Salt losing nephropathy simulating congenital adrenal hyperplasia in infants with obstructive uropathy and/or vesicoureteral reflux – value of ultrasonography in diagnosis


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Abstract. Salt losing nephropathy, occurring predominantly in male infants, has been reported in association with a spectrum of urologic diseases including obstructive uropathy and massive, infected vesicoureteral reflux (VUR). This has been called pseudo-hypoaldosteronism (PHA) or alternatively, pseudo salt-losing congenital adrenal hyperplasia (CAH), and is thought to reflect a tubular unresponsiveness to aldosterone. We report our experience with six cases, discuss one case in detail and review the 39 cases previously reported. A one month old male infant presented with a left upper quadrant mass. Signs and symptoms included vomiting, dehydration, hyponatremia and hyperkalemia. This suggested the diagnosis of CAH for which therapy was instituted. Ultrasonographic examination subsequently revealed the mass to be a urinoma in an infant with posterior urethral valve (PUV) and obstructive hydronephrosis.

Pseudo-hypoaldosteronism (PHA), as originally described in 1958 refers to a salt losing condition seen in infants with vomiting, dehydration, hyponatremia and hyperkalemia. These infants have normal adrenal function and normal renal morphology [1, 2]. A subsequent group of infants have been reported using the same term PHA, or alternatively pseudo salt-losing CAH, in whom a variety of obstructive and non-obstructive renal lesions have been identified [3–6]. Treatment of the urological abnormality often leads to resolution of the salt wasting syndrome. Clinical presentation in these infants is confusing and may lead to an incorrect diagnosis of CAH. We present our experience with six such cases, one of which is described in detail, as well as a review of the literature.

Case report

The findings in Case 1 are discussed in detail. Pertinent information regarding the remaining cases is presented in Table 1.

T.J., a one month old previously well male infant, presented to an outside institution with vomiting and dehydration and was admitted to rule out hypertrophic pyloric stenosis. On admission, a left upper quadrant abdominal mass was palpated. Ultrasonography, limited to the upper abdomen, demonstrated a cystic structure in the left upper quadrant and following a pediatric surgical consultation, the diagnosis of gastric duplication was made. Admission laboratory results revealed hyponatremia, hyperkalemia and metabolic acidosis, which led to a diagnosis of salt-losing CAH.

The infant was transferred to Babies Hospital for endocrinologic treatment of CAH and for surgical correction of the gastric duplication. Upon transfer, the infant was seen by an endocrinologist and received intramuscular cortisone acetate. A repeat ultrasonogram was obtained to evaluate the abdominal mass, and in the setting of possible CAH, to visualize both adrenal glands, and to confirm the presence or absence of a uterus. The ultrasonogram demonstrated a thick-walled bladder, dilated posterior urethra (Fig. 1), bilateral hydrenephrosis, and a left perinephric fluid collection consistent with urinoma (Fig. 2). The urinoma was the source of the initial ultrasonographic diagnosis of gastric duplication. Voiding cystourethrography confirmed posterior urethral valve, and demonstrated bilateral VUR and filling of a left urinoma (Fig. 3).

The patient underwent drainage of the kidney and left urinoma, and transurethral resection of the posterior urethral valve. The patient’s electrolytes subsequently returned to normal and he has done well.

Fig. 1. Longitudinal sonogram of the bladder demonstrates a thickened bladder wall (straight black arrow) and a dilated posterior urethra (curved black arrow)
Table 1. Clinical and laboratory findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Sex</th>
<th>Clinical features</th>
<th>Serum values</th>
<th>Urine Na (mEq/L)</th>
<th>Diagnosis</th>
<th>Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>TJ</td>
<td>1 mo</td>
<td>M</td>
<td>Poor feeding Vomiting Abdominal mass</td>
<td>121</td>
<td>85</td>
<td>PUV Left urinoma Bilateral VUR (Grade 4)</td>
<td>Inappropriate cortisone acetate injection. Open drainage of urinoma. Transurethral resection of valve.</td>
</tr>
<tr>
<td>JR</td>
<td>1 mo</td>
<td>M</td>
<td>Fever Vomiting Irritability</td>
<td>119</td>
<td>-</td>
<td>PUV Sepsis Bilateral VUR (Grade 3)</td>
<td>Inappropriate cortisone acetate injection. Transurethral resection of valve. Bilateral urethral reimplantation.</td>
</tr>
<tr>
<td>JS</td>
<td>3 wks</td>
<td>M</td>
<td>Vomiting</td>
<td>117</td>
<td>75</td>
<td>PUV Right urinoma (Fig 4)</td>
<td>Transurethral resection of valve.</td>
</tr>
<tr>
<td>DB</td>
<td>6 mo</td>
<td>M</td>
<td>Dehydration</td>
<td>124</td>
<td>-</td>
<td>Left ectopic ureterocele Bilateral renal duplication Bilateral VUR (Grade 4)</td>
<td>Left ureterocele excision. Bilateral ureteral reimplantation.</td>
</tr>
<tr>
<td>KG</td>
<td>3 wks</td>
<td>M</td>
<td>Fever Vomiting Left flank mass</td>
<td>111</td>
<td>52</td>
<td>Bilateral renal duplication. Staph. aureus sepsis Bilateral VUR (Grade 4)</td>
<td>Bilateral ureteral reimplantation.</td>
</tr>
<tr>
<td>SW</td>
<td>2 mo</td>
<td>F</td>
<td>Poor feeding Dehydration</td>
<td>116</td>
<td>-</td>
<td>Bilateral VUR (Grade 3)</td>
<td>Bilateral ureteral reimplantation.</td>
</tr>
</tbody>
</table>

Discussion

Fetal hydronephrosis, whether due to obstruction or VUR, is recognized with increasing frequency due to the wide use of prenatal ultrasound. Once the finding is made, post natal confirmation of the diagnosis includes ultrasound, voiding cystourethrography, intravenous pyelography or renal scintigraphy.

There are a group of infants in whom the diagnosis of a primary urologic lesion has not been made prenatally. These infants present days to weeks postnatally with a severe salt-losing renal dysfunction termed PHA or pseudo salt losing CAH. This is characterized by hyponatremia, hyperkalemia, dehydration and metabolic acidosis. Although adrenal function is normal in these infants, their presentation has suggested CAH and has led to inappropriate therapy.

A wide variety of urologic lesions has been reported in association with this salt wasting syndrome [3–6]. These include both obstructive and non-obstructive lesions (Table 2). The obstructive lesions that have been described include PUV, ectopic ureterocele and ureteropelvic junction obstruction. Non-obstructive lesions consist of massive VUR, usually accompanied by urinary tract infection. Reflux may be unilateral or bilateral, or may be segmental as occurs with reflux into the lower pole of a renal duplication.

Assessment of the urologic abnormalities in the reported cases is limited by the lack of imaging studies in the majority of the previously reports [3–6]. These papers have defined “obstructive uropathy” in a very broad sense; included in these cases of obstructive uropathies were a significant number of infants described as having

Fig. 2. Longitudinal sonogram of the left flank shows a large cystic collection compressing the left kidney (arrow). This proved to be a urinoma. Mild dilatation of the left renal collecting system is also shown.