CONGENITAL INTESTINAL LYMPHANGACTASIA

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Mrs. H. A. 32 years old lady admitted with the complaints of huge abdominal swelling and abdominal pain for 5 month. The pain was continuous & dull aching in nature which was localized on her right side of abdomen. There was no history of weight loss, jaundice, oliguria, dysuria, cough, breathlessness, palpitation, joint pain, vomiting, constipation or diarrhoea .She didn't visit to any filaria prone zone since her childhood. On query we were informed that the similar type of symptoms first developed when she was of only 9 months age. Since then, she had recurrent abdominal swelling which responded occasionally with frusemide. In 1998 and 2012, she again developed similar type of symptoms with severity and admitted in a tertiary care hospital and paracentesis of 3.5 Litres of milky fluid(chyleascites) (Fig.-1) was done. She was prescribed with tablet Frusemide which she consumed for 1 year. On abdominal examination there was huge ascites evaluated by fluid thrill. There was no lymphadenopathy. Liver was enlarged in size ,about 2 cm from rt. Costal margin along the rt. Mid clavicular line. It was mildly tender with a smooth surface,sharp margin,no associated bruit. Respiratory, Cardiovascular, Neurological & all other system examination revealed normal findings.

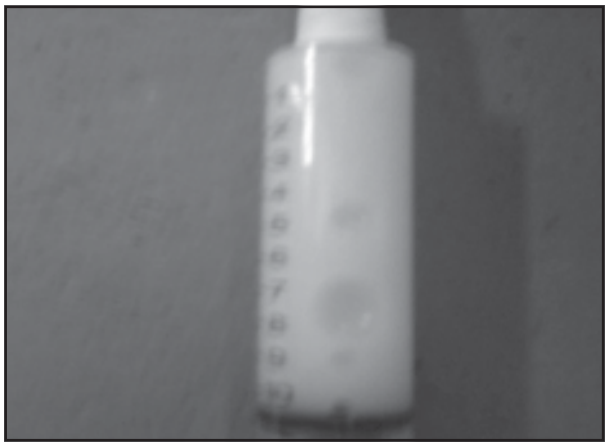

Fig.-1: *Chyleascites at paracentesis shown milky fluid*

Lymphoscintigraphy of both lower limb with 99m-Tc-Dextran Nanocolloids was done which suggest patent lymphatic drainage in both lower limbs with obstruction in abdominal drainage in both side (Fig 2). CT scan of abdomen was unremarkable and Liver

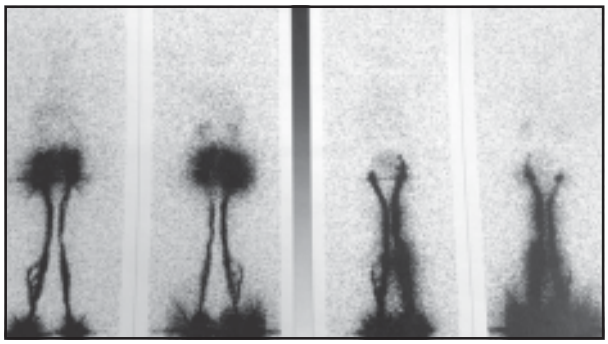

Fig.-2: *Lymphocentigram shows normal lymphatics in both lower limbs but complete absence of tracing at abdomen suggesting absence of lymphatic channels.*

biopsy and Fibroscan of liver revealed incidental mild chronic hepatitis which can not explain the chyleascites since 9 months of age. The patient was diagnosed as **Congenital Intestinal Lymphangactasias.** Lymphoscintigraphy is safe, easily performed and relatively noninvasive. By contrast, lymphangiography is time-consuming, invasive and not without risk of lymphangitis, pulmonary embolus and, under some circumstances, cerebral embolization and anaphylactoid reactions to the iodine-containing contrast medium[1,2]. This diagnostic test, however, does have a place in the preoperative evaluation of patients undergoing surgery for primary chylous disorders and, when expertly performed,complements the data provided by lymphoscintigraphy.[3,4]

References:

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