was followed by more similar reports of children with COVID-19 and clinical features that are similar to those of toxic shock syndrome and atypical Kawasaki disease and laboratory findings associated with increased inflammation [3-5]. Royal College of Paediatrics and Child Health (RCPCH) labeled this new inflammatory entity as Paediatric multisystem inflam-matory syndrome temporally associated with COVID-19 [8]. Case definitions include persistent fever, inflammation and evidence of single or multi-organ dysfunction after exclusion of other microbial causes. The case mentioned showed a rise of CRP without any neutrophilia, lymphopenia or organ dysfunction.

Our case was very similar to that described by Jones, *et al.* [2] but that girl had persistent tachycardia and most of the clinical features of KD with normal echocardiography. Riphagen, *et al*. [3] reported a case series of 8 children (only 3 tested COVID-19 positive) needing intensive care support with a hyper-inflammatory shock. One child died after a massive cerebral infarction. All of them had features mentioned in RCPCH guidelines with minimal respiratory symptoms [3]. These children and the two infants with Kawasaki disease most likely had a similar pathogenesis with varied consequences, which needs further research to define it.

India is still in the early stage of this pandemic and has not yet had many children with severe COVID-19. This 4-month-old child presenting as typical Kawasaki disease represents a novel presentation among the very young population with COVID-19.

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# Portal Hypertension in a Case of Klippel Trenaunay Syndrome

Klippel Trenaunay syndrome (KTS) is a sporadic disorder that belongs to PIK3CA-related overgrowth spectrum of disorders. The diagnostic criteria for KTS comprise of presence of capillary malformation, venous with or without lymphatic malformation and limb overgrowth. Only 63% patients have all three clinical manifestations [1]. Here we describe a case of KTS presenting as mixed venolymphatic malformation with complication in the form of portal hypertension due to dysplastic portomesentric veins.

We report an 11-year-old girl who presented with swelling of the right gluteal region noticed since birth. This swelling slowly progressed to involve the whole of the right lower limb accompanied by dilated veins over lateral aspect of the ankle. At six years of age, she developed clusters of small vesicles with warty appearance in the affected lower limb which ruptured spontaneously discharging serous fluid. Baseline hemogram, kidney and liver function tests were normal. Skin wedge biopsy performed was consistent with lymphangioma circumscriptum. Ultrasound doppler of gluteal region revealed dilated anechoic tortuous channels showing no flow within suggestive of lymphangioma. Ultrasonography of abdomen was normal. MR angiography of limb revealed extensive soft tissue hypertrophy involving right gluteal, thigh and upper leg

with dilated vascular channels in posterior compartment of leg, suggestive of venolymphatic malformation.

At eight years of age, she developed severe pallor with anasarca and bleeding per rectum. On examination, ascites and splenomegaly were evident. Investigations revealed hemoglobin of 3.9 g/dL, total leucocyte count of 4200/$mm^3$ and platelet count of 50000/µL. She was transfused with packed RBC and platelets. Liver and kidney function tests were normal. Ultrasound of the liver unveiled portal vein thrombosis with periportal collaterals and massive splenomegaly. Upper gastrointestinal endoscopy was normal. External hemorrhoids was identified by proctoscopy. CT angiography of the abdomen revealed dilated main portal vein with fusiform aneurysmal dilatation of left branch of portal vein and superior mesenteric vein with foci of thrombus within. Multiple collaterals were seen at porta (***Web Fig*.1a**), pericholecystic region, head and body of pancreas with dilatation of left gonadal vein and splenomegaly. There were abnormal soft tissue and vascular channels within the subcutaneous and intramuscular plane of right gluteal region with grossly dilated right internal iliac veins and presence of abnormal draining vein arising from soft tissue (***Web Fig*. 1b**)

At 11 years of age, she again presented with pancytopenia. On examination, there was autoamputation of terminal phalanx of 4th toe of the affected limb with surrounding skin necrosis. Limb deformity was also present with previous existing features (***Web Fig* 1c**). The patient was transfused and was put on prophylaxis for portal hypertension and referred to vascular surgery department for further intervention.

KTS is a low flow vascular malformation in an overgrown limb. Somatic heterozygous gain of function mutations in a mosaic pattern in the *PIK3CA* gene was recently identified in patients with KTS [2]. These somatic mosaic mutations affect only a portion of the body and since these mutations occur as a post zygotic event, they are not transmitted to progeny.

Absence of central nervous findings, truncal fatty-vascular growth, paraspinal fast-flow lesions and skeletal abnormalities distinguishes KTS from other *PIK3CA* related disorders. Amongst other overgrowth disorders, absence of arterial involvement distinguishes it from Parker Weber syndrome. Lack of nevi differentiates it from Proteus syndrome. Our patient exhibited two of the cardinal features of KTS. Portal hypertension could have resulted from development of thrombosis in ectatic portomesentric veins. External haemorrhoids and splenomegaly could be a complication of the primary disease or portal hypertension.

Patients with large and complex vascular malformations in KTS generally tend to have a higher risk for thromboembolic disease. Visceral involvement, as a consequence can result in significant morbidity and mortality [3]. Lymphatic malformations are also prone to rupture and recurrent infections. In KTS patients, magnetic resonance imaging of the abdomen, pelvis and lower extremities should therefore be performed in the early infantile period or at the time of initial presentation. Colour and spectral Doppler ultrasound can also be used to image the vascular malformations when available. In contrast to other *PIK3CA* disorders, occurrence of malignancies are less likely.

Symptom specific management includes surgical debulking for complex lymphatic malformations, sclerotherapy for minor vascular malformations, pulsed dye laser for capillary lesions and orthopedic interventions for deformities. Sirolimus has been found to have a promising role in complex slow flow vascular malformations [4]. PI3K and mTOR inhibitors are also being actively investigated [5]. Prophylactic anticoagulant therapy can be considered in patients with complex vascular malformations prior to radiological or surgical procedure.

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