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Case Report

An unusual cause of edema in a child with congenital heart disease post Fontan procedure

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ABSTRACT

Protein Losing Enteropathy Post Fontan procedure. Protein Losing Enteropathy (PLE) is an uncommon cause of edema in children with congenital heart disease. Protein-Losing Enteropathy may be defined as excessive loss of proteins across the intestinal mucosa and is due to either a primary gastrointestinal abnormality or secondary to cardiac disease. Protein-losing enteropathy (PLE) is a rare complication of the Fontan palliation for functional single-ventricle. Although PLE occurs in about 3.5% of patients post-Fontan, it confers marked morbidity and high mortality within 5 years of diagnosis. The pathogenesis of Fontan-related PLE is not completely understood, and it is unclear why it develops in some patients post-Fontan and not others. We describe a child with Double Inlet Right Ventricle who had undergone Fontan procedure, and presented to us with generalised oedema. The child had hypoproteinaemia, the common causes for which were ruled out and was diagnosed as Protein Losing Enteropathy (PLE) related to his surgical intervention. Though, not frequently encountered it should be kept in mind as one of the causes of anasarca.

Keywords: Alpha -1- antitrypsin, Congenital heart disease, Edema, Fontan procedure, Heparin, Lymphatic pressure, Protein Losing Enteropathy, Steroids

INTRODUCTION

In children operated for complex congenital heart diseases, multitudes of complications occur which affects their morbidity and mortality. The Fontan procedure was developed as a means for separating the systemic and pulmonary circulation in patients with tricuspid atresia and was then applied to other patients with a functionally single ventricle.[1]

Over next decade, the observation of complication related to loss of proteins through intestinal mucosa among these patients, gained importance. Initial reports of PLE secondary to cardiac disease, namely congestive heart failure, constrictive pericarditis and myocarditis were published in the early 1960.[2,3] The triggers of PLE are unknown, various mechanism like changes in venous pressure, infection trigger, perioperatively factors have been implicated. It is imperative to diagnose this condition before the child has severely malnourished with multiple dietary deficiencies and consider specific surgical intervention wherever possible.

CASE REPORT

Eleven year old male child case of Double inlet left ventricle (DILV) with inverted outlet chambers, severe pulmonary stenosis with ostium secundum ASD with right to left shunt diagnosed at 2 month of age, had undergone bidirectional Glenn anastomosis at 1.5 years of age and extracardiac Fontan operation done at 7 years of age was discharged on Bosentan, sildenafil and aspirin.

He was hospitalized with us with complaints of periorbital puffiness, abdominal pain, one episode of hematuria. There was history of similar such episode two month back which subsided on its own. On examination patient was afebrile heart rate 108/ min, respiratory rate 22/ min blood pressure at 50 centile, periorbital puffiness with pedal oedema and with grade 2 clubbing. Patient had right sided hydrocele. On systemic examination systolic murmur, grade 3 was present. Abdominal examination and central nervous system examination were normal. Initial impression was Urinary tract infection to rule out Nephrotic Syndrome in a child operated for Complex congenital heart disease.

Investigation done on admission showed hemoglobin of 12gm/dl, White cell count of 5900 with differential of Polymorph 83, lymphocytes 15, Eosinophil 02 platelet count of 1.45 lakh. Total protein 4.3gm and albumin of 2.0 gm/dl. with normal liver enzymes and renal function test. Urine routine microscopy showed occasional epithelial cells and occasional RBCs. Later, during ward stay, patient had developed abdominal distension with loose stools, with increase in oedema. Urine and Blood cultures were showing no growth. Ultrasonography and Doppler of abdomen and chest showed normal renal scan and liver echotexture, mild to moderate ascites with mild pleural effusion. Serum albumin dropped to 1.4 mg/dl. A 2 D- echocardiography was done to access cardiovascular status, which showed no evidence of failure of the Fontan procedure. He was started on spironolactone. Calculated and high amount of protein with micronutrients were supplemented orally.

Child required albumin infusion on three occasions. Yet the child failed to show reasonable improvement. Thus, child was diagnosed to have Post Fontan, Protein Losing Enteropathy. Patient was treated with furosemide, spironolactone, albumin infusion thrice. Oral steroids and intravenous Heparin sulphate infusion was started and after about a week patient improved symptomatically. He was referred to cardiovascular surgery opinion and was doing well on his follow up visit after one month but is lost to follow since then.

DISCUSSION

Protein losing enteropathy (PLE) is a complex disorder associated with enteral loss of proteins, entity that occurs in a variety of gastrointestinal as well as non-gastrointestinal conditions like diseases caused from bacterial or parasite infection, celiac disease, Crohn's disease, lymphoma, portal hypertension and cardiovascular corrective surgeries. It complicates many diseases; its existence is blurred due to features of the underlying condition. The disease processes that lead to protein-losing enteropathy can be classified into the following 3 major categories: a) lymphatic obstruction; b) mucosal erosion or ulceration, and c) epithelial cell dysfunction in the absence of macroscopic compromise (collagen vascular disease).

The current case had developed PLE in the background of Fontan procedure for congenital heart disease. Protein losing enteropathy (PLE) in cardiac disease is mostly associated with disorders in which chronically elevated systemic venous pressures and especially elevated superior caval venous pressures are present (constrictive pericarditis, long-standing congestive heart failure, baffle obstruction after the Mustard operation for transposition of the great arteries; after the Glenn operation and Fontan operation.[1] It may occur within months or even several years after surgery. The exact pathophysiology of PLE is unknown. It is hypothesized that chronic venous congestion with elevated superior vena cava pressures leads to disturbed lymph drainage through the thoracic duct. More-over, the increased inferior vena cava and portal vein pressures lead to increased intestinal congestion and lymph production. Proteins and lymphocytes leak from the dilated lymphatics. However, PLE has also been observed in patients with normal pressures for the Fontan circuit. Multivariate analysis of pre- and postoperative risk factors revealed that PLE is associated with non-left ventricular anatomy, long hospital stay at the time of Fontan surgery and renal failure in the immediate postoperative period.[4] The majority of reported PLE patients are from among the patients who underwent an atrio-pulmonary connection-type of Fontan procedure.[4] While PLE has been reported in a total cavo-pulmonary connection-type of Fontan, prevalence in the total cavo-pulmonary group may be lower; a 1.2% prevalence was reported in one recent study.[5] Also it comes as all or none phenomenon, those who do not develop it have no gut protein losses. In an earlier study, increased right atrial diastolic pressure is shown to be associated with PLE.[6]

The diagnosis of PLE is made on the basis of history and data for surgical correction of Complex Congenital Heart Diseases, clinical symptoms and laboratory confirmation. Alpha-1-antitrypsin is a protein normally found in the blood and in the stool. In presence of PLE, the amount of alpha - 1- antitrypsin, in the stool increases in relation to the blood sample, indicating that protein loss from the gut origin cells.

Once diagnosed with PLE, immediate cardiac catheterization is required for complete hemodynamic evaluation. The initial management includes appropriate adjustment of anticongestive therapy (digoxin, diuretics and afterload-reducing agents), administration of parenteral albumin, sodium restriction and a low fat, high-protein, medium-chain triglyceride diet. Replacement of immunoglobulin and calcium may also be considered.[7,8] In patients whose protein-losing enteropathy is related to lymphatic pathology, decreasing the lymphatic circulation provides some benefit. This requires dietary limitation of long-chain triglycerides because their absorption from the gut stimulates lymphatic flow. In order to provide adequate energy, medium-chain triglycerides must be added as an alternative source of lipid calories. As described below,

fat soluble vitamins must also be supplemented because their absorption is compromised in these patients.[9] Long-term response to this type of diet therapy was registered in 11 of the 18 patients.[10]

Obstructive lesions, if any, in the Fontan circuit must be evaluated for and, if present, relieved by means of transcatheter or surgical therapy. Similarly, aorto-pulmonary connections, naturally-occurring or prior surgical shunts, should be sought out and closed by either transcatheter methodology (coils, devices, etc.) or surgery.[11] Reduction of right atrial pressure by creating an atrial septal defect has been helpful in some patients; a number of reports show success with this method. In patients with an atrio-pulmonary-type of Fontan, conversion to total cavo-pulmonary connection may be helpful, although such converting operations are likely to have high mortality rates. Current treatments for PLE, both medical and surgical, are associated with a very high mortality rate.[12] Further investigation is needed to elucidate the exact mechanism of protein losing enteropathy and to find new therapeutic approaches.

CONCLUSION

In conclusion, Protein losing enteropathy is a life-threatening complication after the Fontan operation. The condition may be elusive initially, hence awareness about it may decrease morbidity and morbidity. Diagnosis is made by ruling out other conditions and can be confirmed by fecal α-1-antitrypsin. Comprehensive treatment plan should be inducted in the management of PLE. Patients need regular evaluation as these patients are at risk of dietary deficiencies, infections apart from hypoproteinemia.

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