Portomesenteric thrombosis and hypertension in Klippel–Trenaunay syndrome

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To The Editor,

In the report entitled "Preoperative splenic artery embolization in Klippel–Trenaunay syndrome (KTS) with massive splenomegaly," Zhang *et al*. described a 29-year-old woman who was diagnosed with KTS and hypersplenism who underwent splenic artery embolization followed by splenectomy.[1] We compliment the authors for their successful management of hypersplenism. Nevertheless, we have a few comments on the etiology of this complication and the diagnosis of KTS.

1. The authors reported that the patient had "noncirrhotic portal hypertension of unknown etiology, complicated by portal vein thrombosis." The etiology of portal hypertension in KTS is likely related to portomesenteric venous thrombosis. Kulungowski *et al*. reported the association between portomesenteric venous thrombosis and anomalous ectatic mesenteric veins in eight patients with portal hypertension in six patients.[2] Four of these patients had typical KTS. The treatment recommendation is prophylactic surgical proximal ligation of the ectatic mesenteric vein
2. The authors described "lymphedema of the lower extremities and cutaneous port-wine stain." KTS is characterized by fatty overgrowth of a limb, capillary malformation (or port-wine stain), venous malformation and lymphatic malformation.[3] Nevertheless, the usual lymphatic spectrum in KTS includes macrocystic and microcystic lymphatic lesions as well as cutaneous vesicles, rather than a typical lymphedema
3. Splenic arteriovenous malformations do not typically occur in patients with KTS. Splenic involvement in KTS is typically limited to lymphatic malformation with or without significant splenomegaly. In addition, portal hypertension may cause splenomegaly
4. In discussing the etiology of coagulopathy in KTS, the authors stated that "thrombocytopenia, in the context of KTS is due to Kasabach–Merritt syndrome secondary to the peripheral vascular malformations." Thrombocytopenic coagulopathy, more accurately called "Kasabach–Merritt phenomenon" rather than a true syndrome, specifically occurs in kaposiform hemangioendothelioma[4] and is not a known feature of KTS.

Though the genetic etiology of KTS has recently been found to be related to somatic mosaic mutations in PIK3CA,[5] the reported wide variety of related and unrelated clinical features created a real diagnostic confusion about this uncommon disorder.

REFERENCES

1. Zhang Z, Dasika NL, Englesbe MJ, Owens SR, Vellody R, Novelli PM, *et al*. Preoperative splenic artery embolization in Klippel-Trenaunay syndrome with massive splenomegaly: A case report. Avicenna J Med 2014;4:40-3.
2. Kulungowski AM, Fox VL, Burrows PE, Alomari AI, Fishman SJ. Portomesenteric venous thrombosis associated with rectal venous malformations. J Pediatr Surg 2010;45:1221-7.
3. Christison-Lagay ER, Fishman SJ. Vascular anomalies. Surg Clin North Am 2006;86:393-425.
4. Sarkar M, Mulliken JB, Kozakewich HP, Robertson RL, Burrows PE. Thrombocytopenic coagulopathy (Kasabach-Merritt phenomenon) is associated with Kaposiform hemangioendothelioma and not with common infantile hemangioma. Plast Reconstr Surg 1997;100:1377-86.
5. Kurek KC, Luks VL, Ayturk UM, Alomari AI, Fishman SJ, Spencer SA, *et al*. Somatic mosaic activating mutations in PIK3CA cause CLOVES syndrome. Am J Hum Genet 2012;90:1108-15.

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