Effect of Plasmapheresis on Treatment of Acute Pancreatitis in Infant with Familial Chylomicronemia

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Abstract

Familial chylomicronemia is a rare genetical disorder, (autosomal recessive) with incidence 1/1000000 that there is difficulty in clearance serum content of triglyceride & cholesterol due to deficiency of Apo-B-lipoprotein lipase or lipoprotein lipase (LPL) or cofactor Apo-C II. Different between them is decrease level of serum TG after prescription FFP in Apo-CII and without response in LPL deficiency.

Clinical features are: Hepatosplenomegalia, Pancreatitis attack, Xanthem eruptions on limbs & butects, Paleness of retina (libemia reticolaris).

We introduce a female baby 41 days old that referred by hyperlipidemia (TG=25000mg/dl & cholesterol=1500mg/dl) and acute pancreatitis attack and signs & symptoms denoting to familial chylomicronemia who treated by plasmapheresis.

Keywords: Plasmapherisis; Acute Pancreatitis; Familial Chylomicronemia

Introduction

Familial chylomicronemia syndrome is a rare genetical disorder with Incidence 1/1000000 that there is difficulty in clear content of Triglycerides and cholesterol due to deficiency in (Apo-c2) [1,2,5].

The different of deficiency is in from relative increase of serum's Triglycerid inform chylomicronemia in deficiency (cofactor Apo-c2) and clear increase in deficiency LPL more than 10000mg/dlit. From the viewpoint of clinical the patients have signs like Xezanthom on knee and arm and hepatosplenomegalia and finally clear attacks pancreatitis in infancy and childhood. Also their eyes fondoscopy is due to lipmia retinalis [3-5].

We introduce a female infant 41 days age, effected with familial chylomicronemia that has referred With Triglicerid level 25000mg/dlit and cholesterol 1500mg/dlit and pancreatitis acute attack and has treated by plasmapheresis.

Case Presentation

Full name: Newshe Akbari Father’s name: Hassan Age:34yrs. Education: Diploma Occupation: Farmer BG: o+ Mother’s name: Zahra, Age:25 yrs. BG: o+

Date of admission:07.05.2009 CC: so black stool, agitation, fever

PI: The Infency 41days,brought to Emam Reza hospital with complain of fever,azhite,cry,black stool since last week. She's
had diarrhea since 4 days ago that stopped after referring to physician and taking cephalaxin. She was agitated since 3 days ago and she’s had black stool-poor feeding – he- matemesis-vomiting.

Table 1. Laboratory findings.

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In sonography: Liver span 13 cm

PMH: She’s the second child from 28-year-old mother G2P2AB, product of NVD, term, birth weight: 3100gr

In inspection of birth, tested for ichter that been showed. She’s TG:1800mg/dlit & Total.Bill:13mg/dlit the second day admitted in Hazrat masume hospital. She has been treated with UTI diagnosed for 12 days in this hospital.

DH: Amp. Cephalaxin/IV ,Amp. Vit k /Im,Drop.Acetamino phen

PH: her parents are related(cousin)

GA: the infancy 41 days, alert , pale, but she isn’t ill and toxic.

BP: 80mmHg/puls. PR:160/min.RR:42/min T:37 grading(After taking Acetaminophen)

Head & neck: normocephal, anterior fontanel: 28*25mm

Posterior fontanel tip finger. The fontanel isn’t bulge. Cnjunctives were pale. Scholraes weren’t Ichteric.-mucous were little dry. Neonate reflexes were normal. Breath sound: normal. heart acchie cardy. Abdomen: was distened. Her liver was length 3-4 cm from right 7th rib. The spleen touched under the edge of left 7th rib. Her genitalia: normal. Her skin had petechi & purpura-and her limbs were edematose. She had not lymphadenopathy.

Abdomenal X Ray: evidences not seen provied dilata- tion of intestinal loops & obstruction. Fecal with fizz seen into rectom. Fizz pattern suddenly has taper in connection desental colon to sigmoid. For investigation request ed nonemergency barium enema at Emam Reza hospital.

In four times with heparin infusion 50µg/Kg/24h). Her TG:22800mg/dlit&Cholestrol:1970mg/dlit were before plasmapherisis.

She discharged in 20.05.2009 with TG:400mg/dlit,Chol:23 mg/dlit and medicinal orders contain:Syrup Ranitidin (15mg/Bid/Po), Cap simvastatin 100mg/Bid/Po and Cap Gimfibrozil 50mg?daily Po also recommended to come back 3 days later for continuing of investigation.

Discussion

Increasing in fasting TG level more than 1000 mg/dlit in children is a reflexation of intense hyperchylomicronemia and refers to background disorder [1-3,5].

In some cases the genitical factor and other medical conditions or hormonical or environmental factors. Main clinical signs in intensive hypertriglyceridemia is acute pancreatitis and the primary treatment for reducing this serious compli cation is reduction TG level less than 1000mg/dlit [4,5].

Some of patients that affected with intensive hypertriglyceridemia are at high risk for CHD and premature atheroscle rotic and they need to more aggressive treatment even in the condition their TG level is less than threshold 1000mg/dlit [3-5].

Familial chylomicronemia syndrome, lipase lipoprotein deficiency, and APO lipoprotein c-2 deficiency. Characters of familial chylomicronemia syndrome is acute pancreatitis in background of is creasing TG more than 1000mg/dlit in childhood [1-5].

1-The current sign in medical history is intermittent ab-
dominal pain. Seldom the Infant refer with colic from clinical point increasing Plasma's TG level is silent and sometimes discover random by apparent of fatblood [4,5].

2-In physical exams seen abrptive Xezanthom(small papular loss that appears a lot on bottom and back of trunk [5].

3-lipmia retinals (Pale apparent of retina's veins) is the trail of intensive hyper Triglycerideia. Often seen hepatospleeno-megali due to digestion of chylomicrons by reticuloendothelial system [4,5].

4-Generally cardiovascular atherosclerotic isn't a sign of this disease. Two different genetical deficiency causes familial chylomicronemia syndrome: LPL and apo-2 deficiency [3-5].

5-Hydrolise of Triglicerides in chylomicrons needs to activ-

ity of LPL in background tissue liver's capillary and apco-2 acts as cofactor of LPL activity [2,3,5].

6-Mutation in the deletion gen LPL or apco-2 causes nonhy-
drolise TG in chylomicrons and doesn't cause hyper chylomi-
cronemia. The disorder come down by inheritance in form Autosomal and both of alleles of LPL or apco-2 gens should be involvement. So TG level in parents of patients generally are normal or close to normal and usually not seen the fam-
ily history of intensive hyper lipidemia [5].

7-Both of these disorders are rare but deficiency of LPL is current than apco-2 (approximately 1 in 1 million people). Hyper chylomicronemia distinguish by clinical signs and some important laboratoriual founds.

One of them is plasma lasent and after putting it in the refrig-
erator for one night form a cake of chylomicrons on the sur-
face of it. TG level is more than 10000mg/dl and even more, also full cholesterol level goes up due to existence of chole-
sterol in the chylomicrone [3-5].

Lipoprotein electrophoresis shows obvious increasing of chylomicrons but it's not necessary for diagnosis. The diagnosis of deficiency LPL and apco-2 verify in specialty centers with quantitative measurement of LPL activity in plasma after intra venus injection of heparin (Lipolitic activity post heparin) [5].

8-In order of diagnosis the doubtful patients to chylomicro-
nemia syndrome should refer to lipid specialty centers [2,3].

9- Treatment in familial chylomicronemia syndrome is full limitation of fat in diet [1-3].

10-Consultation with a nutrition specialist familial to this disorder is necessary. Make use of nutritious complemen-
tary with middle chain Triglycerids(MCT)will be useful that directly absorb into portvein and does'n't cause chylomicronemia.

Some of patients response to oil fish if fat limitation alone isn't successful [2-5].

11-In patients of affected with apco-2 deficiency for treatment of acute pancreatitis for elimination intensive hyper triglyceridemia and pancreatitis can use FFP transfusion that be exogenic source for apco-2 [4,5].

12- Used of plasmapheresis is a common method in internal medicine for poison remover or rapid reduction some of toxic and sometimes it's useful in rapid reduction of serum's lipid level but not reported yet make use of this method control of acute pancreatitis attack in familial chylomicronemia in very high level of triglyceride. In this case existence of hyper triglyceridemia (25000mg/dlt), hyper cholestrolemi(1500mg/dlt) lactic plasma, hepatospleenome-galia and limpa retinalis, venus heparin advise test and FFP transfusion in sometimes and non reduction serum lipids level, cause that for the patient distinguished familial chylomicronemia due to LPL deficiency (lipoprotein lipase)and because of fever and azhitation (colic attacks), Increasing of anylase and lipase level and sonography and abdominal CT and scan that shows pancreas inflammation, moots likeli-
hoods acute pancreatitis after hyperlipidemi.

So we kept the patient NPO and inserted NGT tube, gave injection anti acid (H2BLOKER),and injection analgesic(pethedin) and injection suitable antibiotic with 4 times plasmapheresis infant's plasma, lipids reduced rapidly to less that 400mg/dlt(TG) and 232mg/dlt(cholesterol) finally seen clinical and laboratory obvious improvement acute pancreatitis.

References


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