CASE REPORT

# Klippel-Trenaunay-Weber Syndrome with Kasabach-Merritt Coagulopathy and Hydronephrosis

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*Received: March 04, 2015;*
*Initial review: June 05, 2015;*
*Accepted: August 28, 2015 .*

**Background**: Klippel-Trenaunay-Weber Syndrome is a rare syndrome, consisting of vascular malformation of blood and lymph vessels. **Case characteristics**: A newborn female with respiratory distress from birth, and having vascular malformation involving left thigh. **Observation:** The neonate also had hydronephrosis and developed complication of Kasabach Merritt syndrome. **Message:** Urogenital abnormalities can be present in Klippel-Trenaunay-Weber syndrome but hydronephrosis is rare. Mortality is high with development of Kasabach Merritt syndrome.

**Keywords:** *Bleeding neonate, Coagulopathy, Neonate, Vascular malformation.*

Klippel-Trenaunay-Weber syndrome (KTWS) consists of combined vascular malformation of the capillary, venous and lymphatic types [1]. Varicosities of unusual distribution is observed during infancy or childhood, along with limb enlargement [2]. Hypertrophy of the soft or hard tissues results in asymmetry of the involved extremity [3]. The clinical presentation can be extremely variable. Although urogenital abnormalities can be present, hydronephrosis is rare in KTWS [4]. Kasabach-Merritt syndrome, a consumptive coagulopathy, can complicate around 45% of cases of KTWS. High output cardiac failure in KTWS is usually secondary to anemia and large arterio-venous malformation [5]. We present a case of KTWS with hydronephrosis, who developed Kasabach-Merritt syndrome.

## CASE REPORT

A newborn female was referred to our hospital at six hours of life with complaints of respiratory difficulty since birth, and enlarged left thigh with vascular malformation. She was born by caesarian section at 36 weeks gestation, and cried immediately after birth. Her weight was 3.5 kg. Mother had infrequent antenatal check-up and an unremarkable medical history.

At admission, she had tachycardia, tachypnea, grunting, prolonged capillary refill time, hypoxia ($SpO_2$ 85% on oxygen by prongs) and feeble pulses. Systemic examination revealed bilateral crepitations, grade III systolic murmur at upper right sternal border, and a palpable liver (2.5 cm). There was a large vascular malformation on the left thigh with splitting of overlying skin (***Fig.* 1**). There was no other anomaly or dysmorphism. Investigations were suggestive of anemia (hemoglobin 7.5 g/dL) and severe thrombocytopenia (platelet count: $8\times10^9$/L). Total leucocyte count (13.3 $\times10^9$/L), serum electrolytes and serum calcium were normal; C-reactive protein was negative.

She received intravenous antibiotics, packed red cells, platelet concentrates and fresh frozen plasma. Intravenous furosemide was administered in view of congestive heart failure. By 12 hours of life, infant's condition remained critical, and she required mechanical ventilation. *X*-ray chest showed cardiomegaly (CT ratio 0.7), and liver was palpable (3 cm below costal margin). Congestive cardiac failure further worsened, requiring digitalization and inotrope support to maintain perfusion. Echocardio-graphy revealed dilated right atrium, right ventricle and major pulmonary artery, mild to moderate TR, small ASD, and PDA. Doppler ultrasound of the enlarged left lower limb showed large arterio-venous malformation from left inguinal region including labia, extending upto foot. Computed tomography (CT) angiography of the affected left thigh revealed tufts of vessels in subcutaneous planes arising from superficial femoral artery, with enhancing soft tissue component and large draining veins sugges-tive of large arterio-venous malformation (***Fig.* 2**). Urinary bladder was also grossly distended with bilateral hydronephrosis. Interventional radiologist suggested angio-embolization of the affected arterio-venous malformations, but the parents refused the procedure. Steroids were not administered because of fulminant sepsis. Child's condition worsened and coagulation profile deteriorated further. She developed pulmonary hemorrhage and died on day four of life due to severe

**FIG.1** *Large arterio-venous malformation affecting left thigh.*

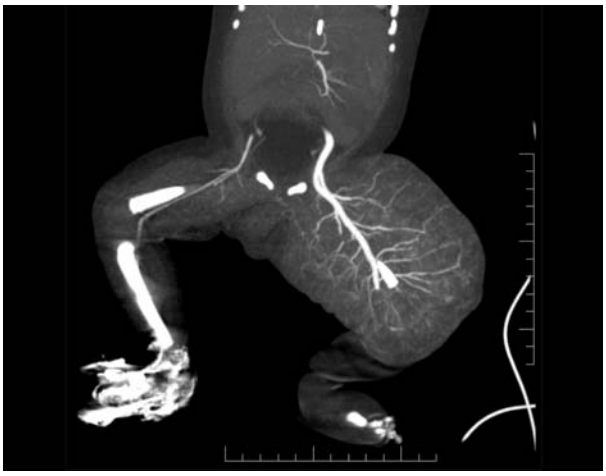

**FIG. 2** *Coronal image of CT angiography showing large soft tissue swelling of left thigh and multiple arterial feeders along with dilated femoral artery.*

bleeding manifestations, overwhelming sepsis and cardiac failure.

## DISCUSSION

Kasabach-Merritt syndrome, a potentially life-threatening coagulopathy characterized by enlarging hemangioma with severe thrombocytopenia [6], can complicate KTWS. Analysis of prenatal presentations and perinatal outcomes of KTWS suggested that the involvement of fetal thigh is rare [7]. Bilateral hydronephrosis is very rarely reported with KTWS [4]. Although urine output and renal function tests were normal, further workup of renal system involvement could not be done. Mortality rate in the neonatal period is high with complications of Kasabach-Merritt syndrome observed in about one-third of cases [7]. The causes of neonatal mortality in KTWS includes Kasabach-Merritt syndrome, cardiac failure, sepsis, and prematurity [7,8]. Treatment options include steroids, compression, pulsed-dye laser treatment, surgical reduction or embolization. Surgical therapy is often precluded by the risk of severe hemorrhage and high mortality in extensive lesions [7].

*Contributors*: LB: Patient management. SB: manuscript writing.; KK: investigation support and the care of the patient. *Funding*: None; *Competing interest*: None stated.

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