LETTER TO EDITOR (VIEWERS CHOICE)

PSEUDOHYPOALDOSTERONISM IN A NEONATE

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A male neonate was admitted to the nursery on Day 7 of life with neonatal jaundice (serum bilirubin 19.3 mg%). Baby was kept on phototherapy and breast feeding was continued. Twenty four hours after admission the child started vomiting, was noticed to have cold extremities, weak pulses and peripheral cyanosis. There were no features of virilisation or pigmented scrotum. Child was treated with saline boluses and intravenous fluid was started. After sending the sepsis screen and blood culture, IV antibiotics were started. With this treatment child started taking oral feeds. But as soon as the fluids were stopped, child again developed cold clammy extremities. Investigations showed white cell count of 22,300/cumm, serum creatinine of 0.8 mg/dl, serum sodium of 110 meq/l (hyponatremia), serum potassium of 8 meq/l (hyperkalemia), and ionized calcium of 5.38mg/dl. Ultrasound of abdomen showed mild increase in the echogenicity in both the kidneys. 17- hydroxy progesterone levels were normal, serum cortisol was raised [65.27 ug/dl (normal = 3.09-16.66ug/dl)] and serum aldosterone were also raised [> 150 ng/dl (normal =1-16 ng/ml)] suggesting a diagnosis of pseudohypoaldosteronism. The child was treated with salt, IV fluids, fludrocortisone to which he responded. However, the child developed sepsis and died.

Pseudohypoaldosteronism (PHA) should be considered in the differential diagnosis of a salt wasting syndrome in infants, especially when it is accompanied by infections or congenital defects of the urinary tract. (1) PHA is classified into PHA type I (PHA-I), which is the classic form, and PHA type II (PHA-II), which is also referred to as Gordon syndrome or chloride shunt syndrome. Children with PHA-I have hyperkalemia, metabolic acidosis and salt wasting leading to hyponatremia and volume depletion. Serum aldosterone values are elevated. (2) Secondary pseudohypoaldosteronism may resemble congenital adrenal hyperplasia. On clinical basis, PHA-1, has 2 clinically distinguishable entities with either renal or multiple target organ defects (MTOD). (4) PHA-II is a rare familial renal tubular defect characterized by hypertension and hyperkalemic metabolic acidosis in the presence of low renin and aldosterone levels. (4) Early diagnosis is essential since both conditions, when untreated, are fatal, and treatment of the two differs significantly. (3) Patients with PHA who are experiencing hypovolemia and shock should receive fluid resuscitation with isotonic saline. For the correction of hyperkalemia and acidosis, potassium-binding resins, prostaglandin inhibitors, alkalizing agent, hydrochlorothiazide (in PHA type II) is used.

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