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SCIENTIFIC LETTER

Hot Summer Shines Light on Diagnosis of Pseudo-Bartter Syndrome

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To the Editor: Cystic fibrosis manifesting as Pseudo-Bartter syndrome is a rare presentation. Pseudo-Bartter syndrome, as the initial presentation, is reported in 9% of cystic fibrosis infants. In patients over 1 year, 12% of cases have been reported [1]. We describe a one-month-old infant who got admitted with recurrent episodes of dehydration, electrolyte derangement and metabolic alkalosis due to excess of chloride loss from sweating. This unusual presentation is noted in cystic fibrosis infants during hot summer climate.

A 1-mo-old exclusively breastfed infant born to primigravida mother by non-consanguineous marriage was referred to our hospital due to severe dehydration, metabolic alkalosis, and electrolyte derangement. There was no family or antenatal history of infant deaths present. There was no previous comorbid illness present. At admission pH–7.56; HCO_3–32; sodium − 125 mmol/L and potassium - 2.1 mmol/L was found. The chest X-ray suggested right upper lobe collapse. There was no history of diarrhea, vomiting or polyuria explaining the severe dehydration noted. Hence at admission, the reason underlying Pseudo-Bartter syndrome and lung collapse was not known. The metabolic alkalosis and electrolyte derangement gradually resolved with severe dehydration correction and hence the infant was discharged and planned for follow-up. The infant was noted to develop recurrent episodes of mild to moderate dehydration, metabolic alkalosis and electrolyte derangement (Pseudo-Bartter syndrome) within 2 wk of discharge. Hence dehydration due to insensible water losses in the form of sweating was considered. This raised the suspicion of cystic fibrosis, which was supported by the persistence of lung collapse in X-ray, low plasma chloride (55 mmol/L), low urine chloride (8.9 mmol/L), high sweat chloride levels (70 mmol/L) and normal blood pressure (65/40 mmHg). Genetic mutation for the delta F508 gene was negative. The infant was treated with oral rehydration solution and nebulization for lung collapse management. The treatment follow-up plan during infancy includes additional sodium intake during hot weather, weight gain monitoring, and immunization (influenza and pneumococcal vaccine).

Cystic fibrosis presenting like Pseudo-Bartter syndrome is a rare entity. These infants are initially asymptomatic but later develop dehydration and Pseudo-Bartter syndrome [2]. The presence of thick viscid secretions predisposes to lung collapse in these infants [3]. Hence one should suspect cystic fibrosis when there is chloride responsive dehydration, metabolic alkalosis and low urine chloride levels due to chloride losses from sweating in hot summer climate [4].

Compliance with Ethical Standards

Conflict of Interest None.

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