In Utero Renal Vein Thrombosis in a Preterm Newborn with Palpable Abdominal Mass at Birth Associated with Feto-Fetal Transfusion: A Case Report

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Abstract

Renal vein thrombosis (RVT) is a rare clinical condition that occurs in neonates with various underlying risk factors and it is associated with a serious prognosis for the affected kidney. Clinical suspect emerges in presence of the classical triad of RVT (macroscopic hematuria, palpable abdominal mass and thrombocytopenia), but these three elements are not always simultaneously found at the presentation. Imaging is usually needed to confirm the diagnosis in order to start, as soon as possible, a proper therapy to reduce RVT severity. Radiological examination show kidney involvement with increased size and reduced function in the acute phase, while small and atrophic features in a later stage. Anticoagulant and fibrinolytic therapies have been promoted in the past with possible clinical improvement in some circumstances even if RVT in neonates often leads to irreversible damage. We report a case of a preterm twin girl who presented at birth with abdominal mass in the left hypochondrium in absence of hematuria and thrombocytopenia: ultrasound examination was consistent with RVT and MRI and CT scan confirmed the diagnosis. The large intertwin hemoglobin difference without the degree of amniotic fluid discordance suggest a twin anemia-polycytemia sequence (TAPS), describe as a new form of chronic feto-fetal transfusion. RVT should always be considered in differential diagnosis in case of abdominal mass and hematuria in newborn.

Keywords: Renal vein thrombosis; Preterm; Newborn; Feto-fetal transfusion

Introduction

Renal vein thrombosis (RVT) is the most common thrombotic event in infancy and it occurs primarily in the neonatal population, especially within the first three days of life. Males are affected two times more than females. The clinical presentation is monolateral in about 2/3 of cases, with predominance of the left side. Many risk factors have been identified coexisting up to 80% of the affected neonates in the absence of placement of central venous catheter. Hypercoagulability, perinatal asphyxia, dehydration, infection, maternal diabetes mellitus and congenital heart disease have been reported in patients with RVT. Moreover preterm birth is considered a risk factor since approximately 1/3 of the affected infants born prematurely.

RVT occurs as an acute event and the features of its classic pathognomonic triad of presentation (abdominal mass, haematuria and thrombocytopenia) are not always present at the same time [1]. We report this case to underline the importance of early imaging to confirm the diagnosis of RVT especially in case of an incomplete clinical presentation.

Case Report

B. M. was a preterm monochorionic diamniotic monozygotic twin girl born at 35 weeks of gestational age by planned caesarean section for poor growth of one twin in the last two weeks of gestation. No antenatal administration of betamethasone was performed. Birth weight was appropriate for gestational age in both twins. Amniotic fluid was meconium stained. First and fifth minutes Apgar scores were 5 and 7 respectively; at birth she presented bradycardia, paleness and hypotonia that recovered after one minute of ventilation with facial mask, suggesting mild perinatal hypoxia. Physical examination in delivery room revealed a palpable abdominal mass in the left hypochondrium. Respiratory distress lasted for three days supported by nCPAP. Blood pressure and vital signs were normal. No macro or micro hematuria was detected.

Prenatal medical history included gestational diabetes not treated and ultrasound examination, performed 2 weeks before delivery, showed two anatomically normal foetuses. However abdominal ultrasound examination, performed 1 hour after birth, revealed an enlarged left kidney (longitudinal diameter of the left kidney was 55 mm vs. 45 mm of the right one) with low renal corticomedullary differentiation and echogenic intramedullar streaks. Moreover a big adrenal haemorrhage (45 × 20 mm) was detected (Figure 1). Doppler US (Figure 2) and MRI (Figure 3) performed 1 week later, after a complete multidisciplinary re-evaluation of the ultrasounds, confirmed the adrenal haemorrhage and showed haemorrhagic infarction of pyramids in the left kidney. Left renal vein was smaller than the right one, and blood flow in the left kidney was reduced. Inferior vena cava was not involved by thrombosis. CT scan examination showed the absence of renal filtration after intravenous contrast administration. The reports were consistent with left renal vein thrombosis.

Macroscopic haematuria appeared about two weeks after birth. Blood test performed at birth revealed severe anemia (RBC 2.830.000/ml; Hb 10,2 g/dl; Hct 29,2%) without thrombocytopenia (PLT 224000/μl) or hypercoagulability status (protein C 62%, protein S 79%, factor V Leiden mutation and MTHFR mutation absent). High D dimer level (15 mg/L vs normal <0.3 mg/L) confirmed a recent thrombotic event. The reticulocyte count increased, reflecting chronic blood loss. On the other hand, the other twin had polycytemia (Hct 49%). The presence of anemia in the twin affected by RVT and polycytemia in the other one suggested a twin anemia-polycytemia sequence (TAPS).
Blood pressure was normal during all the 23 days of hospitalization, as well as renal function (normal values of blood urea nitrogen and serum creatinine) and no antithrombotic therapy was administered. She was discharged in good clinical conditions and follow-up performed at six months of age showed left kidney atrophy with normal blood pressure.

**Figure 1:** Abdominal ultrasound scan performed 1 hour after birth. The left kidney is enlarged with low renal corticomedullary differentiation and echogenic intramedullar streaks (black arrows). It was associated with a big adrenal haemorrhage (white arrow).

**Figure 2:** Color Doppler US exam. Left renal vein was smaller than the right one (black arrow) and blood flow in the left kidney was reduced (white arrow).

**Figure 3:** MRI coronal scan confirmed the adrenal haemorrhage (white arrow) and showed hemorrhagic infarction of pyramids in the left kidney (black arrow).

**Discussion**

The RVT accounts for 16-20% of all thromboembolic events in newborn. The incidence of symptomatic RVT in neonates has been estimated to be about 2.2/100.000 live births [2]. In utero presentation is quite rare, reflecting about 7% of all renal vein thrombosis. The evocative “triad” of RVT (macroscopic hematuria, palpable abdominal mass, thrombocytopenia) is described in literature in about only 23% of affected newborns [3,4], and, as confirmed by our case, only abdominal mass may be present after delivery, while mild hematuria appeared later. Regardless to what reported in literature no prothrombotic risk factors (normal protein C, protein S, absence of factor V mutation or MTHFR mutation) were found in our patient [5]. Pathogenesis of RVT in this newborn was probably due to prenatal suffering as suggested by low Apgar score, needs of ventilation at birth and the presence of meconium stained amniotic fluid. Perinatal hypoxia and dehydration are established risk factors for RVT. The newborn had also a severe chronic anemia (donor) without a clear cause and the presence of polycytemia in the other twin (recipient) suggested a spontaneous TAPS, because no amniotic fluid discordance was detected in the fetal ultrasound performed 15 days before the delivery [6].

Monochorionic twins share a single placenta with intertwin vascular anastomoses, allowing the transfer of blood one fetus to other one and vice versa. These anastomoses are the essential anatomical substrate for the development of several complications, including twin-twin transfusion syndrome (TTTS) and twin anemia-polycythemia sequence (TAPS) [7]. TAPS is an atypical form of chronic feto-fetal blood transfusion which may occur in uncomplicated monochorionic twin pregnancies (spontaneous form) or after laser surgery for TTTS. Both form are characterized by the presence of large intertwin haemoglobin difference without the degree of amniotic fluid discordance that is required for the diagnosis of TTTS [8]. The spontaneous form of TAPS complicates approximately 3 to 5% of monochorionic twin pregnancies [6,7].
The thrombus inside the left renal vein was not directly detected at radiological examination. Abdominal ultrasound may show increased size of the affected kidney, increased echogenicity and loss of corticomedullary differentiation in the acute phase, and it is widely useful to early detect a RVT, especially in those cases in which only one of the three cardinal signs is present. However for a definitive diagnosis an MRI and a CT scan were performed. Blood pressure, as well as renal function, persisted normal and the baby did not develop hypertension during follow-up. Unfortunately left renal function was lost and the kidney became atrophic at 6 months follow-up, confirming the severity of this disease and the need of a prompt diagnosis. We only managed this baby with supportive care (hydration and prophylactic antibiotic therapy), without fibrinolytic drugs, low molecular weight heparin or other therapies, on the basis of the absence of agreement of efficacy of antithrombotic treatments in the literature [9]. Similar clinical outcome and proportion of affected kidneys becoming atrophic has been observed in neonates treated either with supportive care or heparin 3. Previous studies reported conflicting data regarding the benefit of anticoagulation with regard to long-term renal function, particularly in cases of bilateral rein vein thrombosis. Thrombolytic therapy may be considered in cases of bilateral renal vein thrombosis, especially if there is concomitant renal failure [10].

In conclusion RVT is a serious clinical condition that should always be kept in mind when a newborn presents at least abdominal mass or hematuria. Due to the variability in the clinical presentation, ultrasound appears to be very useful for the diagnosis. Unfortunately, even if rapid recognition is critical for proper management, outcome is still poor regardless of therapeutic approach. Prospective studies are needed to elucidate the optimal treatment for neonatal RVT as well as the effects of anticoagulation and thrombolytic therapies on renal short and long term outcomes of these patients.

Conflict of Interest

All the authors declared that they have no conflict of interest to declare.

References


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