Case Report

Congenital primary hypothyroidism associated with the rare form of nonimmune hydrops fetalis

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Abstract. We present a male newborn born with diffuse edema and ascites. A diagnosis of congenital primary hypothyroidism was made based on thyroid hormone levels of total T_4 1.74 µg/dL, free T_4 0.30 ng/dL and TSH >75 µIU/mL and thyroid hormone replacement therapy was initiated. At day 15 of therapy, the thyroid function tests of the patient reached normal limits, and his edema and ascites regressed. In this report we present a newborn case of hypothyroidism that was accompanied by nonhydrops fetalis. We want to emphasize that congenital hypothyroidism may present with severe symptoms such as hydrops.

Key words: Hypothyroidis, hydrops fetalis, newborn

1. Introduction

Nonimmune hydrops fetalis (NIHF) characterized by generalized soft tissue edema and cavity effusion is a life-threatening syndrome in the fetus and newborn (1). It is seen in approximately 1 / 3000 live births and has high mortality rate with about half of cases presenting with in-utero death (2). The etiology of NIHF has many different. these may result of fetal anomalies, placenta or the mother the disorder (2-4). In this report, a newborn is presented who diagnosed as NIHF with congenital primary hypothyroidism at birth.

2. Case report

The patient was the fourth child of consangious parents. The pregnancy had been uneventful until 32 weeks. The baby had no complication at birth and during intrauterine life. Apgar score was ten at five minutes but there was a placental edema.

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He presented with generalized edema and respiratory distress on first day of his life, not necessitating cardiopulmonary resuscitation and mechanical ventilation. After stabilization of his condition, he was transferred to the neonatal intensive care unit.

His weight was 2230 grams, height 43 cm, and head circumference was 30 cm. There was severe generalized edema, abdominal distention and hepatomegaly (4 cm palpable) on physical examination. No malformations or dysmorphic features were noticed (Fig. 1).

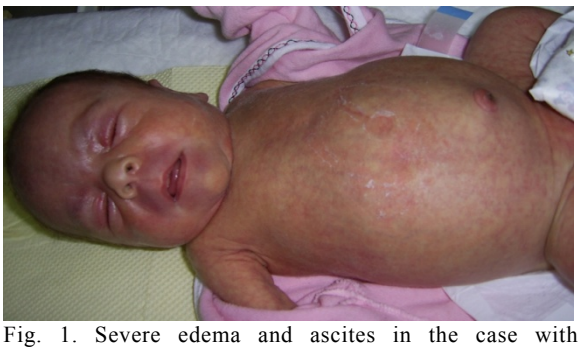

Fig. 1. Severe edema and ascites in the case with congenital hypothyroidsm.

Serum electrolytes, creatinine, blood urea nitrogen, and liver enzymes were in the normal range (aspartate aminotransaminase 63.45 U/L and alanine aminotransaminase 20.9 U/L). The serum protein level was low (3.7 g/dL), as well as the serum albumin level (2.5 g/dL), hemoglobin

20.5 g/dL; platelet count 251000/mm^3; white blood cells 16700/mm^3, lymphocyte 40%, neutrophils 70%, with normal red cell morphology. There was no evidence of hemolysis or hyperbilirubinemia; therefore levels of fetal hemoglobin and glucose 6–phosphate dehydrogenase were not obtained. Additional laboratory tests were performed in an attempt to detect a possible cause of hydrops fetalis (HF). An immune-based process was unlikely, because the blood groups of the mother and infant was A-positive; results of an indirect coombs test in the mother and a direct coombs test in the infant were negative. There was no metabolic acidosis; IgM antibodies for parvovirus B19, toxoplasmosis, rubella, cytomegalovirus, herpes simplex virus, Epstein–Barr virus, and adenovirus were negative. Urinary amino acids, organic acids, and mucopolysaccharides were normal. ventricular septal defect and persistent foramen ovale, no pericardial effusion. Central venous pressure was not measured. Results of ultrasound examination of the brain were normal.

Ascites was demonstrated on abdominal ultrasound scan. At day 16 of age, serum free thyroxine (FT4) level was 0.30 ng/dL (normal values, 0.6 to 2 ng/dL) and thyroid-stimulating hormone (TSH) level was 75 µIU/mL (normal values, <10 µ IU/mL). Low FT4 and high TSH levels confirmed the diagnosis of congenital hypothyroidism. Auto antibodies of thyroid were not detected in the mother or in the infant. Thyroid ultrasound examination revealed hypoplasia of the thyroid gland. Thyroid hormone supplementation was started, and within 4 days the infant became more vigorous and after 10 days, skin edema and ascites disappeared, and serum thyroxine level normalized (FT4 1.7 ng/dL). The infant started to gain weight and was discharged from the hospital at 25 days of life (Fig. 2).

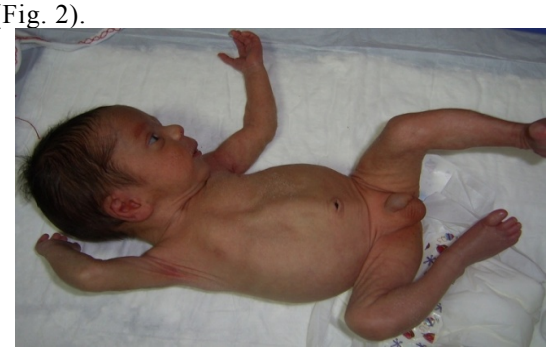

Fig. 2. The view of the case after thyroxin therapy.

His mother's urine iodine concentration was low (20, 3 µg/L) but her thyroid functions were normal levels. The iodine concentration is normal in our patient's urine.

3. Discussion

Hydrops fetalis may be related to maternal, fetal, placental and umbilical cord causes (1-4). The physiopathology of NIHF is releated abnormal fluid transposition between the plasma and tissues (5,6). The medical history of our patient revealed that his mother had no complaints during the prenatal period and her thyroid function tests and antibodies were in the normal ranges in the postnatal period though the placenta was swollen. He was born at 32 weeks by spontaneous vaginal delivery perinatal asphyxia and meconium aspiration were absent during birth. Laboratory studies showed no signs of blood conflict between the mother and infant and no metabolic acidosis was present. Mother's indirect coombs test and infant's direct coombs test were negative and hepatitis, evidence of TORCH group and parvovirus B19 infections were absent.

In the literature, cardiac tamponade and pleural effusion have been described in patients with hypothyroidism (7-10). Also pericardial effusion with or without cardiac tamponade symptoms is reported in children with hypothyroidism in whom the diagnosis and treatment is delayed (11-14). However it is not a frequently encountered condition that an infant with congenital hypothyroidism has clinical finding at birth. Our patient was born with hydrops fetalis, abdominal ascites and dermal diffuse edema; however pleural or pericardial effusion were absent. With thyroxine replacement therapy edema and ascites improved rapidly.

In infants born with NIHF, it is necessary to consider congenital hypothyroidism even if the condition occurs rarely, and we want to emphasize that severe symtom such as hydrops could occur during the neonatal period. Perhaps, the patient's findings occurred in the newborn period due to iodine deficiency, because his mother had lived in an endemic iodine deficiency area and her iodine level was low in her urine.

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