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Case Report

A viral trigger for a metabolic crisis: parainfluenza induced decompensation in a child with glycogen storage disease type III and complex congenital heart disease

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ABSTRACT

Glycogen storage disease type III (GSD III) also known as Cori's disease is a rare autosomal recessive metabolic disorder caused by the deficiency of glycogen debranching enzyme (amylo-1,6-glucosidase), leading to hepatomegaly, hypoglycemia, ketosis, and variable cardiac involvement. Intercurrent infections may precipitate metabolic decompensation, particularly in young children. This case report describes a 21-month-old baby boy, known case of GSD III with complex congenital heart disease who presented with fever and respiratory symptoms secondary to parainfluenza infection, complicated by metabolic acidosis, ketosis, electrolyte imbalance and liver enzyme derangement. Prompt recognition and early initiation of dextrose containing fluids prevented further deterioration. This case highlights the importance of early metabolic support during acute illness in children with GSD III, especially in those with coexisting cardiac disease.

Keywords: Glycogen storage disease type III, Cori's disease, Debranching enzyme deficiency, Metabolic decompensation, Parainfluenza, Congenital heart disease, Toddler

INTRODUCTION

Glycogen storage disease type III (GSD III) also known as Cori's disease due to deficiency of the glycogen debranching enzyme (amylo-1,6-glucosidase), defect at AGL gene on chromosome band 1p21.1.[1] It is an autosomal recessive disorder caused by the buildup of a complex sugar called glycogen in the body's cells. The accumulated glycogen is structurally abnormal and impairs the function of certain organs and tissues especially liver, muscles and heart. Affected children typically present with hepatomegaly, fasting hypoglycemia, ketosis, hyperlipidemia, and elevated transaminases. The estimated incidence is approximately 1 in 100,000 live births. Clinically, GSD III are usually diagnosed in infancy or in early childhood with hepatomegaly, hypoglycemia, ketosis and elevated transaminases. Intercurrent illnesses, especially viral infections, can precipitate catabolic stress leading to metabolic decompensation.[2] The coexistence of congenital heart disease (CHD) further increases clinical complexity. Here describes a 21-month-old baby boy with GSD III with complex CHD presenting with acute metabolic derangement triggered by parainfluenza infection.

CASE REPORT

A 21-month-old baby boy, born at 37 weeks and 5 days of gestation via spontaneous vaginal delivery with a birth weight of 3.45 kg. He is a known case of glycogen storage disease type III with complex congenital heart disease and was brought to the emergency department with a history of fever (maximum recorded temperature 38.5 °C), dry cough, and nasal congestion. The fever was noted early in the morning, and an attempt to administer paracetamol syrup resulted in a single episode of vomiting. There was

no history of persistent vomiting, apnea, cyanosis, reduced oral intake, diarrhea, urinary symptoms, or rash. The mother reported slightly reduced activity. However, the child was waking up, interactive, and asking for food. A significant sick contact was present, as the mother had symptoms of upper respiratory tract infection. The child was on regular medications for heart failure management.

On examination, the child was febrile with mild dehydration but hemodynamically stable. Respiratory examination revealed mild tachypnea during febrile episodes without signs of respiratory distress. Abdomen was distended with huge hepatomegaly, not tender and spleen palpable. CVS showed normal S1 and S2 with murmur and no gallop.

Initial laboratory evaluation revealed metabolic acidosis with respiratory compensation on blood gas analysis. Serum bicarbonate was 13 mmol/l, potassium was 3 mmol/l, sodium was 142 mmol/l, and lactate was elevated at 4.9 mmol/l. Capillary blood glucose was 152 mg/dl. Blood ketones were elevated at 1.9 mmol/l, suggestive of ketotic stress. Liver function tests showed deranged transaminases compared to previous trends, with a markedly elevated gamma-glutamyl transferase (GGT) in the 400s, suggestive of cholestasis. Alkaline phosphatase was within normal limits. CPK was elevated at 1874 U/l. Procalcitonin was elevated at 2.25 ng/ml, while CRP was negative. Respiratory viral panel was positive for parainfluenza virus.

Chest radiography demonstrated apparent cardiomegaly with a cardiothoracic ratio of 0.62 but no focal consolidation or infiltrates. USG of abdomen revealed moderate hepatomegaly with diffuse increased coarse liver echo texture. Echocardiography performed previously showed a large secundum atrial septal defect, multiple muscular ventricular septal defects (Swiss cheese type), pulmonary valve and supravalvular stenosis, and a dilated right heart (Figure 1).

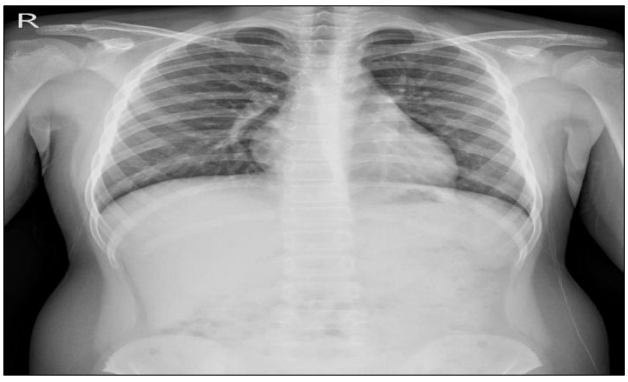

Figure 1: Chest radiography.

The child was managed with initial normal saline and 10% dextrose boluses followed by maintenance IV fluid using D10 normal saline with potassium supplementation. Enteral feeds were continued to prevent catabolism and supportive care was provided for upper respiratory symptoms. The mainstay treatment for GSD III is mainly dietary. Long term dietary management with frequent carbohydrate intake, uncooked cornstarch, and high protein diet remains the cornerstone of therapy.

The child remained hemodynamically stable with improvement in metabolic parameters following dextrose containing fluids. Ketone levels and acid base status showed gradual normalization. No progression to hypoglycemia or cardiac decompensation was noted.

DISCUSSION

GSD III due to the deficiency of the glycogen debranching enzyme, causing defective glycogen breakdown and results in fasting hypoglycemia, hepatomegaly, hyperlipidemia, ketosis, myopathy and cardiac involvement. Intercurrent illnesses like viral infections precipitate metabolic decompensation by increasing catabolic demand and reduce glycogen availability.[3]

In this child, parainfluenza viral infection acted as the precipitating factor for acute metabolic stress. Despite adequate oral intake and absence of hypoglycemia, the child developed ketotic metabolic acidosis with elevated lactate which indicates impaired hepatic glucose release and increased anaerobic metabolism. Ketosis in the presence of euglycemia highlights the susceptibility to early metabolic instability during intercurrent illness. Elevated CPK suggests skeletal muscle involvement or myopathy. The markedly elevated GGT with relatively normal alkaline phosphatase suggests evolving cholestatic liver involvement, which is a recognised hepatic manifestation in GSD III due to chronic glycogen accumulation and progressive hepatic injury. Transaminase derangement indicates acute hepatic stress. Raised procalcitonin with normal CRP and no bacterial source suggest that the inflammation is more likely due to viral infection.

The coexistence of complex congenital heart disease significantly increases the risk of decompensation in this patient. Fever, dehydration, metabolic acidosis, and electrolyte disturbances can exacerbate heart failure and arrhythmias. Early recognition of mild dehydration and prompt correction with isotonic fluids, followed by dextrose containing infusions prevented progression to hypoglycemia, severe acidosis, or cardiac instability.[4]

Management of acute illness in GSD III focus on prevention of catabolism. Continuous glucose delivery through intravenous dextrose, continuation of enteral feeds, and avoidance of fasting are critical. In this case, the use of D10 normal saline with potassium supplementation effectively corrected metabolic derangements while maintaining euglycemia.[5]

The cornerstone of management for GSD III is dietary therapy. Carbohydrates should be given frequently at

every 3-to-4-hour interval to maintain euglycemia. But it was associated with high insulin levels and life-threatening hypoglycaemia when feeds were interrupted. Uncooked cornstarch emerged as the preferred therapy due to its low glucose release and lower insulin stimulation. And it has neuroprotective effect during hypoglycaemia and improved metabolic stability when administered frequently. A high protein diet is recommended while fat intake should be reduced. Dietary and nutritional therapy has turned GSD from a fatal disorder into one with an excellent long-term prognosis.[5,6]

This case emphasizes that metabolic decompensation in GSD III may present with ketosis and acidosis without overt hypoglycemia and that viral infections can significantly disturb metabolic homeostasis. Early intervention based on biochemical trends rather than waiting for symptomatic hypoglycemia is essential.

CONCLUSION

This case highlights an acute metabolic decompensation triggered by a viral upper respiratory infection in a child with GSD III with congenital heart disease. GSD III is a rare genetic disorder with nonspecific findings and may lead to severe complications. It is important to realise the symptoms of this disease and appropriate genetic testing should be done immediately to ensure initiation of early treatment. Children with GSD III are highly vulnerable to metabolic instability during intercurrent illnesses. This case emphasises that viral infections can precipitate ketotic metabolic acidosis without hypoglycemia and also highlights the importance of early metabolic support to prevent life threatening complications, especially in the presence of complex congenital heart diseases. The case underscores the importance dietary therapy, particularly frequent carbohydrate intake and uncooked starch as the cornerstone of both acute and long-term management of GSD III.

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