Case Report

The Challenges of Managing and Following-up a Case of Short Bowel Syndrome in Eastern Europe

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Abstract

The problems faced by hospitals in the developing countries and how they handle difficult and complex cases as in short bowel syndrome is usually not discussed in international journals. The aim of this paper is to attract attention to the plight of patients with rare and complex diseases in Eastern Europe. Data’s were collected and analyzed from 3 different hospitals in two different countries from November 2013 to 2016. Patient had an emergency surgery for intestinal malrotation (volvulus), as a result of extensive ischemic necrosis, with just 80 cm of the bowel without ileocecal valves after surgery. The resultant short bowel syndrome symptoms forced parents to seek medical help in a bigger regional hospital. But despite intensive care and subsequent surgeries with just 70 cm of the intestine left, patient remained in catabolic state and was transferred to a neighboring western hospital where two more surgeries and intensive care helped patient to finally achieve enteral feeding at the optimal time. The collaboration between our hospitals and the pediatric unit across the border was the last hope that helped save this patient’s life at the last minute. The major problem is the lack of sufficient mucosal surface in order to gain enteral nutrition, as enteral feeding is tolerated TPN was gradually weaned proportionally, it should be noted that fluid and electrolyte imbalance is frequent during this process, so hydration and serum electrolyte level should be closely monitored and corrected promptly.

INTRODUCTION

We sought to follow up an individual case of short bowel syndrome (SBS) and to show what patients with similar cases have to go through in a developing countries with less healthcare facilities or under financed healthcare system. These cases are rare and complex so many healthcare planners in government don’t give it the attention it rightly deserves. The complex nature of our case makes the management and follow up pain staking, but the successful outcome makes the obstacles encountered worthwhile. Medical records were gathered from 3 different hospitals in 2 different countries and we followed up this case from November 2013 to 2016, this wasn’t an easy task. Patient’s weight and height were regularly monitored, while hypercaloric and hyperproteic diet were very well structured with the aim to improve nourishment and to gain weight. Follow-up blood work was carried out every forth night till date. Patient initially was placed on total parenteral nutrition (TPN), then followed by parenteral nutrition (PN) combined with enteral feeding, later progressed to just enteral feeding. Continuous sip of oral rehydration should be noted. Life saving surgeries played a key role in patient’s survival. We wish to guide our readers through the complex and complicated nature of this case and all odds being against this patient coupled with a life threatening enterocolitis and salmonella infection 2 years after surgery, patient survived. Also the roles parents play at home with reference to continuous

outpatient treatment should never be overlooked. The high cost of managing a SBS patient and the complicated nature of this disease makes it more difficult and life threatening for patient’s from developing countries and less financed healthcare system such as ours. Most of these patients die because of the inadequate monitoring system and lack of fund for rare diseases. There is no close coordinated interdisciplinary collaboration among caregivers which includes pediatric surgeons, gastroenterologist, nutritionist, psychologist, nurses, parents, and volunteer’s [1]. The only option left for these patient’s is to seek extra medical help from the west if the parents can afford it otherwise they are simply left to fate or to the little the healthcare system can offer which is often inadequate. The collaboration between our hospital and the pediatric unit in Graz was the last hope that helped save this patient’s life at the last minute. Short bowel syndrome can be defined as a patient requiring PN for more than 42 days due to gastrointestinal intolerance and residual small bowel length of less than 25% of predicted gestational age [2].

**CASE PRESENTATION**

A 6 yrs old, female patient was brought in a serious condition, abdominal pain, fever, nausea, bilious vomiting, no passage of stool, abdominal distention and tachycardia. No history of surgery. Abdominal ultrasound and CT scan showed signs of intestinal obstruction.

Emergency surgery was performed while common mesentery, intestinal obstruction and volvulus with extensive ischemic necrosis of the ileocecal intestinal loop were discovered. Intestinal resection followed by side to side ileocolic anastomosis was performed. Remaining intestinal loop was just 80 cm. Patient postoperative status in ICU continued to deteriorate. (Blood pressure 70/50, urine output for 24h 400ml, gastric tube 300 ml of blood strained fluid, central venous catheter was placed, Blood gas: Ph 7.24, CO2 46, pO2 25, Na 140, K 6.3 blood sugar 85, Lac 2.5, Ca 0.91, HC03 19.7 BE 7.7, Hem 9.9).

Pathology result: Macroscopic; Ileum and jejunum 70 cm long, dark pinkish color, hemorrhagic serosa with ulcerorehrea greenish content, multiple mesenteric adenoapathy. Lymphatic ganglion 4/3 cm at ileocecal angle, subileal ganglion group of 0.5-2cm diameter, of dark pinkish color. Appendix of 10 cm. Omentum of about 12/15 cm. Microscopic aspect: 1. Ulcerative ileocejunocecal mucosa, with intestinal villi and ulcerative lesions, lymphoid follicles, lymphocytic infiltration of submucosa and mucosa, thrombotic vessels of the sub mucosa and the intestinal wall lesions mimicking enterocolitis follicles overlapping the ischemic lesions. 2. The 16 mesenteric lymphatic gangions have wall lesions mimicking enterocolitis follicles overlapping the mucosa, thrombotic vessels of the sub mucosa and the intestinal ileojejunocolic mucosa, with intestinal villi and ulcerative lesions, Omentum of about 12/15 cm. Microscopic aspect: 1. Ulcerative ileocejunocecal mucosa, with intestinal villi and ulcerative lesions, lymphoid follicles, lymphocytic infiltration of submucosa and mucosa, thrombotic vessels of the sub mucosa and the intestinal wall lesions mimicking enterocolitis follicles overlapping the ischemic lesions. 2. The 16 mesenteric lymphatic gangions have wall lesions mimicking enterocolitis follicles overlapping the mucosa, thrombotic vessels of the sub mucosa and the intestinal ileojejunocolic mucosa, with intestinal villi and ulcerative lesions, Omentum of about 8.3 cm, intestinal lumen of about 1.6 cm, Ash color with brownish elastic areas. Microscopic Intestinal fragment with large surface of thrombotic extravasation, hyperemic vessels, with thrombus of different stages and highly infiltrated polymorphic inflammation (Ischemic type of circulatory modification).

Laboratory analysis: RBC 3,56x10⁷/mm3, HGB 10.1g/dl, HCT 39.9%, PLT 900,000/mm3, Neutrophil 83.9%, WBC 30,12x10⁹/mm3, CRP 65mg/l, PCT 0.32mg/ml, Protein 60mg/dl. Parents requested to be transferred to children’s hospital in Graz. Patient arrived in Graz in a catabolic state with large amount of bilious vomiting, metabolic acidosis, pyrexia, confusion, speechlessness. Vitamin A, D, and E deficiency, selenium deficiency and axial hiatal hernia with gastro esophageal reflux, hepatomegaly. Upper GI study showed good contrast passage with 2 suspicious areas of possible stenosis. Swabs showed multi-resistant germs (Klebsiella pneumoniae) requiring antibiotics (imipenem and amikacine). Parenteral nutrition, fresh frozen plasma, albumin and balancing of the electrolyte levels helped achieve anabolism, mobilization with the help of physical therapy. Patient continued to loose large amount of bilious fluid requiring further investigation by upper and lower endoscopy (gastroduodenoscopy and sigmoidoscopy) and biopsy. Intestinal adhesion was lysed through a medial laparotomy followed by peritoneal lavage (for colon and abdominal wall abscess), while 2 stomas (jejunostomy and colostomy respectively) were place at the stenotic sites to improve nutrition. Stomas were closed after 6 weeks. Central venous catheter was removed, while patient was discharged with right Broviac catheter. Patient was treated with nephrotans , kalioral and olevitD3. Enteral feeding was gradually introduced and tolerated (dietary food with low lactose and fructose content). Parent’s were instructed on how to continue PN nutrition at home especially at night 7x daily smolkabien peripheral emulsion infusion (at the beginning 16 h iv infusion daily, then 12h iv infusion/24h, 8h iv infusion/24h, 5h iv infusion for 24h, finally 3h iv infusion for 24h respectively for a period of about 18 months). Blood gas analysis is to be repeated
every 2 weeks at patient's country of origin. Gradually patient's condition improved and stool frequency reduced to just 3 stools per day (no more watery diarrhea), and patient went back to school. Patient comes in (at our hospital in Arad) every 2 weeks for blood gas and follow up, gradually patient gained weight and now lives a normal life. Broviac catheter was removed at the end of treatment. Laboratory analysis of patient also improved. Hemoglobin 11.4g/dL, Hem 33.1%, Ly 62%, Chlorine 112mmol/L, phosphate 5.74mg/dL, uric acid 2.2mg/L, Alanine transaminase 58 U/L, fe 58 U/L, transferin saturation 15%, BE 6.4mmol/L, HCO3 18.5mmol/L, pH 7.322, D-Dimere 1.35mg/L APTT 41.1 sec, VitaminD3 25.2 ng/ml, WBC, RBC, Platelets, trace elements, creatinine, other hepatic values, CK, LDH, Blood sugar, Folic acid, Vitamin B12, Cholesterol, Triglyceride, Protein, Ferritin and thyroid values were all within the normal range.

Two years after surgery our patient now 8 year and 4 months was admitted at the pediatric unit of county hospital Arad with the following symptoms: fever, bilious vomiting, watery diarrhea (9 stools per day), she was later transferred to pediatric unit of children's hospital Timisoara as her condition worsened. On admission she was apyretic, but in a serious condition, pale, thin, weak, white tongue, pharyngeal congestion, acetone breath, asthenic thorax, heart beat of 120b/min, no pulmonary rales, painless palpable abdomen, watery diarrhea (9 stools daily), repeated vomiting and absence of meningeal irritation, signs of dehydration >15%, prerenal azotemia, hepatocytolysis (hydroelectrolytic imbalance, metabolic acidosis, hypoponatemia, hypopatassemia), carential syndrome (hyposideremia, hyposideremia) and inflammatory syndrome. Patient was placed on ICU and hydroelectrolytic imbalance was corrected, antibiotics (metamizole) were administered. Ultrasound and x-ray of the thorax and abdomen were normal.

Laboratory analysis: Alanine transamylase 108 U/L, calcium 0.95 mmol/L, creatinine 141mg/ml, ferritin 314ng/ml, phosphate 2.56 mmol/L, magnesium 0.61mmol/L, WBC 183.83 mg/L, sideremia 1.3 umol/L, urea 12.33 mmol/L, CPR 19.48, neutrophil 17.88, lymphocyte 0.84, NEUT% 91.7, LYMP% 4.3, E00% 0.1, IGM 2.31g/L, ESR 80mm/h, procalcitonin 9.43 ng/ml, urine density 1010, urine pH 5, bacteria present, leucocytes present, Diagnosis: Acute gastroenterocolitis, renal insufficiency (prerenal type), hypopotassemia (moderate type), respiratory compensation metabolic acidosis, hepatocytolysis, SBS, urinary tract infection, mild malnutrition, lymphpoenia, iron deficiency. Parent's insisted on transfer to pediatric unit in Graz, as patient health wasn't improving.

Patient was readmitted in Graz for gastro-intestinal infection, watery diarrhea, repeated vomiting, metabolic syndrome, consciousness disturbances (confusion, speechlessness and hypotonic state). The amount of liquid and parenteral nutrition administered at the country of origin was considered inadequate thus worsening drastically patient's health.

Laboratory analysis on arrival: WBC 12.85g/L, neutrophil 8.9g/L, monocyte 1.5g/L, chloride 114mmol/L, blood sugar 110mg/dL, CPR 15.0mg/L, BE 5.8mmol/L. 2nd day: Urine test: erythrocyte 5280/ul, leukocyte 20/ul, protein +++ positive, serum creatinine 1.20mg/dl, P-amylase 293U/L, lipase 156U/L, CRP 6,6mg/dl, monocyte 1.2g/L. Other hepatic and electrolyte values, CK, LDH, blood sugar, serum protein were normal. Stool test (bacteriologic and virusologic): was negative. 3rd day: serum protein: 2065mg/L, protein/gcreatin 864mg/gCrea, albumin 1031mg/L, albumin/gCrea 431mg/gCrea, leukocyte 120/ug, erythrocyte 2880/ul, protein ++++. 4th day ultrasound: SBS, Salmonella infection, renal changes due to infectious toxic nephritis probably caused by salmonella infection. Moderate hepatopatropemonogal, pancreatic edema is also as a result of infection, moderat intra peritoneal fluid, hydrops and gall bladder sludge but no peripancreatic exudate. 8th day laboratory analysis: WBC 17.21g/L, GGT 1120U/L, ALT 42 U/L, P-amylase 196U/L, Lipase 83U/L, renal, hepatic and electrolyte values, CK, LDL, CRP, blood glucose, cholesterol, triglyceride and serum protein values were all normal. Urine test was normal. Abdominal x-ray image: SBS as a result of volvulus. No free intra peritoneal air or fluid, intestinal dilatation (4,7cm). Stool test: No pathogenic bacteria. On the 9th day: Ultrasound of the soft tissues of the neck, Doppler ultrasound of the neck vein and sub clavian vein on both sides. The jugular vein was as well visualized and there was no sign of obstruction, it wasn't possible to continue to right atrium. On the 12th day stool test was repeated and no pathogenic organism was found. The 15th day laboratory test: erythrocyte 1012/L, Hb 9,5, Hct 26.4%, Potassium 3.4mmol/L, Ca2+ 2.17, Mg2+ 0.68mmol/dl, GGT 67 U/L, ALT 41 U/L, P-amylase 78U/L, blood sugar 134mg/dl, other hepatic, renal values, CK, LDL, CRP, cholesterol, triglyceride were all normal values. The 19th day laboratory test: hemoglobin 9.7, creatinine 1.21, others were all normal.

On the 21st day: Spontaneous and palpable abdominal pain: 0 (on a scale of 0-10). At the time of admission patient weighs just 18kg having lost 4 kg of body weight as a result of metabolic imbalance and prerenal insufficiency. PN and ceftriaxone was continued, while the level of hydration was significantly increased. The presence of salmonella in stool was confirmed, and because of the seriousness of the infection antibiotics was changed to Unasyn. Sodium bicarbonate and kalium were administered. Port venous catheter was placed on the 10th day through which PN was continued until the 15th day. Patient was sent home with port catheter should in case supplementary nutrition and hydration is needed (this has to be flushed once every month with saline solution and heparin). Antibiotic was also stopped the same day, and patient tolerated enteral feeding, while the acid base values gradually normalized. Upon discharge creatine level was still a little high and has to be monitored.

DISCUSSION

The term short bowel syndrome is a malabsorptive state that occurs after the resection of a large portion of the small intestine, be it congenital or acquired such as intestinal atresia, enterocolitis, volvulus, gastrochisis, intestinal aganglionicosis, dysmotility syndromes etc. This can also mean the need for prolonged PN as a result of intestinal failure [2]. These patients require long term hospitalization and PN. The major problem here is the lack of sufficient mucosal surface in other to gain enteral nutrition [3]. The degree of malnutrition depends on the remaining intestinal length, this is crucial in determining bowel functional capacity. Our case with just 70cm of the intestine left and with
no ileocecal sphincter capacity, severe malnutrition coupled with watery diarrhea and metabolic acidosis were life threatening and challenging [4]. The management of SBS always starts with a total parenteral nutrition (TPN), this is gradually followed by the combination of PN and enteral feeding [5]. The optimal goal is to achieve full enteral feeding at the optimal time. Long time TPN can lead to intestinal failure associated liver disease (IFALD). IFALD may require at the long run liver and intestinal transplant as the last hope for survival [6,7]. As enteral feeding is tolerated TPN should be weaned proportionally, it should be noted that fluid and electrolyte imbalance is frequent during this process, so hydration and serum electrolyte level should be closely monitored and corrected promptly [8]. Excessive osmotic fluid load is the most common cause of diarrhea, and so continuous enteral feedings coupled with altering enteral type formula to low carbohydrate and high fat formula may help to reduce output [9]. Please note that our patient received a low carbohydrate diet reduced lactose and fructose intake, we also monitored closely the blood gas once every 2 weeks for nutritional support. Lack of enteral feeding leads to gastrointestinal hormones reduction such as cholecystokinin (CCK), motilin, secretin, pancreatic polypeptide, glucose-dependent insulinotropic polypeptide and glucagon, also vaso active intestinal peptide reduces gallbladder contractility and intestinal stasis [10]. Disruption of enterohepatic circulation caused by ileal resection and the absence of enteral feeding leads to hepatocellular bile acid reduction, bile secretion and gallbladder contraction increases the chances of liver disease [11]. Excess lipids and TPN which lacks aminoacids (taurine or cysteine) are linked to IFALD [12]. Biliary disease (sludge, gallstone, acalculous cholecystitis) occur in patients dependent on PN [13]. Restriction of diet is recommended in case of high level of oxaluria. To avoid metabolic bone disease calcium, vitamin D and alkaline phosphate levels should be checked periodically as well as soluble vitamins (A, D, E, K), also vitamin B12. Our patient required follow-up nutritional support (supplemented through the Broviac catheter at home) for as long as 18 months, weight was gained but was again lost due to the gastroenteritis caused by the salmononella infection. With the enterocolitis resolved we gained but was again lost due to the gastroenteritis caused by the salmononella infection. With the enteral feeding started patient has started gaining weight again. The fragile nature of SBS patient should not be underestimated. The best food group to start with that provides less osmotic load is protein and fat, they provide additional stimulant for intestinal adaptation. Adequate PN and later enteral nutrition helps to reduce diarrhea from 12 stools to 3 stools per day. For the maintenance of good hydration and electrolyte balance normal or half saline, potassium, sodium bicarbonate need to be supplemented as indicated by the blood gas [14]. Continuous sip of oral rehydration solution (ORS), through the day helps maintain a positive fluid balance. If enteral route does not meet patient’s fluid need, extra iv fluid may be needed at night [15]. Small bowel biopsies in SBS always demonstrates inflammatory changes, villous blunting, while the present of adherent or intracellular bacteria proves the presence of short bowel bacterial overgrowth [16]. Catheter related blood stream infection (CRBSI) is another source of infection in SBS[17]. It is though difficult to predict the source of infection (Pseudomonas aeruginosa and Klebsiella pneumoniae), it may be as a result of SBBO due to low immunity and the use of proton pump inhibitors. CRBSI is another source of infection in SBS patients. Bacterial translocation has been noted in animal models, but data supporting its occurrence in humans are limited. Bacterial overgrowth and impaired mucosal immunity puts patient’s with SBS at risk of bacterial translocation [18]. Intestinal bacterial growth is controlled by many mechanisms, such as: gastric acidity, pancreatic enzyme activity, enterocyte turnover, normal peristaltic activity and the presence of ileocecal sphincter. In SBS patient’s, these factors are altered and thus bowel dilatation with reduced peristalsis may develop adaptive mechanism to improve enteral adaptation, while these factors may favors bacterial overgrowth by reducing bowel ability to expel microorganisms [19]. Intestinal endotoxines increases in children without ileocecal valve and can also impair liver function by decreasing body’s bactericidal defense mechanism. Fecal lactate concentration in SBS is as high as 60mmol/L, in contrast with just 1mmol/L in normal individuals. Also note that SBS patients have low fecal pH. Elevated d-lactic is a serious complication responsible for acidosis in children with SBS, this condition is associated with confusion, speech disturbance, and severe metabolic acidosis. It is necessary to determine the levels of IgA, IgM and gG. Increase in IgM number and M cell apoptosis are seen in chronic intestinal inflammation and it enhances the uptake of microorganisms. Reduction of IgA levels results in an increased frequency of gastrointestinal infections and impaired reticuloendothelial of macrophage function, this predisposes patient to systemic bacteria. Growth retardation is as a result of severe metabolic disturbances and impaired immune system. Multi disciplinary rehabilitation centers helps improve outcome and prevents the advancement of this disease to IFALD requiring liver and intestinal transplantation. [7,11].

REFERENCES

10. Greenberg GR, Wolman SL, Christofides ND, Bloom SR, Jeejeebhoy KN. Effect of total parenteral nutrition on gut hormone release in humans.


