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Pseudohypoaldosteronism with Skin Rash

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Introduction

Pseudohypoaldosteronism (PHA) is characterized by electrolyte abnormalities due to renal tubular unresponsiveness to aldosterone. PHA type 1 is a rare disease which can present with renal salt wasting, hypovolemia, hyperchloremic acidosis and hyperkalemia. The autosomal dominant form is milder than autosomal recessive.

Case Report

A male Syrian infant was admitted on day 4 with poor oral intake, excessive crying and multiple episodes of diarrhoea. The patient was severely dehydrated, lethargic, a febrile. There was a papular skin rash covering all the body. Biochemistry showed hyponatremia Na 119.9mEq/L, hyperkalemia K 9.44mEq/L and hypercalcemia Ca 12.6mg/dL. Hyperkalemia was treated, and hydrocortisone and fludrocortisone were started as the results were suspicious of congenital adrenal hyperplasia (CAH). The 17-hydroxyprogesterone was elevated 16.49ng/ml (1.70-4.00), with markedly elevated plasma renin (500uIU/ml (4.4-46)) and aldosterone (100ng/dL (2.2-35.3)). These results and the lack of response to treatment suggested PHA. NaCl 3% was given orally in addition to Na polystyrene. He was admitted for 3 months. Subsequently he has had several admissions with similar clinical picture. There is a family history of early death of two siblings with similar symptoms but no diagnosis was made at that time.

Discussion

In patients presenting with electrolyte disturbances of hyperkalemia and hyponatremia, CAH must be ruled out. However in this case the elevated renin and aldosterone levels led to a diagnosis of PHA despite high level of 17-hydroxy progesterone. A maculopapular rash affecting the face and trunk is a characteristic feature which can accompany this condition.