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Case Report
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A Phenomenal Case Report of an Autoimmune Thyroiditis Masquerading Symptomatic Pericardial Effusion in a Child with Down's Syndrome

Betsy Mathew and Ananta Jois*

Department of Pediatric Critical Care, GG Hospital, India

Abstract

Hypothyroidism is a common endocrinal disorder with multi-organ involvement including cardiovascular system. Although small Pericardial Effusion (PE) is a common clinical finding but moderate to massive PE with or without tamponade is rare. Incidence of hypothyroidism in patients with Down's syndrome is high, but diagnosis is often delayed or missed for lack of specific clinical clues. In a study done on congenital hypothyroidism in neonates with Down's syndrome, a prevalence of 0.12% was found. Hashimoto Thyroiditis (HT)/Autoimmune thyroiditis (secondary hypothyroidism) is the most frequent cause of acquired hypothyroidism in pediatric age groups. HT is usually diagnosed in adolescents, especially females. It is rare in infants and toddlers with cardiac involvement, including pericardial effusion that is seen in 10% to 30% of adult HT cases. Pericardial effusions have been reported in 50% to 73% of pediatric patients with hypothyroidism in various series but none of these describes the symptomatic pericardial effusions or cardiac tamponade.

Keywords: Thyroiditis; Down's syndrome; Hypothyroidism; Hashimoto thyroiditis

Introduction

Hypothyroidism is a common endocrinal disorder with multi-organ involvement including cardiovascular system. Although small Pericardial Effusion (PE) is a common clinical finding but moderate to massive PE with or without tamponade is rare [1]. This is often associated with severe form of the hypothyroidism [2]. The accumulation of fluid in pericardial space occurs in a slow manner, and is seen in severe form of untreated primary/secondary hypothyroidism. Incidence of hypothyroidism in patients with Down's syndrome is high, but diagnosis is often delayed or missed for lack of specific clinical clues [3]. In a study done on congenital hypothyroidism in neonates with Down's syndrome, a prevalence of 0.12% was found [4].

Hashimoto Thyroiditis (HT)/autoimmune thyroiditis (secondary hypothyroidism) is the most frequent cause of acquired hypothyroidism in pediatric age groups. HT is usually diagnosed in adolescents, especially females. It is rare in infants and toddlers with cardiac involvement, including pericardial effusion that is seen in 10% to 30% of adult HT cases [5].

Pericardial effusions have been reported in 50% to 73% of pediatric patients with hypothyroidism in various series but none of these describes the symptomatic pericardial effusions or cardiac tamponade [6].

Case Presentation

A 11 years old, male child with congenital heart disease (mild VSD) corrected with age, immunized to date, with developmental delay, on regular home diet who was diagnosed as Down's syndrome at few days of life (chromosomal analysis) was admitted to the Pediatric Intensive Care Unit (PICU) with complaints of progressive breathlessness since 3 days. No history of fever or previous hospitalization for the same. Patient was not on any regular medication.

At admission

Airway: Not maintained in room air.

Breathing: RR-46/min, Chest expansion –R>L, No added sounds, Intercostal retraction {+}, Sub costal retraction {+}, Spo2- 70% to 72% in room air.

Non-Invasive Ventilation (HHHFNC) initiated at 60% Fio2, 35 L/min flow. Blood gas analysis

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Correspondence:
Betsy Mathew, Department of Pediatric Critical Care, GG Hospital, Flat 12E, Artech Deepam, Anayara, Trivandrum, 695029, Kerala, India, Tel: 8281736759; E-mail: betsymathew001@gmail.com

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showed metabolic acidosis with hypoxemia. After 1 h to 2 h of initiation, his RR-28 to 30/min & Spo2>94%. NIV support given for 5 days, weaned & stopped as tolerated. On Day 6, low flow O2 initiated. His respiratory status & blood gas analysis maintained normal. On Day 7, he was shifted to ward on portable low flow O2 systems. His mother was trained for its use & educated on Spo2 monitoring. During PICU stay, he had daily episodes of deep sleep apnea at night which gradually reduced by day 6. He was discharged home on portable O2 support (o.5 L/min to 1 L/min).

Circulation: At admission, HR-102/min (NSR), all central & peripheral pulses well felt, color & temperature normal, CFT<2 sec, BP-112/80(88) mmHg.

ECHO (at admission)-moderate pericardial effusion (posterior-7 mm, anterior-2 mm, apical- 2 mm, lateral- 3-4 mm), Normal LVEF function, No significant PAH.

Throughout PICU stay, no evidence of hemodynamic instability. Daily ECG monitoring done to assess QRS voltage & no evidence of cardiac tamponade.

Disability: GCS-15 at admission, pupils equal & reactive, no hypoglycemia throughout PICU stay.

Exposure: No evidence of fever/rash throughout hospital stay.

Head to toe examination & general examination in Figure 1

- Short stature (constitutional): Protuberant belly
- Obesity (BMI-21): Macroglossia
- Short hands & limbs: Flat feet & Genital normal
- Short neck & upward slanting of eyes
- Moon face & depressed nasal bridge

Systemic examination: CVS - Muffling of heart sounds +, all other systemic examination WNL

Supportive care:

- Mild fluid restriction based on daily intake - output monitoring was done.
- Daily weight monitoring done. At admission-38 kg, At discharge-35 kg
- Third generation Cephalosporin IV antibiotic given as prophylaxis.

Figure 1: Child showing features of Down's syndrome.

Diagnostic interventions

- Complete blood counts, C- reactive protein, blood culture, renal & liver function – normal
- Endocrinology consultation done. Thyroid function showed TSH>100 with low T3 & T4 & Anti-Thyroprotein antibody elevated (113) at admission. Bone Age assessment done with left hand-wrist X-ray (corresponding to 6.5 years old). Thyroxin tablet was initiated at 115 mcg from day 1 of admission & dose adjusted as improvement seen.
- ECHO done on daily basis by Pediatric cardiologist & by day 5, mild pericardial effusion was documented.
- TSH at discharge reduced to 60.
- 2 weeks after discharge, follow up ECHO-No pericardial effusion, Normal LVEF, No significant PAH. Repeat TSH- normalized after one month (Figures 2-4).

Discussion

PE in hypothyroidism is considered to be caused due to the generalized polyserousopathy and the main pathogenic mechanism being a combination of extravasation of albumin and inadequate lymphatic drainage. It accounts for the exudative nature of the

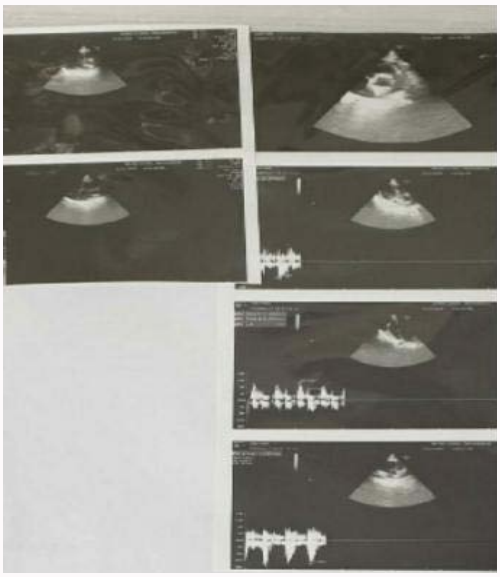

Figure 2: Echo shows moderate PE.

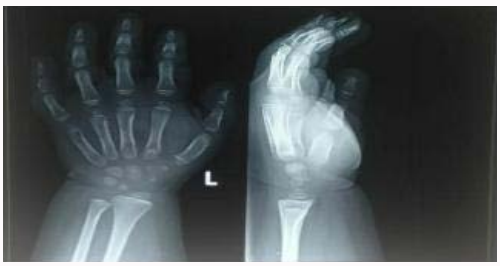

Figure 3: Bone age estimation - left hand/wrist x-ray.

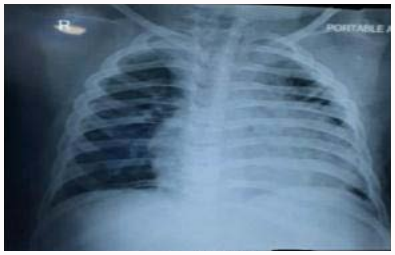

Figure 4: Chest X-ray shows moderate PE.

accumulated fluid in this condition. The pericardial fluid in hypothyroidism is characteristically straw color due to high content of alpha and beta globulins with predominance of lymphocytes [7]. In our case, pericardiocentesis was not indicated as the effusion (moderate) got resolved with adequate airway support (HHHFNC) and medical management [8]. Treatment of hypothyroidism is always mandatory following resolution of the acute crisis. There is generally a residue of effusion, which disappears following appropriate therapy over a period between one month and one year. It is for this reason, treatment with increasing doses of Levo-thyroxine is essential. Thyroid replacement therapy is indicated for a long period of time and requires follow-up.

Conclusion

Hypothyroidism should be kept in mind for any child with unexplained pericardial effusions. Early diagnostic treatment with thyroid hormone replacement will eliminate the need for unnecessary diagnostic procedures and invasive treatment measures. Appropriate control of pericardial effusion can reduce the risk of progression to cardiac tamponade.

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