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Case Report

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/ 1000,000 that there is difficulty in clearance serum content of triglyceride & cholesterol due to deficiency of Apo-B-lipoprotein lipase or lipoproteine 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, Xantem 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 plasmapherisis.

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 hepatospleenomegalia 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: Newsha 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,brouth 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 cephalexin. She was agitated since 3days ago and she's had black stool-poor feeding – hematemesis-vomiting-

Table1. Laboratory findings.

	Na	K	BS	BC1000*	HCT	PH*1000	ESR	PT	INR	PTA	Tg	chol	HDL	AST	ALT	♁ALkP	Amilase	Lipase
07.05.2009			48				34	14.1	1.2	84.1	22890	1970	265	30	40	980		
//08/05.2009				21.9	16.1	561											100	80
09.05.2009					21.9													
70.05.2009				15.1	22o2	110												
/09.05.2009	136	3.9	75	13.8	22.6	165					13110	1120						
10./05.2009			65								6360	680		38	40	510		
11.05.2009			80								1730	360						
12.05.2009	135	4.7	88								400	232		24	15	460		

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 for12 days in this hospital.

DH: Amp. Cephalexin/IV ,Amp. Vit k /Im,Drop.Acetaminophen

PH: her parents are related(cousin)

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

BP: 8ommHg/puls. PR:160/min.RR:42/min T:37C(after taking Acetaminophen)

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

Posterior fontanel tip finger. The fontanel isn't bulge. Cnjunctivaes were pale. Scholraes weren't Ichteric.-mucous were little dry. Neonate reflexes were normal. Breath sound: normal. heart achie 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 dilatation of intestinal loops & obstruction. Fecal with fizz seen into rectom. Fizz pattern suddenly has taper in connection desental colon to sigmoid. For investigation requested nonemergency barium enema at Emam Reza hospital. Gastric washing done that wasn't bloody. Treated with ceftazidim and keflin and Vit K/ IM. transfused P.C 2 times in 07.05.2009 and 09.05.2009 also transfused FFP. Inserted cat down in 09.05.2009. Plasmapherisis done 4 times in dates 09 .05.2009/10.052009/11.05.2009/12.05.2009(exchange50ml/Kg of whole of her plasma fluid by serum albumin 5%.

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

She discharged in 20.05.2009withTG:400mgdlit,chol:232mg/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 complication 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 atherosclerotic 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 abraptive 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 hepatospleenomegali due to digestion of chylomicrons by reticoloendotelial 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 activity 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 apc-2 causes nonhydrolise TG in chylomicrons and doesn't cause hyper chylomicronemia. 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 family history of intensive hyper lipidemia [5].

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

One of them is plasma lasent and after putting it in the refrigerator for one night form a cake of chylomicrons on the surface of it.TG level is more than 10000mg/dl and even more, also full cholesterol level goes up due to existence of cholesterol 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 apoc-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 chylomicronemia 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 complementary with middle chain Triglycerids(MCT)will be useful that directly absorb into portvein and does'nt cause chylomicronemia.

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

11-In patients of affected with apoc-2 deficiency for treatment of acute pancreatitis for elimination intensive hyper triglyceridemia and pancreatitis can use FFP transfusion that be exogenic source for apoc-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/dlit), hyper cholestrolemi(1500mg/dl)lactic plasma, hepatospleenomegalia 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 amylase and lipase level and sonography and abdominal CT and scan that shows pancreas inflammation, moots likelihoods 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/dlit(TG) and 232mg/dlit(cholesterol) finally seen clinical and laboratory obvious improvement acute pancreatitis.

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