Delayed Presentation of Short Bowel Syndrome Complicated with Severe Degree of Nutritional Deficiencies, Nephrocalcinosis and Distal Renal Tubular Acidosis

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Abstract

Background: Short bowel syndrome (SBS) is a malabsorptive state due to functional or anatomic loss of extensive segments of small intestine which can lead to nutritional deficiencies and metabolic disarrangements. Here we describe a young patient with short bowel presenting with severe nutritional deficiencies, nephrocalcinosis and distal renal tubular acidosis. Nephrocalcinosis and distal renal tubular acidosis are closely associated and each can lead to the other. There are only rare case reports of short bowel syndrome complicated with nephrocalcinosis and distal RTA and severe degree of metabolic derangement with nutritional deficiencies. And this case highlights

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low plasma HCO_3^-. He also complained of progressive proximal muscle wasting and weakness with bone pain for the last f feen years and had a fragility fracture of right femur (Figure 1). His growth was stunted and was also found to have a mixed def ciency anemia. He has not attended medical clinics regularly and so was not followed up properly till this admission.

Figure 1: X-Ray right femur showing fragility fracture.

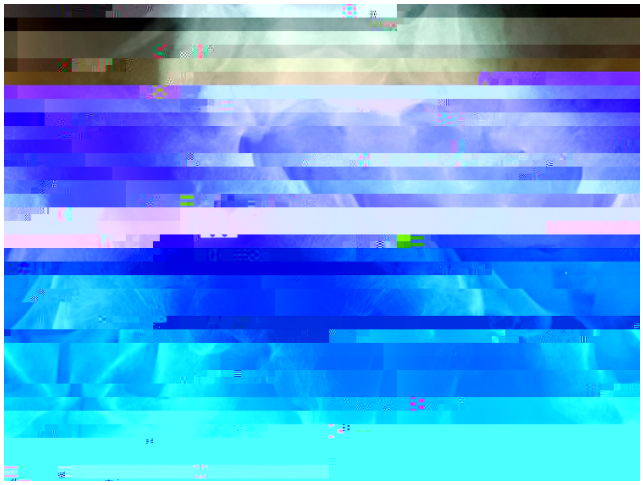

Figure 2: X-Ray pelvis showing loozer zones.

On examination his height was 145 cm and weighted 45 kg with a BMI of 21.4. He was pale but did not have angular stomatitis, glossitis, bitot spots or gum bleeding. His cardiovascular, respiratory and abdominal examination was normal. Cranial nerve examination was also normal with normal fundoscopy. However he had both upper and lower limb proximal muscle wasting and weakness of grade 4 with preserved ref exes. His Heamoglobin was progressively decreasing with a rising MCV. Ten years af er the surgery his Hb was 10.1 g/dl with a MCV of 89 f and at this presentation (af er 22 years later from the surgery), Hb was 6.8 g/dl with a MCV of 105 f. White cell count and platelet count were normal throughout. Blood picture showed hypochromic microcytic RBC as well as normochromic normocytic RBC with ovalmacrocytes, pencil shaped cells and hyper-segmented neutrophils suggesting a mixed def ciency anemia which was con f rmed bio chemically (Table 1). Liver functions were normal with a marginally low albumin and globulin (Table 2).

Serum iron 51.9 µg/dl (59-156)	T.I.B.C. 249 (291-430)
Transferrin saturation 20.8% (20% -50%)	Ferritin 225 ng/ml (28-365).
Serum B12 111.5 pg/ml (223 -915)	Red cell folate 240 ng/ml (252-813)

Table 1: Mixed def ciency anemia, Serum iron studies, B12 levels and Folic acid levels.

Serum creatinine was progressively rising with 84 µmol/l 10 years af er the surgery and now, 22 years later, 257 µmol/l. Serum calcium and phosphate were normal with progressively rising serum Alkaline phosphatase and parathyroid hormone (PTH) (Table 3). Serum vitamin D (25 hydroxy) was low at 10.56 ng/ml (<20 def cient) X-ray pelvis revealed Looser zones (Figure 2).

Albumin of 34 g/L (36-48)	Globulin of 14 g/L (22-40)
AST 18 U/L (10-35)	ALT 30 U/L (10-40)
Total bilirubin 13 µmol/L (5-21)	INR 1.4
Amylase 70 U/L (22-80)	

Table 2: Liver function tests.

Serum potassium was low initially (3.3, 3.7 and 3.4 mmol/l) prior to starting potassium replacement and replaced to a level of 4.4 mmol/l. Other ions were within normal range with serum sodium 138 mmol/l and serum Chloride 110 mmol/l. Urine sodium was 62.2 mmol/l (14-144), urine potassium 14.4 mmol/l (13-62) and urine Chloride 58.0 mmol/l (55-125). Arterial pH showed acidosis with pH values of 7.3, 7.28, 7.32 with the urine pH persistently above 5.5 (6 - (ů ny3wea

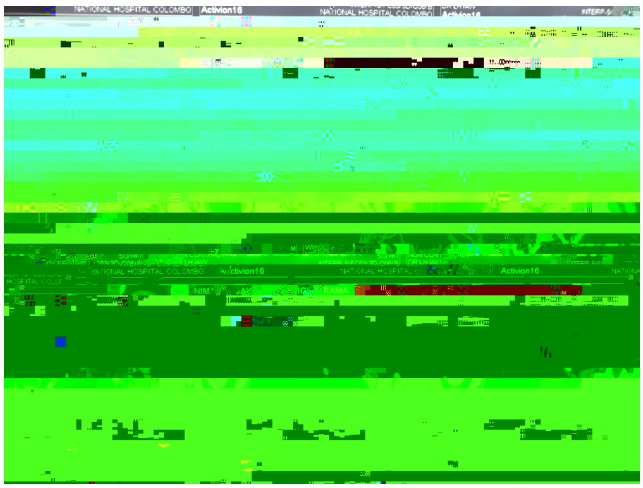

Figure 3: NCCT KUB showing bilateral nephrocalcinosis.

Renal stone analysis was positive for calcium, oxalate and cysteine and negative for uric acid, ammonia, magnesium, phosphate and carbonate.

Time following surgery	10 years	14 years	20 years	22 years
Serum ionized Calcium (1.12 -1.32)	1.20	1.0	1.2	1.14
Serum Phosphate mg/dl (2.5-4.3)	2.99	2.8	2.9	2.1
Serum Alkaline phosphatase µmol/l (100-360)	399	494	718	694
Intact PTH (14-72) pg/ml	92.6	Not analyzed	Not analyzed	349

Table 3: Serum ionized calcium, phosphate, alkaline phosphatase and intact PTH.

ESR was normal. Sigmoidoscopy performed was normal and biopsy revealed nonspecifc colitis only. ANA, Rheumatoid factor, C-ANCA and P-ANCA were negative. TSH 1.58 mIU/l (0.3-4.2) Fasting blood sugar and lipid profle were normal. HIV 1 and 2 and VDRL were negative.

Discussion

T e short bowel syndrome (SBS) is a malabsorptive state of both macronutrients and micronutrients inadequate to maintain his or her nutrient and hydration status without intravenous or enteral supplementation that may follow massive resection of the small intestine [1] SBS in adults usually results from surgical resection of bowel for Crohn's disease, malignancy, radiation, vascular insu f ciency and in infants and small children it can be due to necrotizing enterocolitis and congenital intestinal anomalies such as atresia or gastroschisis. Adult small intestine is approximately 480 cm and adults with residual small intestine of less than 180 cm are at risk for developing SBS and its complications and those who have less than 60 cm of small intestine (but with a colon) are more likely to be dependent on parental nutrition [2]. In our patient about 240 cm of the small bowel had been resected as a complication of appendicitis a f er which he was largely lost to follow up until he developed complications of SBS.

Chronic complications of short bowel syndrome include malabsorptive nutritional abnormalities including metabolic bone disease, mixed de fciency anemia, small bowel bacterial overgrowth, D-lactic acidosis, chole fs

oxalate stone formation and nephrocalcinosis. So we believe our patient develop nephrocalcinosis secondary to hyperoxalruia and later contributed by distal RTA. He had urolithiasis due to hyperoxaluria, hypercalciuria and hypocitrateuria [9] caused by distal RTA, and hyperuricemia caused by CKD. His urine stone analysis was positive for calcium, oxalate. Urolithiasis and hypocitraturia secondary to distal RTA led to recurrent urinary tract infections. In here we were unable to confrm hyperuricemia, hyperoxaluria and hypocitraturia by measuring twenty-four hour urinary studies due to fnancial di f culties.

Our patient had osteomalacia with bone pain, muscle pain, bone tenderness, a fragility fracture and proximal muscle weakness. In this case osteomalacia is due to impaired vitamin D absorption, impaired calcium absorption, CKD (reduced formation of 1,25-dihydroxyvitamin D, metabolic acidosis) and distal RTA (causing metabolic acidosis and hypercalciuria). Here the serum calcium was almost normal due to secondary hyperparathyrodism and metabolic acidosis causing bone resorption and release of calcium. But increased serum alkaline phosphatase (elevated in 95 to 100 percent), PTH (elevated in 100 percent) and low 25-hydroxyvitamin D <15 ng/ml (low in 100 percent) and looser zones in X-ray confrmed the diagnosis. Hypercalciuria is defned as urinary excretion in excess of

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