Reminder of important clinical lesson

Renal tubular acidosis and superior mesenteric artery syndrome

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Summary

A 12-year-old girl presented with significant vomiting, and generalised muscular weakness. She had normal anion gap metabolic acidosis, hypokalemia and alkaline urine. Vomiting generally leads to metabolic alkalosis but this patient had acidosis which suggested either renal tubular acidosis (RTA) or diarrhoea. Investigations showed distal RTA. There was no family history of similar illness. Abdominal ultrasound showed features of the superior mesenteric artery syndrome, but a barium study showed no duodenal obstruction, making this an unlikely cause of symptoms. Weight loss as a result of the renal condition may have caused loss of mesenteric or retro-duodenal fat and explain the ultrasound appearance.

BACKGROUND

Renal tubular acidosis (RTA) and the superior mesenteric artery (SMA) syndrome are rare conditions. Their association is so unusual that it warrants reporting.

CASE PRESENTATION

A 12-year-old girl was admitted with a 3-month history of intermittent fever and weakness. She developed recurrent episodes of vomiting a month after the onset of illness. The vomitus contained ingested food and was both bilious and non-bilious. There was no abdominal pain or haemorrhage. She required hospitalisations three times. There was no history of drug intake.

On examination she was thin and weighed 24 kg (expected 45 kg), which was below third percentile, and 146 cm tall (expected 155 cm). Her fingers were long and slender, but she was not dysmorphic or hyper-pigmented. Her pulse was 84 beats per minute and blood pressure was 90/60 mm Hg. She had generalised hypotonia, but physical examination was otherwise normal. Her sexual maturity rating was tanner stage II. The vomiting decreased but on third day she developed marked generalised weakness and was not able to hold her head up or grasp objects. Intercostals muscle movements were normal. Gastro-intestinal obstruction was considered.

INVESTIGATIONS

Urine culture was sterile.

Abdominal ultrasound (figure 1) showed

- ▶ The angle of origin of the SMA (aorto-mesenteric angle) was reduced at about 18° (normal 38–60°).
- ▶ Aorto-mesenteric distance that is the distance between the aorta and SMA was reduced, measuring about 3.6 mm (normal 10–20 mm).
- ▶ Dopplers showed normal flow in the SMA with a peak systolic velocity of 102 cm/s, peak end diastolic velocity of 11.8 cm/s, resistive index 0.88 and pulsatility index 3.02.
- ▶ There was no nephrocalcinosis.

These findings suggested a SMA syndrome.¹

An upper gastro-intestinal barium study showed no dilatation of the duodenum, excluding SMA syndrome as a cause of her symptom.

TREATMENT

She was given intravenously 0.45% sodium chloride with 5% dextrose and 40 mmol/l potassium chloride followed by 0.18% sodium chloride with 5% dextrose.

Her potassium levels and muscle strength improved and she was prescribed sodium bicarbonate and potassium chloride, 2 mEq/kg, each orally.

Table 1 Serum electrolytes and blood gases

Hospital day	Na (mmol/l)	K (mmol/l)	Ca (mmol/dl)	Cl (mmol/dl)	Urea (mmol/l)	Cr (mol/l)	pH	Anion gap	HCO ₃ (mmol/l)
1	130	1.76		94	9	50			
3	131	0.91		93			7.32	22.9	15.7
4	147	2.70		124					
11	136	1.42	2.25	104			7.30	12.2	21.0

OUTCOME AND FOLLOW-UP

Serum potassium level was normal and she was well when she went home.

DISCUSSION

We made a diagnosis of distal RTA on the basis of severe hypokalemia, urinary loss of potassium (>15 mEq/l), hypercalciuria, normal anion gap metabolic acidosis and alkaline urine.² Normal linear growth and severe weight loss suggested an acute insult. Furthermore, there was no nephrocalcinosis and long standing RTA seemed unlikely. Although rare, a dominant form of distal RTA may present in adolescents and be milder than disease presenting in infancy.³

The SMA artery syndrome describes extrinsic compression of the duodenum causing obstruction and occurs in children after rapid weight loss and is most marked when

they lie supine. The compression may occur when the mesentery loses fat and allows the SMA to compress the duodenum, between the SMA anteriorly and the aorta posteriorly. Alternatively, loss of supporting fat in the second and third portions of the duodenum may allow the duodenum to collapse against the spine.⁴

A classic example is an adolescent who starts vomiting after application of a body cast for orthopaedic surgery. Other factors include anorexia, prolonged bed rest, weight loss, abdominal surgery and exaggerated lumbar lordosis. The diagnosis is established radiologically with the demonstration of a cut-off of the duodenum just to the right of the midline. The duodenal obstruction may be accompanied by proximal duodenal and gastric dilatation.⁴ In this child, there was no evidence of obstruction and her vomiting could not be attributed to SMA syndrome, which may have been due to loss of weight and her slim habitus.

Table 2 Urinary electrolytes

Hospital day	Na (mmol/l)	K (mmol/l)	pH	Ca/Cr (normal 0.2 for the age)	Glucose	Proteins	Ketones	Specific gravity
6	56	25	7.0	0.96	Nil	Trace	Nil	1015
11	64	38.2	7.0	–	Nil	Trace	Nil

Figure 1 Sagittal gray-scale image through the epigastrium showing origin of the superior mesenteric artery (SMA) from the aorta. The aorto-mesenteric angle and the aorto-mesenteric distance are greatly reduced.

Learning points

Intractable recurrent vomiting in a child should prompt a search for both obstructive and non-obstructive causes.

- ▶ Metabolic acidosis in presence of severe hypokalemia suggest RTA, diarrhoea or diabetic ketoacidosis as possible diagnoses.
- ▶ Normal Anion gap acidosis and alkaline urine indicates distal RTA.
- ▶ Rarely, RTA may present late.
- ▶ Ultrasonic features of SMA may be found in thin children, who have lost weight and mesenteric or retroduodenal fat but this may not be responsible for symptoms.

Competing interests None.

Patient consent Obtained.

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