benzoate 250mg/kg/day and Arginine 400 mg/kg/d and dialysis was planned. The child became responsive within 12 hours of medical management and could be extubated in next 24 hours. Repeat serum ammonia 3 days later was 264 µg/dl. It showed a downward trend on weekly assessment.

Breastfeeds were resumed to total in 7 days and child was discharged on sodium benzoate and arginine. Tandem mass spectroscopy, TMS later came normal. Serum ammonia normalized by 3 weeks. Levels of serum ammonia remained normal even after stopping sodium benzoate and arginine. Electroencephalogram, EEG showed no seizure activity and Magnetic resonance imaging, MRI was also normal. The baby had normal growth and development till a follow up of 9 months of postnatal age.

DISCUSSION

In our case initial presentation and evidence was in support of urea cycle defect. The rapid response to medical management, normal TMS and urine amino-acidogram and normal values of serum ammonia after stopping the treatment all supported the diagnosis of transient hyperammonemia of neonate.

Transient Hyperammonemia of neonate, THAN was first reported in 1978 (34-36 wks). Ammonia returned to normal in 72 hours with normal outcome in four out of five babies (1). It presents within first few days of postnatal life in form of severe hyperammonaemia, a comatose state, absence of abnormal organic aciduria, normal activity of urea cycle enzymes and usually, complete recovery. In contrast to some of the hereditary disorders of the urea cycle, no specific structural changes could be found in THAN in liver (2). This indicates that a different biochemical system may be pathogenetically involved in THAN. Transient platelet activation in the portal system is one possible pathogenesis of transient neonatal hyperammonaemia syndrome (3). Other possible hypothesis is there is vascular complication of prematurity caused by shunting of blood away from the portal circulation of the liver into the systemic circulation with subsequent lack of ammonia removal (4).

A nationwide survey of transient hyperammonemia in newborns was carried out in Japan. A total of 18 patients 12

male and 6 female infants, were reported. The multivariate analysis revealed that the Apgar score at 1 minute, peak plasma ammonia concentration, birth weight and sex were significant factors affecting the prognosis of life (5). Rapidity of Toxin removal (ammonia) is important in preventing mortality. Most relevant indicator for prognosis is coma duration before the start of dialysis (6). It is not associated with short-term or long-term neurologic deficits (7). In our case also child was neurologically normal till 9 months of age.

This condition has been earlier reported in literature (1-7) and was reported from India also first in 1990 (8), but presentation with such high values of ammonia responding rapidly to medical treatment within 12 hours without dialysis is not reported.

CONCLUSION

Transient neonatal hyperammonemia can present in early postnatal life with value as high as 30 times to the normal.

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