Eastern Journal of Medicine 20 (2015) 235-237

Case Report

A rare cause of metabolic acidosis; Hypoplastic left heart syndrome

Nihat Demir[a], İbrahim Ece[b], Kaan Demirören[c], Nesrin Ceylan[c,*], Erdal Peker[a], Oğuz Tuncer[a]

[a]*Department of Pediatrics, Division of Neonatology, Yuzuncu Yil University School of Medicine, Van, Turkey*
[b]*Department of Pediatrics, Division of Cardiology, Yuzuncu Yil University School of Medicine, Van, Turkey*
[c]*Department of Pediatrics, Yuzuncu Yil University School of Medicine, Van, Turkey*

Abstract. Metabolic acidosis is a common problem especially in newborns. Metabolic acidosis develops due to acid deposition rather than HCO_3 loss in body fluids. It separates into two groups as anion gap normal and anion gap increased depending on anion gap level. Metabolic diseases, neonatal sepsis, congenital adrenal hyperplasia and congenital heart diseases should be thought with the observation of metabolic acidosis in a previously healthy infants. $NaHCO_3$ infusion, renal replacement treatment and tamps like karbicarb, dichlora acetate, tromethamine are used for treatment of congenital heart diseases or lactic acidosis secondary to hypoxia. In this article we want to emphasize there are not only congenital metabolic illnesses under this non-response acidosis table but also there are congenital heart diseases which are in substantial rate.

Key words: Metabolic acidosis, hypoplastic left heart syndrome, neonate

1. Introduction

Metabolic acidosis is a common problem especially in newborns. Metabolic acidosis develops due to acid deposition rather than HCO_3 loss in body fluids. It separates into two groups as anion gap normal and anion gap increased depending on anion gap level. The gastrointestinal and renal reasons which cause the loss of normal anion gap metabolic acidosis develop due to aldosterone lackness, overtaking of intravenous chlor and over extracellular volume expansion. The reasons of metabolic acidosis with increased anion gap are asphyxia, hypothermia, severe respiratory distress, sepsis, hipotermi, and secondarily developed lactic acidosis due to the serious diseases of newborn. The lower frequency rate of reasons are renal failure, inherited metabolic diseases and overtaking toxine. Observing metabolic acidosis in the healthy newborns in the first three days of life cause to think the possibility of metabolic diseases, neonatal sepsis, congenital adrenal hyperplasia and congenital heart diseases (1, 2).

[*]Corresponding Author: Nesrin Ceylan, M.D.
Department of Pediatrics, Yuzuncu Yil University School of Medicine, 65250, Van, Turkey
Tel: +905324318522
Fax: +904322155281
E-mail: drnesrinceylan@hotmail.com
Received: 28.12.2013
Accepted: 21.02.2014

Hypoplastic left heart syndrome is characterized in anomalies that are close to each other such as atresia and/or critical stenosis in aorta or mitral valve, left ventricular hypoplasia, ascending aorta and aortic arc hypoplasia. Since ductus is a dependent cardiac defect, it is asymptomatic until closure of ductus happens. If it is not interfered in few days after birth, hypoxia, acidosis and shock will happen following closure of the ductus arteriosus. This multifactorial reasoned cardiac defect forms 0.016-0.036% of all live births, 1-3.8% of all congenital heart defects and 9% of critical newborn patients (3,4). With this fact we want to emphasize there are not only congenital metabolic illnesses under this metabolic acidosis unresponsive to treatment table but also there are congenital heart diseases which are in substantial rate.

2. Case report

A 3020 gr (male) baby who was born as normal spontaneous vaginally with 1. and 5. Apgar score respectively 8-9 from the third live birth of 30 years of mother, was learned that his general health condition was fine, actively breastfed and discharged postnatal 12th hour. The baby whose breastfeeding decreased on postnatal second day and stopped totally on the third day and breathing with moan started, was taken to our emergency room. In his first physical examination the patient was in septical appearance, upper extremity tension arterial was 32/22 (systolic/diastolic) mmHg. The capillary (filling) time of the patient lengthened out (> 7 sec) whose lower extremite

tension and pulse could not be taken as well, 4 cm palpable was obtained from liver edge. In his cardiac examination peak heart rate as 181 beat/min, second heart beat was single and hard, 1/6 systolic cardiac murmur from sternum left upper edge, respiration rate as 72/min and intercoastal-subcoastal typical resignation were obtained. Since the patient was a newborn, he was hospitalized to intensive care unit. Then, since the patient was in decompensate shock status, he was given respiratory support and deficits support as 20 cc/kg to 0.9% NaCl with two times. On the other hand the urine was obtained as 89 mg/dL (normal range [NR]:10-50**),** creatinine 2.5 mg/dl (NR:0.5-1.2), Na 147 mmol/L, K 6 mmol/L, Ca 9.9 mg/dL, AST 165 U/L, ALT 97 U/L, ketone negative, ammoniac 71 (16-60) µmol/L, Hb 13.7 gr/dL, Hct 41%, white blood 18500/mm^3, thrombocyte 151000/mm^3, PT 37 sec, aPTT 28 sec, CRP 4 mg/dL. Blood gas values PH 6.96, PO_2:42 mmHg, PCO_2:44 mmHg, HCO_3:9 mmol/L, BE -23, when lactate was obtained as 20 mmol/L, sodium bicarbonate ($NaHCO_3$) deficit was given. The results of blood and urine culture were obtained normal. The continuity of acidosis despite three times of $NaHCO_3$ deficit treatment, the value of bicarbonate did not do over 8 mmol/L and there were symptoms of acute renal failure, peritoneum dialysis was opened. In echocardiography by obtaining hypoplastic left heart, aortic valve atresia, hypoplastic ascendens aorta, ductus dependent systemic circulation and large atrial septal defect**,** Hypoplastic Left Heart Syndrome was diagnosed by pediatric cardiologist (Figure 1A, 1B). Then in order to keep ductus open, prostaglandin E_1 infusion was started. Cardiac computerize tomography (CT) angiography was done to confirm diagnosis. In CT angiography left ventricul exit, until ascending aorta with innominant artery (innominate artery) level, arcus aorta was not observed (Figure 1C). Since it was obtained as in the 12th session of periton dialysis (peritoneal dialysis) PH 7.44, PO_2:71 mmHg, PCO_2:41 mmHg, HCO_3:25 mmol/L, BE 3, in the 50th Session creatin 1 mg/dL, urea 30 mg/dL, Na 134 mmol/L, K 3.8 mmol/L, dialysis was stopped. As the respiratory insufficiency and laboratory parameters were obtained well, the patient was extubated and put under nasal CPAP observation. Since his general condition partially got better, the patient was transferred to surgery to put a stent on ductus. However the patient was lost during surgery.

3. Discussion

It is very important to have an urgent diagnose since the metabolic acidosis occurs with shock in newborn facts. But since the clinical table is wide, its diagnose is also difficult. In this kind complex table the first thing is to support respiratory of patient, provide circulation and amend metabolic problem (5). Because of the general condition of our fact is pretty bad, he was provided mechanical ventilation. Since the patient is in decompensate shock table, he was loaded 20 cc/kg 0.9% NaCl two times. In baby facts since the most frequent reason of metabolic

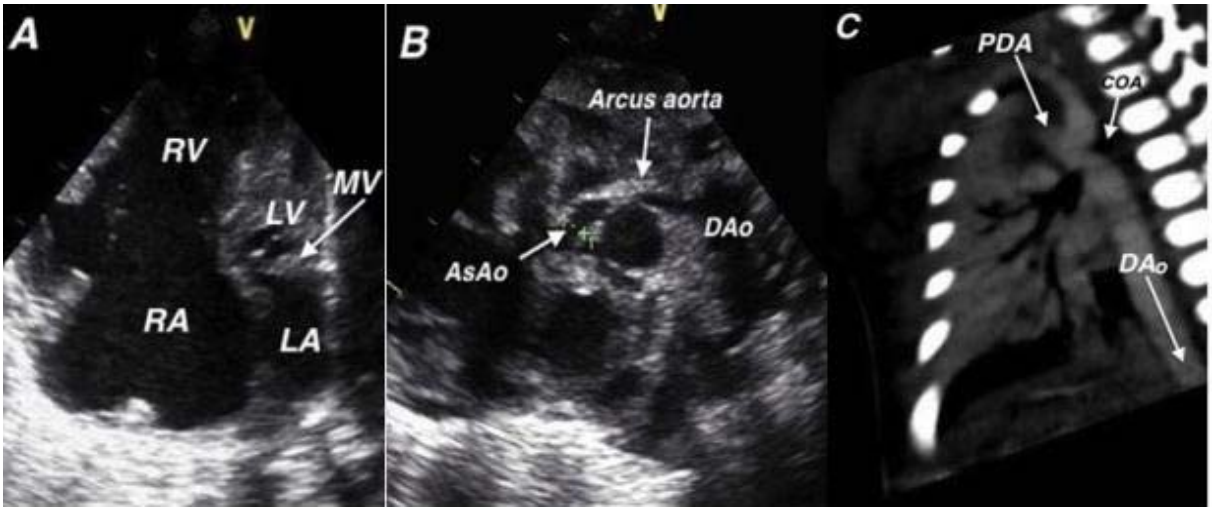

Fig. 1. 1A, 1B; Echocardiogram showing hypoplastic left heart, aortic valve atresia, hypoplastic ascendens aorta, ductus dependent systemic circulation and large atrial septal defect**,** 1C; Cardiac computerize tomography angiography not observing left ventricul exit, ascending aorta and arcus aorta.

RV=Right ventricle. LV=Left ventricle. RA= Right Atrium. LA=Left atrium. MV=mitral valve. AsAo=Ascending aorta. DAo= Descending aorta. CoA= Coarctation of aorta. PDA= Patent ductus arteriosus.

acidosis with high anion gap is lactic acidosis secondary to tissue hypoxia, serum lactate level must be measured. Also renal insufficiency, congenital metabolic diseases and toxins which can cause anion gap increase, must be considered (2). In our fact since serum lactate was obtained as 20 mmol/L, the first reasons were thought to be tissue hypoxia and congenital lactic acidosis. The general condition malfunctions that depend on heart diseases show themselves as congestive heart insufficiency, circulation insufficiency and malfunction of oxigenation. In ductus depending congenital heart diseases, when ductus starts to close, congestive heart insufficiency, metabolic acidosis and shock starts to develop rapidly. Generally these patients are diagnosed as congenital metabolic disease, sepsis and renal insufficiency by mistake (6). As a matter of fact the laboratory and clinical condition in our fact caused to think us at first metabolic acidosis depending on severe dehydration, secondarily developed renal insufficiency. For lactic acidosis treatment $NaHCO_3$ infusion, renal replacement treatment (periton dialysis, hemofiltration, venose-venose hemofiltration) and other tamps (Carbicarb, Dichloroasetate, Trometamine) are used (7). Since our patient has non-response lactic acidosis and acute renal insufficiency, peritone dialysis was started. In extremities in addition to weak pulse, hypotension is also evaluated as shock. When clinical and laboratory results of our fact were evaluated, the condition was obtained as cardiogenic shock developed secondarily to hypoplastic left heart sydrome.

Finally septum lactate level of every newborn must be evaluated in who severe metabolic acidosis is obtained. There tandem mass examination must be done as well as echocardiography in facts having lactic acidosis. We present this fact in order to remind that congenital heart diseases are among the reasons of non-response metabolic acidosis and metabolic acidosis which is resistant to treatment can be treated in short term by periton dialysis.

References

1. Ward KE, Pryor RW, Matson JR, et al. Delayed detection of coarctation in infancy: implications for timing of newborn follow-up. Pediatrics 1990; 86: 972-976.
2. Michael AP, Jacquelyn RE. Acid-Base, Fluid, and Electrolyte Management. In: Christine AG, Sherin UD, editors. Avery's Dıseases of The Newborn. 9th ed. Philedelphia: Mosby Elsevier; 2012: 367-390.
3. Connor JA, Thiagarajan R. Hypoplastic left heart syndrome. Orphanet J Rare Dis 2007; 2: 23.
4. Barron DJ, Kilby MD, Davies B, et al. Hypoplastic left heart syndrome. Lancet 2009; 374: 551-564.
5. AB Z. Metabolic Acidosis. In: Fanaroff A, Martin R, Walsh M, editors. Fanaroff and Martin's Neonatal-Perinatal Medicine: Diseases of the Fetus and Infant.Vol 4. 9th ed. Philedelphia: Mosby Elsevier; 2010: 1659-1660.
6. Park M. Coarctation of the aorta. In: Park M, editor. Pediatric cardiology for practitioners. Philedelphia: Mosby Elsevier; 2008: 205-213.
7. Kanburoğlu MK, Çizmeci MN, Türkay S, Örün UA, Tatlı MM. Yenidoğanlarda Ağır Metabolik Asidozun Nadir Bir Nedeni: Aort Koarktasyonu. Yeni Tıp Dergisi 2012; 29: 183-186.
8. Fall PJ, Szerlip HM. Lactic acidosis: from sour milk to septic shock. J Intensive Care Med 2005; 20: 255-271.