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Case Report

CONGENITAL CHYLOUS ASCITES: A RARE ENTITY IN A 4M BABY

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A B S T R A C T

A 4m female baby came to our Dept. of Paediatrics, M.K.C.G. Medical College, Berhampur, Odisha, India for evaluation of abdominal distension and unilateral limb hypertrophy and diagnosed as a rare case of Congenital Chylous Ascites. Congenital Chylous Ascites is defined as accumulation of chyle into the peritoneal cavity in infants younger than 3 months. This condition is often refractory to therapy and it is responsible for severe malnutrition and immunological deficiency because of the loss of proteins and lymphocytes. Its diagnostic evaluation is difficult. In most of the cases conservative treatment is the treatment of choice. Some times repeated Paracentesis may required but when it fails, exploratory laparotomy can provide a successful alternative.

INTRODUCTION

Case Report

A 4m female baby admitted to our ward complaining of progressive abdominal distension since 1m of age and cough and cold for 15 days. She was undergoing treatment on OPD basis for abdominal distension. There was no history of fever, loose motion, constipation, vomiting, yellowish discolouration, bleeding manifestation, blood transfusion. Perinatal History was uneventful. Achieved all devlopmental milestone as per her age. Immunized as per age. BCG Scar present. Belongs to lower socioeconomic status. Her weight was 6kg, Length was 47cm and Abdominal Girth was 44cm. Bilateral upper limbs equal. Left Mid thigh was 22cm but Right mid thigh was 20cm, Left Lower leg seems to be hypertrophied.

On Examination there was no pallor, icterus, cyanosis, clubbing, lymphadenopathy But there was definite hypertrophy of left lower limb with stable vitals with B/L Rhonchi on auscultation with 1st and 2nd heart sound normally heard with out any murmur. Abdomen was distended with centrally placed everted umbilicus and visible veins without any bruit.

Investigation reports revealed CBC, CRP, CPS, RFT,LFT normal. No hypoalbunemia. Thyroid Function Test cameout to be normal. Urine albumin nil.USG abdomen and pelvis revealed moderate Ascites, B/L Kidneys texture normal, Liver Size, Texture normal. Abdominal Paracentesis done.

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Milky white colour fluid came out. Chylous test (direct visualisation of fat globules under the microscope after extraction with ether) was positive and the diagnosis was confirmed. Patient was treated with nebulization and antibiotic for pneumonia and discharged with Lasilactone (Spironolactone +Frusemide) and advised for follow up for 1month.

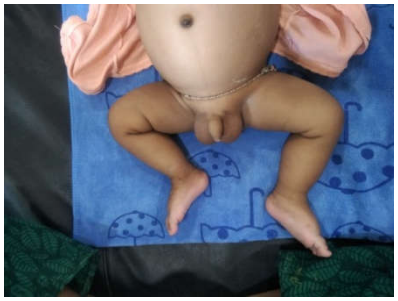

Figure 1 showing left leg swelling

Figure 2 pic of pt showing ascites

Figure 3 showing ascitic fluid after tapping

DISCUSSION

Chylous ascites (CA) is a uncommon form of ascites, defined as the leakage of lipid rich lymph into the peritoneal cavity (1).Damage or obstruction to the lymphatic system or one of its tributaries produces ascites with a turbid or milky appearance from the high triglyceride content (1).The aetiological factors of abdominal chylous ascites in the pediatric population can be primary and secondary. Abnormalities connected with the development and maturation of lymphatic vessels are the cause of primary process which is a most common cause. Chylous ascites may be the result of devlopmental defect of the lymphatic system, nonspecific bacterial, parasitic and tuberculous peritoneal infection, liver cirrhosis, malignant neoplasm, blunt abdominal trauma and surgical injury (3,4). In adults the most common cause is abdominal malignancy, while in children, congenital lymphatic abnormalities are more common. The presence of these anomalies in genetic syndromes- Turner syndrome, Yellow nail syndrome, Klippel-Trenaunay-Weber Syndromes points role of genetic factors in etiopathogenesis of lymph circulation(7). Affected neonates usually presents with abdominal distension and respiratory distress (2).Abnormal function of the GI tract results in malabsorption of nutrients in the intestines.

Ultrasonogram of the abdomen confirms the presence of ascites. The most useful diagnostic method is Paracentesis (5). Although lymphangiography is the gold standard in defining the cause of the lymphatic obstruction, it is an invasive procedure. Lymphoscintigraphy is a method showing the site of lymph leakage, connected with a lower risk of complications, and offering a possibility of repeating the evaluation(6).Invasive techniques-Laparoscopy and Laporotomy, still remain a diagnostic and therapeutic tool used in abdominal chylous as cites resistant to conservative treatment. Visualisation of the site of lymph leakage by providing the patient with pre- and intraoperative administration of fat soluble dyes seems to be the highest importance for the succsess of these methods (6,7).In Lymphangiogram almost always the lymphatic vessels are dilated. But one point to note here is that grossly dilated and tortuous vessels points towards absence of valves having poor prognosis.

After confirmation of the diagnosis, patients should be treated conservatively by dietary correction of any fluid, electrolytes and vitamins D deficiencies, and provided with fat-free diet, with the fat being replaced by medium-chain triglycerides (8). In severe or complicated chylous ascites that persists after a maximum of 6-10weeks of complete bowel rest and total parenteral nutrition should be initiated. Abdominal Paracentesis can rapidly relieve respiratory insufficiency and abdominal discomfort. Parenteral nutritional support appears to decrease the hazard of medical therapy by maintaining an adequate nutritional intake while eliminating obligate losses consequent to repeated paracentesis (9). Somatostatin arrests lymphatic flow through reduction in gastric, intestional, and pancreatic secreations or by a decrease in hepatic venous pressure and splanchnic blood flow. Surgery is advised in cases resistant to conservative treatment, which is usually undertaken for 4-8weeks.Before surgical intervention, CT, MRI and laparoscopic methods are applied to define the type of lymphatic vesssel pathology and its location (6).A shunt joining the peritoneal cavity with the venous system is treated as last resort, due to the possibility of severe complications, and a limited effectiveness (6,7).

CONCLUSION

Congenital chylous ascites is a rare disease in newborn. It is especially important to intensify and individualise the treatment process. Conservative treatment, with an early adminstration of somatostatin seems to be advantageous, as it shortens the time of treatment and hospitalisation. Surgery should be considered after ineffective attempts at conservative treatment continued for 4-8 weeks. Before surgery, it seems necessary to try to establish the type and location of the pathology of lymphatic vessels, with the use of noninvasive and invasive diagnostic methods. Still many babies particularly having valves in lymphatic channel will have normal life.Our case after the diagnosis advised to come for follow up.We have decided to publish this case as it is a rare entity with amikable outcome.

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