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Pediatrics

A Massive Pericardial Effusion Secondary to Hypothyroidism in a Child with Downs Syndrome

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Case Report

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Abstract: Though common in adults, reports of massive pericardial effusion is rare in children secondary to hypothyroidism. Here we report very unusual case of massive pericardial effusion secondary to hypothyroidism in Downs syndrome child. A 10 year old girl presented to pediatric casualty with severe respiratory distress and fever with typical phenotypic features of Downs syndrome and clinical features of hypothyroidism. On further evaluation child was found to have massive pericardial effusion, severe primary hypothyroidism and Downs syndrome. Child improved with thyroxine supplementation.
Keywords: Hypothyroidism, Downs' syndrome, pericardial effusion.

INTRODUCTION

Downs syndrome is the most common genetic cause of moderate intellectual disability. Incidence of Downs syndrome is approximately 1 in 733 live births [1]. Congenital anomalies are common in a child with Downs syndrome. Endocrine disorders in Downs are hypothyroidism, Diabetes Mellitus, Obesity and Hyperthyroidism. Hypothyroidism can be congenital (1%) or acquired (5%) [2].

Hypothyroidism is common endocrinal disorder with multiorgan involvement including cardiovascular system. The occurrence of pericardial effusion in hypothyroidism is related to the severity and duration of the disease. Although small pericardial effusion is a frequent occurrence but moderate to massive pericardial effusion with or without tamponade is rare and often associated with severe form of disease or myxedema [3, 4].

Herein we present 10 year old girl with massive Pericardial Effusion secondary to severe Hypothyroidism in Downs Syndrome.

CASE REPORT

A 10 year old girl born to non consanguineous couple at hospital by full term normal vaginal delivery with no significant neonatal history, with global developmental delay came to pediatric casualty with history of difficulty in breathing from past 7 days and running mild to moderate grade fever from past 7 days. Difficulty in breathing was insidious in onset and gradually progressive which was worsening with sleeping in supine position and was associated with intermittent noisy breathing and breathlessness. It was better when child sits with bending forward position. Fever was insidious in onset, mild degree and was gradually progressive in nature and intermittent type not associated with chills or rigor.

On examination child was conscious, irritable, febrile and in severe respiratory distress with respiratory rate around 64 per minute and SpO_2 60-64% without Oxygen, pulse rate 80 per minute, BP 76/60. General physical examination revealed features of Downs syndrome like depressed nasal bridge, short and stubby fingers, simian crease, generalized hypotonia, protruded tongue, short stature and features of hypothyroidism like short stature with height 95cm, weight 14kg at admission, head circumference 44cm, MAC 14.7 cm, US:LS ratio 1.5:1, dry skin, generalized Hypotonia, mental retardation & umbilical hernia (Fig-1).

Systemic examination revealed reduced air entry on left lung field with bilateral basal crepitations and inspiratory stridor. Cardiac examination revealed distant and muffled heart sounds. Abdomen was protruded with hepatomegaly of 5cm below right costal margin, with umbilical hernia. CNS examination showing obvious mental dullness and obtundation.

Child was stabilized with Oxygen and IV fluids and started on antibiotics in view of pneumonia.

Child was investigated with complete blood count, electrolytes and serum creatinine blood urea and chest x-ray. Blood investigations were within normal limits, but chest x-ray was showing massive pericardial effusion with loss of normal cardiac silhouette (Fig-2). ECG showing low voltage waves with bradycardia. Child was further evaluated for assessment of bone age which revealed 3 years old. (Fig-3) Thyroid function tests revealed severe primary hypothyroidism and rise anti TPO antibodies and anti-Thyroglobulin antibodies suggesting autoimmune hypothyroidism.

Fig-1: Revealing features of Downs syndrome like depressed nasal bridge, short and stubby fingers, simian crease, protruded tongue, short stature

2D echo showed a large pericardial effusion with no signs of tamponade. Child was investigated for Downs syndrome which revealed 21 trisomy by karyotyping.

With these supportive investigations diagnosis of Downs syndrome with severe primary hypothyroidism with massive pericardial effusion was made.

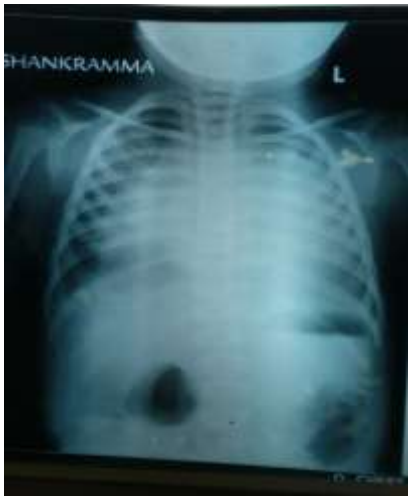

Fig-2: Chest x-ray was showing massive pericardial effusion with loss of normal cardiac silhouette

Child was treated with levothyroxine 4microgram/kg body weight, oxygen, nebulisation and antibiotics for 7-10 days. Her general condition got better by thyroxine supplementation after around 3 weeks. Pericardiocentesis was performed for diagnostic purpose and around 10-15ml of fluid is removed. Since respiratory distress and oxygen dependency got better by thyroxine treatment continued.

Fig-3: Xray of left elbow AP & lateral view revealing bone age of 3years

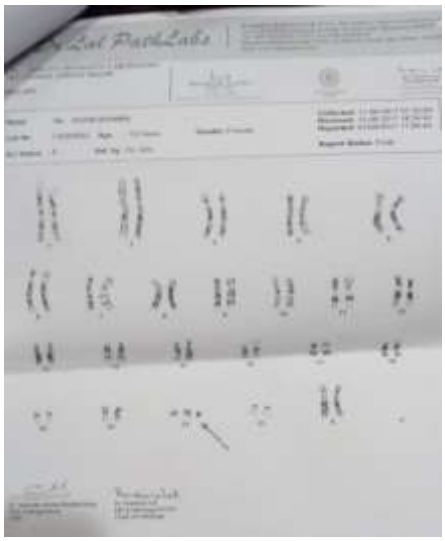

Fig-4: Showing karyotyping of the patient with trisomy 21

DISCUSSIONS

Pericardial Effusion is most common cardiac manifestation of Hypothyroidism in 10-30% of adults [5]. However these findings appear to be rare in children and adolescence [5]. Incidence of Hypothyroidism in patients with Downs Syndrome is high, but diagnosis is often delayed or missed for lack of specific clinical clues [11]. In a large study of congenital hypothyroidism in neonates with Downs syndrome, a prevalence of 0.12 % was found [12] A small PE frequently be found in cases of Downs Syndrome with hypothyroidism. In these patients PE is completely resolved by Thyroxine therapy [5, 6]. Hypothyroidism is associated with increased capillary permeability and subsequent leakage of proteins into the interstitial space , resulting in pericardial effusion, however tamponade is rare [13] In hypothyroidism the accumulation of fluid in pericardial space usually occurs very slowly, which is usually recognized before it goes for massive PE and resolves with thyroxine. Massive PE is rare and is often associated with severe and prolonged form of the disease [7].

To date, 4 children and adolescent with hypothyroidism who have massive PE without Downs Syndrome have been reported in literature [7- 10].

The most common and prominent manifestation of hypothyroidism in children is profound Severe growth failure with Mental retardation[10].

CONCLUSION

Hypothyroidism is a common cause of symptomatic PE in adults. But any child with DS with hypothyroidism who present with cardiomegaly without CHD should be suspected of having PE and echocardiography and TFT should be performed immediately and treated for hypothyroidism.

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