

Refractory cardiogenic shock in an infant with congenital hypothyroidism

Syed Ahmed Zaki, Amit Dolas

Abstract

Thyroid dysfunction causes remarkable cardiovascular derangements. Both systolic and diastolic dysfunction of the heart can occur in hypothyroidism leading to cardiac arrhythmia and congestive heart failure. Refractory cardiogenic shock and hypotension in congenital hypothyroidism is rare. We describe a 5-month-old female infant with congenital hypothyroidism and refractory cardiogenic shock. Cardiac function and hemodynamic stability were restored after starting levothyroxine therapy.

Keywords: Congenital hypothyroidism, hypotension, levothyroxine, refractory shock.

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Introduction

Congenital hypothyroidism is the most common congenital endocrine disorder and occurs at rate of one in 3000-4000 births.^[1] As the clinical features of congenital hypothyroidism are often subtle, many newborns remain undiagnosed at birth. Various unusual presentations of congenital hypothyroidism have been described.^[2] It is important for physicians to be aware of such presentations, especially, in areas where screening for congenital hypothyroidism is absent. We present an infant who had refractory cardiogenic shock as the presenting feature of congenital hypothyroidism.

Case Report

A five-month-old female infant presented with refusal to feed, lethargy and vomiting for three days. There was no history of fever, urinary complaints, recurrent respiratory tract infections or hospitalisation in the past. There was history of passing hard stools once every five days since two months. Birth history was normal. There was no

history of prolonged neonatal jaundice, delayed passage of meconium or feeding difficulties after birth. There was no history of antenatal exposure to antithyroid drugs. The patient had not achieved head holding and social smile. She had a hoarse cry and an apparently normal hearing. Family history was normal. On admission, she was lethargic with delayed capillary refill time. Her peripheral extremities were cold. The heart rate was 80/min with poor peripheral pulses, respiratory rate was 38/min and blood pressure was 52/34 mmHg. She had dry coarse mottled skin. There was excessive hair on forehead, macroglossia, distended abdomen and umbilical hernia [Figure 1]. There was no icterus, edema, cyanosis or lymphadenopathy. Mild pallor was present. Anterior fontanelle was large with wide open sutures. There was no swelling in the neck. Generalised hypotonia with delayed relaxation of deep tendon reflexes was present. On cardiovascular system examination, ejection systolic murmur grade 3/6 was heard in the pulmonary area. Second heart sound was wide and fixed split. Generalised distension of the abdomen was present. Liver was tender with a span of 6 cm in the midclavicular line. Spleen was not palpable and there was no evidence of free fluid.

Fluid resuscitation with 20 ml/kg normal saline was given on admission followed by dopamine infusion @10 µg/kg/min. Intravenous cefotaxim and amikacin

From:

Department of Pediatrics, Lokmanya Tilak Municipal Medical College and General Hospital, Sion, Mumbai-400 022, India

Correspondence:

Dr. Syed Ahmed Zaki, Room no.509, new RMO quarters, Sion, Mumbai-400022, India. E-mail: drzakisyed@gmail.com



Figure 1: Clinical photo of the patient showing excessive hair on forehead, macroglossia, distended abdomen and umbilical hernia

were also started. Investigations revealed: hemoglobin 9.8 gm%, total leucocyte count 8900/cumm and platelet count 4.2 lac/cumm. Her liver and renal function tests, serum electrolytes were normal. Serum calcium was 7.8 mg/dL and alkaline phosphatase was 910 IU. Her blood culture and urine culture were normal. Thyroid profile showed: serum T3 < 10 ng/dL (N-105-245), serum T4 1.20 μ /dL (N 7.8-16.5) and TSH 221.67 mIU/ml (N - 0.8- 8.2), suggestive of primary hypothyroidism. Ultrasound of the neck showed absent thyroid gland. Technetium 99 thyroid scan revealed absent thyroid gland in the neck with no evidence of functioning ectopic thyroid tissue. X-ray of lower limb revealed absent distal femoral epiphyses [Figure 2]. Chest X-ray showed cardiomegaly. Echocardiography revealed a left ventricular ejection fraction (LVEF) of 35% with global hypokinesia. A 5 mm of ostium secundum atrial septal defect was also present. Electrocardiogram (ECG) showed low voltage P waves with diminished amplitude of QRS complexes. The next day in view of persistent hypotension, dopamine infusion was increased to 15 μ /kg/min and dobutamine @10 μ g/kg/min was added which was increased to 15 μ g/kg/min. However, even on the third day of admission the hypotension persisted in spite of high dose dopamine and dobutamine infusion. The heart rate was 96/min with poor peripheral pulses, respiratory rate was 32/min and blood pressure was 54/30 mmHg. The liver was still tender with a span of 7 cm. Hypothyroidism was considered to be responsible for the acute refractory hypotension and cardiogenic shock. Oral levothyroxine was started at 10 mcg/kg and was increased to 15 mcg/kg mg over 48 hours. After 48hrs, the heart rate was 106/min with good peripheral pulses and the blood pressure was 84/60 mmHg. The dopamine and dobutamine infusion was tapered and omitted. The patient was discharged

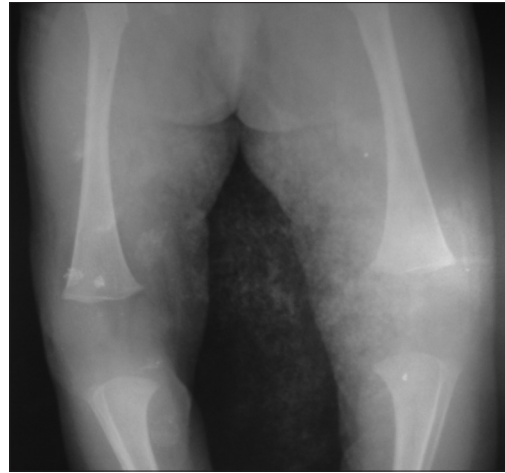


Figure 2: X-ray of lower limb revealed absent distal femoral epiphyses

on levothyroxine after ten days of admission. A repeat echocardiography done after one month revealed normal left ventricular function with LVEF of 60%.

Discussion

The effects of thyroid hormone on cardiovascular system are well documented. Cardiovascular manifestations of hypothyroidism include pericardial effusion, weak arterial pulses, bradycardia, hypotension, non-pitting facial and peripheral edema muffled heart sound, and evidence of congestive heart failure such as ascitis, orthopnea and paroxysmal dyspnea.^[3,4] The chronotropic response and normal tension of the heart muscle in diastole depends on tri-iodothyronine. Moreover, tri-iodothyronine expression in the cardiac muscle affects the number of β -adrenergic receptors and their sensitivity to catecholamines.^[3] Hence, patients with profound hypothyroidism with cardiogenic shock have a poor response to catecholamines. Severe hypothyroidism causes muscle relaxation and the isovolumetric relaxation and contraction time becomes prolonged and myocardial performance index (MPI) increases indicating systolic and diastolic dysfunction.^[5] The ECG changes seen in hypothyroidism include sinus bradycardia, low voltage QRS complexes, prolonged QT interval, low P wave amplitude, right bundle branch block and ventricular dysrhythmias, including torsades de pointes.^[3-6] These cardiovascular effects are usually reversed after starting thyroxin. While cardiac dysfunction is well documented secondary to hypothyroidism, hypotension and cardiogenic shock is rare and is resistant to conventional therapy in the form of intravenous fluids and vasopressors.^[5,7] In such cases, urgent levothyroxine replacement can restore hemodynamic stability by reversing the systolic and diastolic dysfunction, as seen in our case. Similar cases

have been described in adults. Dharmasena *et al* and Gupta *et al*, have described a 52 year old male and a 62-year-old female respectively with hypothyroidism.^[5,7] Both the cases presented with cardiogenic shock and did not respond to intravenous fluids, vasopressors or steroids. They showed improvement only after starting levothyroxine. An extensive PUBMED search with keyword 'congenital hypothyroidism' and 'cardiogenic shock' revealed only a single case described by Drop *et al*. They have described a 6-week-old girl who presented with cardiogenic shock and had a partial thyroid hormone unresponsiveness of the pituitary due to congenital elevation of thyroid binding albumin.^[8] Another unusual feature seen in our patient was the presence of hypertrichosis. Hypothyroidism is usually associated with loss of hair. The association of hypertrichosis with congenital hypothyroidism is rare. The stimulation of hair growth via adrenal androgens by TSH may be the probable pathophysiological mechanism. Zaki *et al*. have suggested that full assessment of thyroid function be carried out in all cases of hypertrichosis or abnormal distribution of body hair.^[9]

Conclusion

Physicians should be aware that refractory cardiogenic shock can occur in congenital hypothyroidism. Urgent

levothyroxine therapy is essential for improving cardiac function and restoring hemodynamic stability.

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