Hypocalcaemic Heart Failure Due to Intestinal Calcium Malabsorption Following Small Bowel Resection

Hypocalcaemia is a relatively uncommon but reversible cause of cardiomyopathy and congestive heart failure. The negative inotropic effect of chronic hypocalcaemia may explain its pathophysiology. We report a case of hypocalcaemia induced reversible heart failure following small bowel resection for intestinal obstruction.

A 22 year old Hindu female presented in emergency department with marked breathlessness, orthopnoea, dull aching pain in right upper abdomen, swelling over feet, muscle cramps and generalized rigidity. She had no significant past history of any cardiovascular disease. On detailed questionnaire it was disclosed that she had small bowel resection at five years of age for intestinal obstruction. However, no records of the site and length of small bowel resected at that time were available.

On physical examination: Pulse - 108/mt. regular, blood pressure 120/80 mmHg, respiratory rate 32/mt., with JVP raised up to the angle of mandible and bilateral pitting pedal oedema. Scar mark of past intestinal resection was visible on abdomen. Liver was palpable 4 cm below right subcostal margin. Cardiovacular system examination revealed cardiomegaly with grade 3/6 systolic murmur heard all over the precordium. Fine crepitations were audible over both lung bases.

Investigations: Hb - 9.6 gm%, TLC - 7,100 cells/cum, DLC - P69, L28, M2E1, ESR - 18 mm 1st hr, blood glucose - 98 mg%, blood urea - 20 mg%, serum creatinine - 0.8 mg%, total serum proteins - 6.9 gm%, with serum albumin 4.2 gm%. Chest skiagram showed cardiomegaly (cardio-thoracic ratio .58) with left ventricular contour and pulmonary congestion. Echocardiography revealed dilated left ventricle, ejection fraction 40% and mild mitral insufficiency. On electrocardiogram QTc interval was increased (QTc = .50 sec.). Ultrasonography of abdomen revealed dilated hepatic vein and congestive hepatomegaly.

She was started intravenous and oral furosemide but her breathlessness was only partly relieved. On next morning her serum calcium was found to be only 1.91 mmol/L. Trousseau’s and Chvostek’s sign suggestive of tetany could be elicited. On the basis of this clinical picture and laboratory investigations, a provisional diagnosis of hypocalcaemia was made and 10 ml of 10% calcium gluconate intravenously every 8 hourly was started supplemented with oral calcium (500 mg tid) and vitamin D3 (0.5 mg/day). She responded dramatically with marked symptomatic and haemodynamic improvement, over next six days. At the time of discharge she was asymptomatic without diuretic and intensity of cardiac murmur was grade 1/6, ejection fraction 63%, QTc interval 0.39 sec, serum calcium 2.4 mmol/L and Hb- 10 gm%.

In this case, the temporal association of hypocalcaemia with myocardial impairment, as well as significant recovery following restoration of serum calcium level to normal by exogenous supplements, proved that hypocalcaemia was responsible for congestive heart failure. Though details were not available, it appears that duodenum was most probably resected because calcium is predominantly absorbed from duodenum. No other features of malabsorption were present because other nutrients were adequately absorbed by intact jejunum and ileum portion of small bowel. Anaemia with haemoglobin levels 9.6 gm% could not contribute to these clinical features since only severe anaemia with haemoglobin less than 6 gms% can produce cardiac dysfunction. Secondly patient showed dramatic recovery within six days while haemoglobin levels could not rise significantly within such a short period of time to explain this rapid recovery. Monig et al have reported heart failure in a patient with serum calcium 1.5 mmol/L who did not respond adequately to diuretics, digitalis and vasodilators but marked improvement in myocardial function became apparent following administration of 50 mmol of calcium intravenously for 10 days. Suzuki et al also described hypocalcaemia induced reversible heart failure following idiopathic hypoparathyroidism.

Intracytoplasmic calcium is the principal mediator of the inotropic state of the heart. In presence of ATP, linkage between actin and myosin filaments are made and broken cyclically as long sufficient calcium is present. These linkages cease when calcium falls below critical level. Normal plasma level of calcium ranges from 2.2 - 2.6 mmol/L. It is primarily absorbed from upper small bowel. Predominantly duodenum in soluble ionized from by a carrier mediated active transport under the influence of vitamin D. Hence hypocalcaemia can result from vitamin D deficient states from intestinal malabsorption as in celiac disease or intestinal bypass (to treat obesity) or after intestinal resection (to treat intestinal obstruction). Clinical presentation of this chronic hypocalcaemic state may vary widely from mild neuromuscular manifestation in form of paraesthesia, muscular twichings, carpo-pedal spasm and facial grimacing to life-threatening complications in form of convulsions, cardiac arrhythmias, hypotension, respiratory arrest and heart failure. Early recognition of clinical signs and symptoms with prompt restoration of serum calcium to normal range by rapid intravenous calcium infusion supplemented with oral cholecalciferol can revert these complications. Hence physician should consider it in differential diagnosis of heart failure in previously well young persons with no evidence of underlying cardiac disease.

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Sir,

Caroli’s disease, is a rare congenital disorder characterized by non-obstructive saccular or fusiform dilatation of intrahepatic bile ducts with two variants: the rare pure type and more common type with congenital hepatic fibrosis (CHF) known a Caroli’s syndrome. We report two cases of Caroli’s syndrome, presenting with upper GI bleeding.

Case one - A nine year female child product of consanguineous marriage who presented with two bouts of painless hematemesis and melena with history of recurrent fever, and abdominal fullness since age of 1 year but no history of jaundice, encephalopathy, blood transfusion, or passage of clay coloured stool. She had pallor; but no icterus, moderate hepatosplenomegaly, and palpable kidneys without free fluid. She was third among four siblings. First and fourth sib expired in neonatal period. Investigation - hemoglobin 8.5 gm%, TWBC - 4000/cc mm, normal liver and renal function. Viral markers were negative.

Ultrasonography showed hepatomegaly with altered echotexture, few cystically dilated biliary radicals, showed hepatomegaly with altered echotexture, few cystically dilated biliary radicals, scattered through both lobes, moderate splenomegaly with perihilar collaterals. Portal vein was 8 mm. Both kidneys showed multiple cysts. CT abdomen confirmed USG findings. Liver biopsy was not done. Upper GI scope showed grade III esophageal varices. Varices were obliterated with sclerotherapy. During 3 years of follow-up there was no recurrence of GI bleeding or attacks of cholangitis.

Case two - Twenty year old male presented at age of 6 months with lump in abdomen and fever. At 4 years he developed hematemesis, melena and was found to have esophageal varices, for which he underwent sclerotherapy. There was no history of jaundice, distension of abdomen or altered sensorium. Recently he required repeated transfusions for anemia. Ultrasonography showed cystic dilatation of biliary radicals involving both lobes of liver. Liver biopsy was suggestive of portal and septal fibrosis with preserved architecture of liver lobule. He had marked pallor with moderate hepatosplenomegaly. CT scan showed cystic dilatation of intrahepatic biliary radicals with splenomegaly. Upper GI scope showed small esophago gastric varices.

Hb 4 gm%, TLC 2000/cumm, platelet 40,000/ccmm. LFT, RFT were normal. Considering the hypersplenism patient underwent splenectomy with devascularization.

Caroli’s disease is a rare congenital disorder, of unknown etiology. Occasional familial clustering suggests possible inheritance, especially associated with polycystic kidney disease. No gene responsible for familial isolated CD is identified. Proposed mechanism is lack of normal involution of ductal plates that surround portal tracts, resulting in epithelium-lined cysts.

Clinical onset usually during childhood, pure form is characterized by segmental, saccular communicating intrahepatic bile duct ectasia, frequently accompanied by stone formation, cholangitis and hepatic abscess formation. Liver involvement may be limited or diffuse. Biliary infection and stones account for fever and abdominal pain. Death is related to septicemia and hepatic abscesses. Cholangiocarcinoma develops in 7%.

CD with CHF presents with abnormalities related to hepatic fibrosis and portal hypertension, as in our patients. Histological intrahepatic bile duct ectasia and proliferation are associated with severe periportal fibrosis. Death is related to liver failure or complications of portal hypertension.

Demonstration of communication between sacculi and bile ducts on cholangiography is important in distinguishing CD from polycystic liver disease. Black pigmented calcium bilirubinate stones are common. Associated conditions include choledochal cysts, medullary sponge kidney, infantile polycystic kidney disease and nephronophthisis.

In pure form, goal of therapy is to optimize biliary drainage. In limited disease, hepatic resection may be curative. In patients with associated CHF, control of varical bleeding is important. Late stage may require liver transplantation. Long term outcome is fairly good unless complicated by recurrent cholangitis or renal failure. Both our patients presented with recurrent variceal bleeding, controlled by sclerotherapy. One patient required splenectomy for hypersplenism. Neither had recurrent cholangitis episodes, both are stable in 4 years follow-up.

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Caroli’s Syndrome - A Rare Cause of Portal Hypertension

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Acute Lymphoblastic Leukemia with Extramedullary Relapse in Bone

Sir,

Acute lymphoblastic leukemia (ALL) is a common malignancy in childhood with high cure rates. Cure rates achieved with standard treatment for ALL in adults is only 30-40%, inspite of 70-80% of patients achieving complete remission after induction chemotherapy.

A 27-year-old male presented with fever, weakness and bleeding from gums for 15 days. Patient was pale. Hepatomegaly and sternal tenderness were present. Rest of the systemic examination was normal. Rest of the systemic examination was normal. Hemogram showed hemoglobin 9.6 g%, total leucocyte count 13000/cumm, blasts 50%, platelets 56000/cumm. Bone marrow aspiration was morphologically suggestive of ALL. Periodic acid schiff stain was block positive in occasional cells and myeloperoxidase stain was negative. Immunophenotyping report was CD19, CD20 positive and negative for CD13, CD33, CD7, CD5. X-ray chest, biochemistry and CSF were normal. Hepatomegaly was documented in ultrasound abdomen.

MCP841 protocol chemotherapy regimen was started which consists of first induction, second induction, repeat induction, consolidation and maintenance chemotherapy. He was in complete remission after induction chemotherapy (vincristine, prednisolone, L-asparaginase, daunomycin and intrathecal methotrexate). During consolidation therapy patient complained of pain, followed by multiple swellings on skull. X-ray skull showed lytic lesions (Fig. 1). Bone survey did not show additional lesion. FNAC done from the skull swelling showed leukemic infiltrates (Fig. 2). Following FNAC, bone marrow aspiration, CSF and ultrasound abdomen and testis were done. They were all reported normal. Central nervous system and testicular examination were normal. Patient refused to take further treatment.

Relapse after achieving complete remission is common in adults with ALL. Most of the ALL patients relapse in marrow. Extramedullary relapse is not uncommon. Central nervous system and testis are the most common sites for extramedullary relapse. Extramedullary relapse other than the above-mentioned sites is very uncommon. Gaynon PS et al in a series of relapsed patients of ALL observed, 38.4% relapse in bone marrow, 16.3% in central nervous system and 8.3% in testis. Other relapse sites were observed in only 1.6% of patients (mediastinum, eye, and bone). Out of these only four patients had isolated bone relapse as seen in our patient. In the same series 33% of patients with isolated testicular relapse died within 6 years of relapse, while 50% died with isolated central nervous system and 80% died with isolated bone marrow relapse after treatment. At each site longer time to relapse was associated with longer survival after relapse. Apart from four earlier cases ten more cases of isolated bone relapse has been reported in the literature.

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