Chronic lower extremity wounds in a patient with Klippel Trenaunay syndrome

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ABSTRACT

Klippel-Trenaunay syndrome is a rare disorder consisting of the triad of vascular and/or lymphatic malformations, capillary malformations, and soft tissue or bony hypertrophy. Symptom control is the mainstay of treatment for these patients, with many of the symptoms never fully being relieved. In this case report, we present the case of a 46-year-old man with chronic lower extremity ulcerations unresponsive to wound care therapy. Owing to the chronic nature of his wounds and associated pain, reconstruction of his iliac vein was performed using polytetrafluoroethylene graft and an arteriovenous fistula. (J Vasc Surg Cases and Innovative Techniques 2019;5:45-8.)

Keywords: Klippel-Trenaunay syndrome; Iliac venous system; Varicose vein; Venous ulcer; Venous insufficiency

CASE REPORT

A 46-year-old man was recently diagnosed with Klippel-Trenaunay syndrome (KTS). He is the only member of his family with this diagnosis and has never undergone any surgical correction related to KTS. A diagnosis of KTS was made when a venous doppler ultrasound and computed tomography scans were performed for the evaluation of extremity swelling. Absence of the proximal right femoral and iliac venous systems were noted (Fig 1). His history includes chronic lower extremity ulcerations and edema present for more than 2 years, and port wine staining present since birth. He denied any trauma to the extremity and had been managed at an outpatient wound care facility with local wound care and compressive therapy. There was minimal improvement to the ulcerations with worsening pain along the medial malleolus.

He presented to our outpatient office with complaints of pain along the gaiter aspect of his right lower extremity with mild drainage of the wounds. On examination, he demonstrated port wine staining throughout the majority of his back and flank with extension down his right buttock, thigh, leg, and onto his forefoot. There was size discrepancy between the lower extremities with the affected side being 2 cm longer and thigh circumference 25% greater on the right side.

Varicosities were present over his lower abdomen, anterior thigh, and leg. The patient's vascular examination demonstrated ulcers along the gaiter distribution of his right lower extremity more consistent with venous stasis disease than an infectious etiology. The largest ulcer measured 5 × 3 cm over the medial malleolus and was without purulence and minimal serous drainage. Pretreatment, the patient was classified as $C_6E_cA_{s,p,d}P_o$ and post-treatment had improved to $C_5E_cA_sP_r$ with reflux secondary to prolonged obstructive dilation. Pitting edema was also appreciated and did obscure his pulses, although they were present. He denied hemorrhoids or hematochezia suggestive of esophageal varicosities. Laboratory results were all within the normal range (Fig 2).

Owing to the worsening pain and failed medical management, including compressive therapy and wound care, the patient was offered surgical reconstruction in an effort to decompress his venous system. A right common femoral vein to inferior vena cava venous reconstruction and creation of a superficial femoral artery to common femoral vein arteriovenous fistula was created using polytetrafluoroethylene (PTFE) grafts. The vena cava was exposed with a right lower quadrant paramedian incision and in a retroperitoneal fashion. Next, a groin incision was made and carried down where a blind ending common femoral vein was identified to be ectatic at its most proximal end. Once exposure was complete, a tunnel was fashioned under the inguinal ligament. A side-of-vein to end-of-graft distal anastomosis was created with a 12-mm ringed PTFE graft, and subsequently anastomosed to the inferior vena cava in a similar fashion. An arteriovenous fistula was also created with a 6-mm PTFE graft between the superficial femoral artery to a branch off the femoral vein (Fig 3).

Local wound care with compressive therapy was continued while in-house with marked improvement of his swelling. He was discharged on rivaroxaban for graft patency. The patient has been seen for follow-up 7 months after the index procedure and has complete resolution of the pain, swelling, and ulcerations (Fig 4). The patient has been adequately informed and consented to involvement in this case report.

DISCUSSION

KTS is a rare congenital disorder first described by Maurice Klippel and Paul Trenaunay in the early 1900s and originally termed naevus vasculosus

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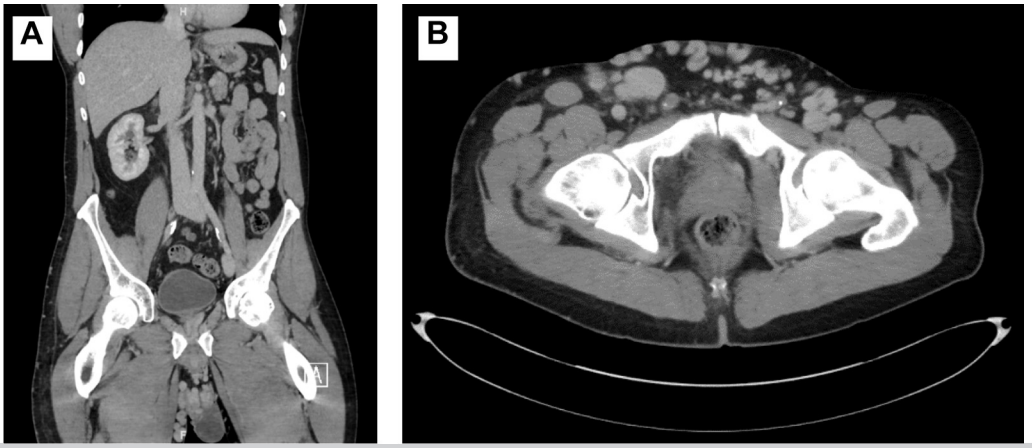

Fig 1. Computed tomography scan. **A,** Absence of right iliac vein with blind ending pouch at the right inferior border of the inferior vena cava. **B,** Abdominal wall varicosities.

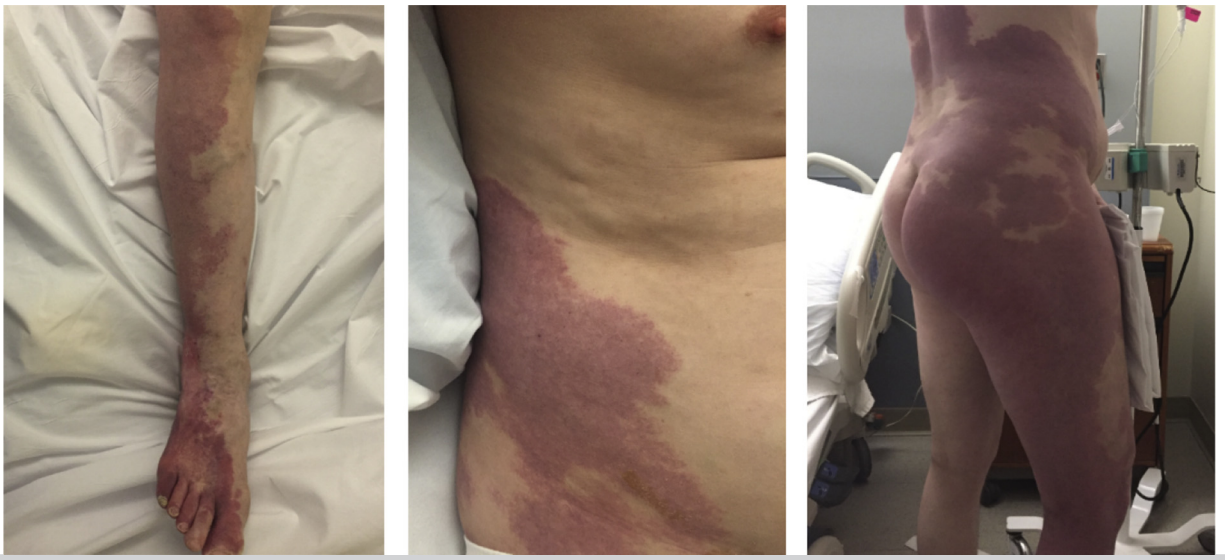

Fig 2. Varicosities and port wine stain along right abdomen and buttock and extending down the leg.

osteohypertrophicus.[1] Currently, the International Society for the Study of Vascular Anomalies describes KTS as the constellation of varicosities and/or venous malformations, capillary malformation, and boney and/or soft tissue malformation of the limbs.[2,3] This should not be confused with Parkes Weber syndrome, which is defined by the triad of capillary malformations, limb disparity, and arteriovenous malformation.[4]

Although most cases of KTS are sporadic, there have been a number of familial cases reported. In most cases, a gain-of-function mutation in the catalytic subunit of the phosphatidylinositol 3-kinase gene is implicated in the pathogenesis, ultimately leading to proliferation and angiogenesis.[5] It has been determined that a small percentage of patients with KTS demonstrate a persistent marginal vein that fails to regress and, if present, represents the major outflow for the leg. This additional malformation was not present on computed tomography scan and duplex ultrasound results of our patient, resulting in extensive tortuous venous collaterals in the subcutaneous tissues of the right inguinal region and lower anterior abdominal wall.

The treatment options for KTS are limited and mainly supportive.[6,7] Venous and lymphatic insufficiency leading to chronic edema is managed with compressive therapy and wound care, limb discrepancy is typically managed with shoe lifts and rarely managed surgically, and capillary malformations are treated with laser and/or ablative therapy if symptomatic.[7-9] To date, there have been no reconstructive surgical attempts for the venous insufficiency portion of the triad.[10-12]

Our patient began exhibiting long-standing issues secondary to venous stasis disease of his right lower extremity. He was also noted to have mild venous

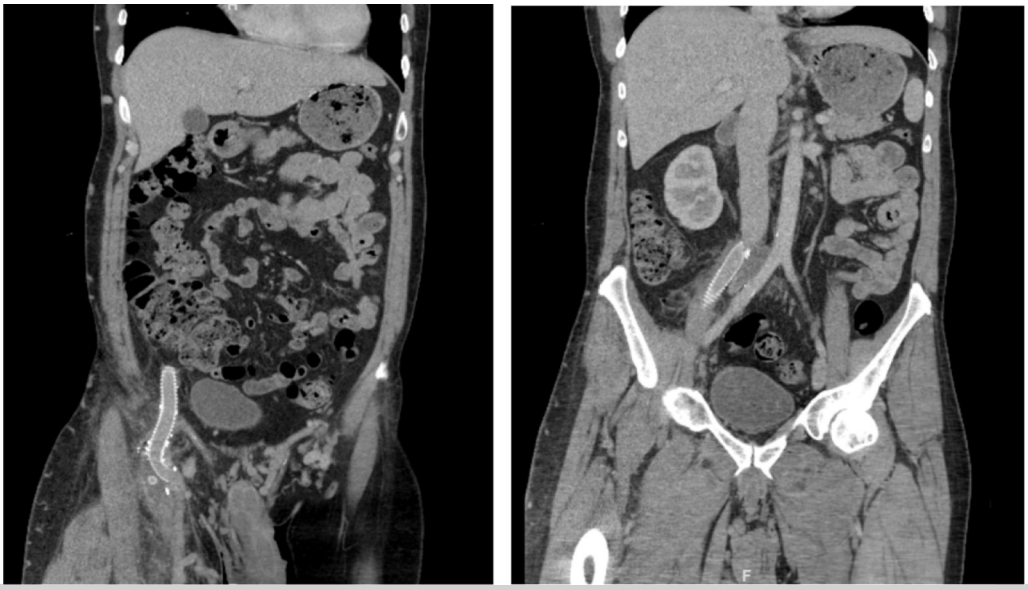

Fig 3. A right common femoral vein to inferior vena cava reconstruction was performed using a polytetrafluoroethylene (PTFE) graft.

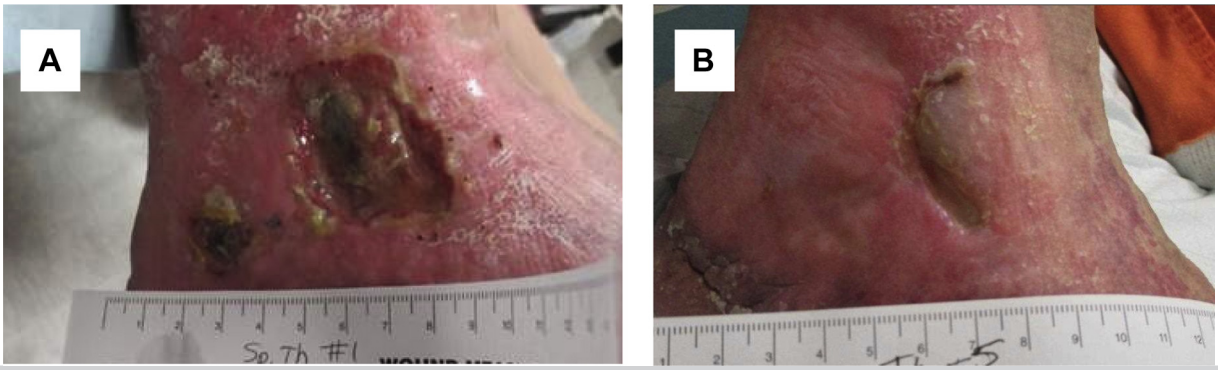

Fig 4. A, Medial malleolus ulceration preoperatively. **B,** The same wound at 6 weeks postoperative.

disease of his left lower extremity and small ulcers that were amenable to local wound care. Medical management of his right lower extremity consisted initially of leg elevation and compressive therapy. Owing to worsening of the ulcerations, this proceeded to wound debridement with Hydrogel type dressing, which was performed for 13 months. Owing to the failure of medical therapy of the right lower extremity, the patient opted to proceed with surgical reconstruction of his missing iliac system in an effort to create venous continuity and drainage of his right lower extremity. A discussion was held concerning a femoral crossover bypass for decompression; however, owing to his preexisting venous disease, we did not want to worsen the drainage of the left lower extremity. A larger 12-mm graft was chosen owing to a better size match at the proximal anastomosis. Initially, the distal anastomosis to the common femoral vein in an end-to-end fashion was considered; however, the distal end of the femoral vein was ectatic and so a side-of-vein to end-of-graft anastomosis was performed. PTFE was chosen owing to its greater rigidity as compared with a spiral vein graft or other vein graft. Prophylactic measures were undertaken to prevent graft thrombosis by creating an arteriovenous fistula to increase flow rates and prevent stasis in a system that was radiographically and physically demonstrated to be ectatic. Postoperatively, the patient did not demonstrate any signs of congestive heart failure.

CONCLUSIONS

Although venous congestion is a known entity of KTS, the consequence of nonhealing ulcerations can become chronic and debilitating, as seen in our patient. When traditional medical management fails to relieve symptoms, and if the venous anatomy is favorable, a reconstruction of the outflow is a plausible treatment modality in such patients, so long as other operative modalities have also been exhausted.

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