

endothelium non-thrombogenic protective factors may have been the cause [2].

There are only a handful of reported cases where deep vein thrombosis have been reported in direct association with dengue fever [2,7]. Thrombotic events in large veins [ileo-femoral deep vein thrombosis (DVT), pulmonary thromboembolism, mesenteric vein thrombosis] in DF patients have been reported from Brazil in 5.4% of all dengue inpatients.

The dilemmas posed in treating a blood clot in a patient who is at risk for excessive bleeding were challenging.

Awareness for these thrombotic complications is recommended to all practitioners who treat dengue in hospital settings

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Neonatal Aortic Thrombosis as a Result of Congenital Homocystinuria

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Background: Arterial thrombosis, that too in aorta is rare in neonates. **Case characteristics:** A 4-day-old presented with non-recordable BP in lower limbs. Doppler ultrasonography of abdomen revealed aortic thrombus. **Observation:** Serum homocysteine level was elevated (25.5 $\mu\text{mol/L}$). **Outcome:** Thrombus resolved with subcutaneous LMW heparin therapy for 2 weeks. **Message:** Congenital classic homocystinuria can rarely cause aortic thrombosis in neonatal period.

Keywords: Congenital aortic thrombosis, Congenital classic Homocystinuria, Neonate.

Thrombotic diseases are rare in neonates. The main known risk factors at this age are perinatal asphyxia, dehydration [1], umbilical arterial catheterization [2] and inherited thrombophilia [3,4]. Inherited thrombotic disorders become manifest in <5% of affected children [5]. Arterial thrombosis, even more rare than venous thrombosis, rarely occurs in the aorta. Most of the described cases of aortic thrombosis are associated with the catheterization of an umbilical artery. We hereby describe a case of abdominal aortic thrombosis due to congenital classic homocystinuria.

CASE REPORT

A four-days-old full term female infant, born vaginally through meconium stained amniotic fluid, presented to us with respiratory distress, lethargy and poor feeding. Infant had cried immediately after birth. There was no history of umbilical arterial catheterization. The mother had pregnancy induced hypertension and two spontaneous second trimester abortions in the past. There was no family history of thrombotic events. Examination revealed mild respiratory distress with Downe's score of

3 and normal heart rate, with absent femoral pulsations. Blood pressure was normal in upper limbs and was not recordable in lower limbs. Pulse oximetry revealed saturation of 95-96% in upper limbs and 91-92% in lower limbs. Systemic examination was normal. A provisional diagnosis of hypoplastic left heart syndrome was made.

Investigations showed normal renal functions, serum electrolytes and serum calcium, and negative sepsis screen. Chest radiograph was suggestive of meconium aspiration syndrome. Echocardiography depicted structurally normal heart. Doppler ultrasonography of abdomen revealed echogenic aortic thrombus (2.7x1.3cm) distal to origin of inferior mesenteric artery upto bifurcation of the abdominal aorta.

The baby was started on low molecular weight heparin (1.5 mg/kg/dose 12 hourly) via subcutaneous route. In view of abdominal aortic thrombosis with no classic predisposing factors, tests for prothrombotic disorders were sent. Lupus anticoagulant, anticardiolipin antibody, antithrombin III levels, protein C, protein S levels were within normal limits and Factor V Leiden mutation was negative. Serum homocystine levels were 25.51 micromole/L (normal range 0-10 micromole/L) by CMIA technology and qualitative test of urine homocystine was positive. After two weeks of therapy, lower limb pulses were palpable. Repeat doppler study revealed complete resolution of thrombus. The aorta was recanalized with restoration of blood flow distal to the obstruction after four weeks of therapy. In view of diagnosis of congenital homocystinuria, infant was started on pyridoxine, folic acid and betaine, and methionine-restricted diet. Additional work-up of homocystinuria revealed normal ophthalmological examination and normal peripheral smear. Serum methionine levels were high 18 mg/dl (normal range <1 mg/dl) and test for MTHR gene mutation was negative. The baby's parents, screened subsequently had normal values of serum homocystine and methionine. This confirmed the diagnosis of classic homocystinuria due to cystathionine beta synthase deficiency. The patient responded to above stated treatment with normalization of serum homocystine values after three months of therapy.

DISCUSSION

Thrombotic diseases are rare in neonates. A German study reported a prevalence of 5.1 per 100,000 live births [5], and a multicenter study reported a prevalence of 2.4 per 1000 NICU admissions [6]. In both studies, the thrombotic manifestations mainly involved large venous vessels as central line complications. To the best of our knowledge, there is no documented case in medical

literature citing congenital classic homocystinuria causing aortic thrombosis in neonatal period.

The optimal treatment depends on the availability of surgical expertise, the associated risk factors for bleeding and degree of organ ischemia. Recent recommendations from the Seventh American College of Chest Physicians Conference on Antithrombotic and Thrombolytic Therapy [7] suggests "the urgent, aggressive use of thrombolytic or surgical therapy supported by anticoagulation with heparin or low molecular weight heparin for children experiencing spontaneous aortic thrombosis with evidence of renal ischemia. It is unclear which thrombolytic agent is most effective; however, tissue plasminogen activator has become the agent of choice for several reasons, including experimental evidence of improved clot lysis *in vitro* compared with that using urokinase and streptokinase, fibrin specificity and low immunogenicity [7].

The support of thrombolysis with concomitant heparin may be synergistic; however, whether LMWH or unfractionated heparin (UFH) is better is still unclear. A case series by Klinger, *et al.* [8] suggests successful treatment of severe aortic thrombosis in two neonates with LMWH alone. Our patient was successfully treated with LMWH and showed complete improvement after completion of therapy.

This case report underlines the importance of inherited thrombophilia as the cause for isolated aortic thrombosis in neonates. Once the diagnosis of homocystinuria is established it is imperative to subclassify it according to the enzyme defect as the treatment modality and future health implications differ amongst the three common subtypes.

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Pheochromocytoma Presenting as Diabetes Insipidus

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Background: Pheochromocytomas are catecholamine producing tumors that classically present with the triad of sweating, palpitations and headache. **Case characteristics:** 9-year-old boy whose only presenting complaints were polyuria and polydipsia for 2 years. **Observation:** Routine measurement of blood pressure detected mild hypertension, and subsequent investigations revealed bilateral pheochromocytoma. **Outcome:** Surgical removal of the tumors resulted in complete resolution of polyuria and polydipsia. **Message:** The case highlights the importance of measuring BP for children as part of physical examination.

Keywords: *Diabetes insipidus, Pheochromocytoma, Polydipsia, Polyuria.*

Pheochromocytomas and paragangliomas are rare, with an estimated annual incidence of 2-8 per million population [1]. Hypertension, paroxysmal or sustained, is the most consistent finding [2]. The classical triad of symptoms, headache, palpitations and excessive sweating, is present in up to 50-70% of patients [2]. Other relatively common symptoms are flushing, pallor, anxiety, diarrhea, fatigue and fever. Here, we report a 9-year-old boy with bilateral pheochromocytoma, whose only presenting complaints were polyuria and polydipsia for two years.

CASE REPORT

A 9-year-old boy was brought to the outpatient department with complaints of increased thirst and urination for last 2 years. The child drank 7-8 L of water and passed 6-7 L of urine per day (10-12 mL/kg/h), with multiple nocturnal awakenings. There was no history of weight loss, polyphagia, fatigue, headache, vomiting, visual complaints or any significant past illness/head injury. Parents had consulted many pediatricians in their city, but no cause had been found. He had been diagnosed as psychogenic polydipsia, and behavioral therapy advised. However, there was no improvement and the child was brought to us. On examination, the child had normal hydration, heart rate of 110/min, blood pressure

of 126/75 mm Hg (at 95th centile), and weight and height between 10th-25th centiles. Urine output was documented as 10 mL/kg/hr. The initial investigative work-up was as follows: blood sugar (fasting and post-prandial) 103 and 146 mg/dL, urine specific gravity-1.002, serum sodium 143 and potassium 4.1 mEq/L. Renal function, blood gas, serum calcium, thyroid function, serum cortisol, urine routine and urine calcium/creatinine ratio were normal. Water deprivation test was planned the next morning, but baseline urinary and plasma osmolarity were 170 and 301 mOsm/kg respectively, which established the diagnosis of diabetes insipidus (DI). It was decided to give vasopressin challenge to differentiate between central and nephrogenic DI. BP was measured before administration of vasopressin, and was found to be 130/90 mm Hg (>95th centile). Vasopressin was not administered in view of the hypertension. Ultrasonography of the abdomen revealed bilateral adrenal masses 4.1 × 3 cm (left) and 2.2 × 1.9 cm (right) with areas of cystic degeneration, suggestive of pheochromocytoma.

Plasma and urinary normetanephrines were markedly elevated (2187 pg/mL (normal <180), and 3810 µg/day (normal 0-600) respectively), while the metanephrines were within normal levels, suggesting that the