Use of Ultrasound and Digital Subtraction Angiography for Arteriovenous Malformations in a Patient with Klippel-Trenaunay-Weber Syndrome

Zhimei Cheng, BS^{a,b}, Shuai Zhang, PhD^a, Ping Wang, BS^a, Hong Zhu, BS^a, Zhijing Wei, BS^a, Shi Zhou, BS^{a,*}

^a School of Medical Imaging, Guizhou Medical University, Guiyang, Guizhou, China; ^b Department of Ultrasound, Xingyi People's Hospital, Xing Yi, Guizhou, China

Received February 21, 2020; revision received April 4, 2020; accepted May 15, 2020

Abstract: We report a case of a 21-year-old woman with Klippel-Trenaunay-Weber syndrome (KTWS), a rare congenital syndrome involving capillary malformations, arteriovenous malformations, and soft tissue or bone hypertrophy. For this patient, ultrasound and digital subtraction angiography (DSA) were used to evaluate arteriovenous malformations. Arteriovenous shunts could be identified with both ultrasound and DSA. DSA was able to provide an overall view of the malformed blood vessels, whereas ultrasound was effective in showing the condition of the blood vessel walls and in assessing blood flow velocity. This case suggests that multiple imaging studies may be needed for the diagnosis and treatment of KTWS disease in these patients.

Key words: Klippel-Trenaunay-Weber syndrome (KTWS); Ultrasound; Arteriovenous malformations; Digital subtraction angiography

Advanced Ultrasound in Diagnosis and Therapy 2020;04:243-247

DOI: 10.37015/AUDT.2020.200009

Klippel-Trenaunay-Weber syndrome (KTWS) is a rare congenital disorder that is characterized by a triad of venous malformations (arteriovenous fistulas), capillary malformations (hemangiomas), and soft tissue or bone hypertrophy of the limb. Although Doppler ultrasound is a valuable method for identifying abnormal vessels and assessing blood flow, few reports have described ultrasound findings for arteriovenous malformations in patients with KTWS [1,2], with most reports focusing on conventional angiography [3-5], computed tomography (CT) [2,6], and magnetic resonance imaging (MRI) [3,6-10]. Here, we mainly describe the use of ultrasound and digital subtraction angiography (DSA) to assess arteriovenous malformations in a patient with KTWS.

Case report

A 21-year-old woman presented with a 5-month

history of ulcers on her right dorsum pedis. The patient stated that she had been born with port-wine stains on the right buttock, and these port-wine stains had gradually increased in size. When she was 11 years old, she had noticed a disproportionate increase in the size of her right lower limb. She complained of joint pain and claudication when she was 11 years old. The patient had no history of trauma and no family history of similar vascular anomalies. On physical examination, pulsation and thrill were detectable, and the cutaneous temperature in the right hip was found to be higher than in the left hip. The right lower extremity was observed to be slightly longer than the left lower extremity (Fig. 1C). Laboratory results were within standard limits.

No substantial abnormalities were noted on electrocardiogram, cardiac ultrasound, or radiographs of the thorax. Radiographs of the right lower limb demonstrated soft tissue hypertrophy, particularly in the right hip. Soft tissue overgrowth of the right lower limb

* Corresponding Author: School of Medical Imaging, Guizhou Medical University, 28 Guiyi Road, Guiyang, Guizhou 550004, China
e-mail: shizhou0128@126.com

and numerous tortuous vessels were observed on MRI, and a mesh-work of vascular malformations in the right lower limb was observed on CT. DSA revealed multiple abnormal vessels along the lateral thigh and draining into the femoral vein in the right thigh (Fig. 2A). When the focus was placed on an abnormal vessel, the

vessel was seen to separate from the trunk of the lower extremity artery and extend subcutaneously (Fig. 2B). The abnormal vessel became large and tortuous in the subcutaneous tissues, branched off, and finally drained into the veins (Fig. 2C, D).

Figure 1 The superficial femoral artery (A) and anterior tibial artery (B) of the right lower limb had blood flow that was high velocity, biphasic, and unidirectional, with low resistance. The right lower limb was slightly longer than the left lower limb (C). Segments of the popliteal artery demonstrated aneurysmal dilatations (D).

Figure 2 Digital subtraction angiography (DSA) demonstrated arteriovenous fistulas in the right lower limb (A). The vessel separated from the trunk of the lower extremity artery and extended subcutaneously (B). The abnormal vessel became large and tortuous in subcutaneous tissues (C), branching off and finally draining into the veins (D).

The internal diameters of the arteries in the right lower limb (including the common femoral, superficial femoral, deep femoral, popliteal, anterior tibial, posterior tibial, and peroneal arteries) were measured on conventional grayscale sonography; when these values were compared with those in the left lower limb, they were found to be significantly higher (paired sample *t* test, $P < 0.001$ Table 1). Segments of some of these arteries (including the popliteal, anterior tibial, and posterior tibial arteries) demonstrated aneurysmal dilatations (Fig. 1D). The blood flow velocity of all vessels was higher in the right lower limb than in the left lower limb (Table 2), with significant differences seen in the peak systolic velocity (PSV) (paired sample *t* test, $P < 0.004$). From the common iliac artery to the dorsal pedal artery of the right lower limb, the blood flow was high velocity, biphasic, and unidirectional, with low resistance (Fig. 1A, B). Color Doppler ultrasound detected multiple fast-flowing arteriovenous shunts or fistulas in the right lower limb, and ultrasound was used to continuously track the abnormal vessels from the

inflow segment (Fig. 3A). In the abnormal vessels, the blood flow velocity and the resistance index gradually decreased (Table 3). The blood flow was biphasic and unidirectional, with low resistance (Fig. 3C, D). Multiple dilated tortuous vascular malformations (Fig. 3B) were

Table 1 Internal diameters of arteries in the lower limbs (cm)

Artery	Affected limb (right)	Contralateral limb (left)
Common femoral (distal)	1.02	0.63
Superficial femoral (initial segment)	0.84	0.49
Deep femoral (initial segment)	0.79	0.46
Superficial femoral (middle segment)	0.58	0.45
Popliteal (initial segment)	0.77	0.58
Anterior tibial (middle segment)	0.55	0.22
Posterior tibial (middle segment)	0.48	0.21
Peroneal (middle segment)	0.5	0.14

observed in the right buttock area, the lateral thigh, and the right crus. The venous blood flow velocity (including the common femoral, superficial femoral, deep femoral,

popliteal, and great saphenous veins) was significantly higher in the right lower limb than in the left lower limb ($P < 0.005$; Table 4).

Table 2 Doppler ultrasound, blood flow parameters in the lower limbs (velocity, cm/s)

Artery	Affected limb (right)				Contralateral limb (left)		
	PSV	EDV	RI	Waveform	PSV	EDV	Waveform
Common femoral	334	161	0.52	2-way	146	−32	3 directions
Superficial femoral	191	59	0.84	2-way	123	−22	3 directions
Deep femoral	267	65	0.78	2-way	142	−34	3 directions
Popliteal	250	100	0.6	2-way	75	−11	3 directions
Anterior tibial	169	76	0.55	2-way	63	−8	3 directions
Posterior tibial	183	96	0.48	2-way	69	−10	3 directions
Peroneal	57	28	0.5	2-way	37	−5	3 directions

PSV, peak systolic velocity; EDV, end-diastolic velocity; RI, resistance index

Table 3 Blood flow parameters for vascular malformation (from the inflow to the outflow)

Peak systolic velocity (cm/s)	End-diastolic velocity (cm/s)	Resistance index
186	81.9	0.56
144	54.3	0.61
100	58.8	0.42
34.3	24.2	0.29

Table 4 Changes in venous blood flow velocity in the lower limbs

Vein	Maximum blood flow velocity (cm/s)	
	Affected limb (right)	Contralateral limb (left)
Common femoral	195	36
Superficial femoral	116	27
Deep femoral	120	28
Great saphenous	67	6
Popliteal	87	11

Based on the clinical findings (port-wine stains and limb hypertrophy) and imaging results (fast-flowing arteriovenous malformations), KTWS was diagnosed. The pros and cons of various management strategies were weighed, and treatment with ethanol combined with coil embolization was chosen. Coils were used to occlude the high-flow vessels, with ethanol used to directly damage

the abnormal vessels. The coils successfully embolized malformed blood vessels in the hip (Fig. 4A, 4B). However, ethanol did not play a role in sclerosing the vessel walls; no change was seen in blood flow velocity on ultrasound (Fig. 4C, 4D). As part of treatment, the patient was also advised to wear compression socks. During follow-up, the patient's symptoms and signs in the right hip were relieved, and she was satisfied with the outcome. The ulcers on the right dorsum pedis were treated successfully with antibiotics.

Discussion

Klippel-Trenaunay syndrome (KTS), a rare congenital disorder involving capillary, venous, and lymphatic malformations and associated with limb overgrowth, was first described in 1900 by Klippel and Trenaunay. Subsequently, Weber added the presence of arteriovenous fistulae to the syndrome, and the syndrome is now commonly described as KTWS [11]. Clinicians must be able to differentiate KTWS from other conditions that cause vascular malformations, as KTWS can be life-threatening if intervention is delayed. Early diagnosis and treatment are therefore essential.

Here we report a case of KTWS presenting with the classic triad of capillary malformations (port-wine stain), arteriovenous malformations, and soft tissue or bone hypertrophy. This case illustrates the usefulness of Doppler imaging in the identification of abnormal vessels and the assessment of blood flow. In this case, ultrasound demonstrated that the blood flow velocity of vessels in the affected (right lower) limb was significantly higher

than in the contralateral limb, with many abnormal vessels identified in the right lower extremity. DSA was able to identify arteriovenous malformations in the right lower extremity, and conventional angiography demonstrated many arteriovenous tunnels in the affected

limb, especially in the right buttock and the lateral thigh area. Compared with ultrasound, DSA was better able to demonstrate the entire structure and complete map of arteriovenous tunnels in this case.

Figure 3 Color Doppler ultrasound demonstrated the primary vessel of the arteriovenous fistula (A). The abnormal vessel became large and tortuous in the subcutaneous tissues (B). The vascular malformations demonstrated gradual decreases in blood flow velocity and resistance index (C, D).

Figure 4 Coils were used to embolize the malformed blood vessels in the hip (A, B). After ethanol was injected into the tortuous dilated abnormal blood vessels, no change was seen in blood flow velocity on ultrasound (C, D).

For patients with KTWS, a multidisciplinary approach to treatment is often helpful. The management strategy will vary based on the size, number, and location of arteriovenous malformations. Previous studies have assessed the use of compression garments, feeding artery ligation, embolization, surgical resection, stent graft placement, and limb amputation for the treatment of KTWS [12–14]. For this patient, we elected to use ethanol combined with coil embolization. However, ethanol was not found to have much effect in this case, perhaps because of the high blood flow velocity in the abnormal vessels.

Through this case, we found that ultrasound and DSA have advantages and disadvantages for imaging patients with KTWS. DSA can show the overall view of malformed blood vessels, however, cannot effectively evaluate the malformed blood vessel walls or blood flow velocity. Ultrasound, on the other hand, can clearly show the condition of the blood vessel walls and can be used to assess the blood flow velocity; however, for deep lesions, the display capability of ultrasound is limited. Therefore, multiple imaging studies may be needed for the diagnosis and treatment of disease in these patients.

In conclusion, early diagnosis and treatment of KTWS are of great importance in clinical setting. This case highlights the usefulness of color Doppler ultrasound

along with DSA in distinguishing between KTWS and other conditions causing vascular malformations.

Acknowledgments

This study was supported by funds from the Guizhou Province Science and Technology Projects (Guizhou foundation [2018] 1129). We would like to acknowledge members of the Interventional Radiology, Affiliated Hospital of Guizhou Medical University for support and technical assistance.

Conflict of Interest

The authors declare that they have no conflict of interest.

References

- [1] Pandey P, Ralli M, Agarwal S, Agarwall R. Klippel-Trenaunay Weber syndrome. *Med J DY Patil Vidyapeeth* 2019; 12: 356–358.
- [2] Volz KR, Kanner CD, Evans J, Evans KD. Klippel-Trenaunay Syndrome: Need for Careful Clinical Classification. *J Ultrasound Med* 2016; 35: 2057–2065.
- [3] Turkmen M, Kavukçu S, Çakmakci H, Soylu A, Aktan S, Çağan Y. A girl of Klippel-Trenaunay Weber syndrome coexistence of recurrent bloody vaginal discharge. *Int Urol Nephrol* 2010; 42: 575–578.
- [4] Ziyeh S, Spreer J, Rossler J, Strecker R, Hochmuth A, Schumacher M, et al. Parkes Weber or Klippel-Trenaunay syndrome? Non-invasive diagnosis with MR projection angiography. *Eur Radiol* 2004; 14: 2025–2029.

- [5] Iizuka Y, Suzuki M, Komura S, Takada T, Shimoji K. Conus medullaris spinal arteriovenous malformation in a patient with klippel-trenaunay-weber syndrome. A case report and review of the literature. *Interv Neuroradiol* 2008; 14: 185-190.
- [6] Khanna V, Singh G, Kumar S, Singh S. A neglected case of Klippel Trenaunay Weber syndrome. *J Orthop Traumatol and Rehabil* 2015; 8: 39-41.
- [7] Opdenakker O, Renson T, Walle JV. Vesical Hemangioma in a Patient with Klippel-Trenaunay-Weber Syndrome. *J Pediatr* 2019; 208: 293-293. e2.
- [8] Onoda S, Komagoe S. Lymphaticovenular anastomosis for Klippel-Trenaunay-Weber syndrome. *Int J Surg Case Rep* 2019; 58: 67-69.
- [9] Chenbhanich J, Leelayuwatanakul N, Phowthongkum P. Klippel-Trenaunay-Weber syndrome as a cause of chronic thromboembolic pulmonary hypertension. *BMJ Case Rep* 2018; 2018.
- [10] Kharat AT, Bhargava R, Bakshi V, Goyal A. Klippel-Trenaunay syndrome: A case report with radiological review. *Med J DY Patil Univ* 2016; 9: 522-526.
- [11] Viljoen DL. Klippel-Trenaunay-Weber syndrome (Angio-osteohypertrophy syndrome). *J Med Genet* 1988; 25: 250-252.
- [12] Girón-Vallejo Ó, López-Gutiérrez JC, Fernández-Pineda I. Diagnosis and treatment of Parkes Weber syndrome: a review of 10 consecutive patients. *Ann Vasc Surg* 2013; 27: 820-825.
- [13] Servelle M. Klippel and Trénaunay's syndrome. 768 operated cases. *Ann Surg* 1985; 201: 365-373.
- [14] Gloviczki P, Stanson AW, Stickler GB, Johnson CM, Toomey BJ, Meland NB, et al. Klippel-Trenaunay syndrome: The risks and benefits of vascular interventions. *Surgery* 1991; 110: 469-479.