

Bilateral Klippel–Trénaunay Syndrome in an Adolescent Male with Deep Vein Thrombosis Complication: A Case Report and Literature Review

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INTRODUCTION

Klippel–Trénaunay syndrome (KTS) represents one of the most complex entities in the fields of vascular medicine and diagnostic radiology. This rare congenital disorder is characterized by a triad of cutaneous capillary malformations, venous anomalies, and soft tissue or bone hypertrophy of the affected extremity. First identified by Maurice Klippel and Paul Trénaunay in 1900, the condition was initially considered a simple vascular malformation; however, modern research has revealed a much deeper genetic and molecular basis [1].

Terminologically, KTS is often associated with the term Capillary-Lymphatic-Venous Malformation (CLVM), reflecting the multimodal involvement of the low-flow vascular system [1]. The key distinguishing feature of KTS from other vascular syndromes, such as Parkes Weber syndrome (PWS), is its hemodynamic nature. KTS is a low-flow disorder,

meaning there is no significant direct connection between the arterial and venous systems that form a shunt or fistula [4]. This understanding is vital because management for high-flow disorders is radically different, and if incorrectly applied to a KTS patient, it can lead to severe iatrogenic complications.

In clinical practice, KTS typically appears sporadically with highly variable manifestations, ranging from cosmetically limited port-wine stains to massive extremity overgrowth causing permanent physical disability [6]. One of the greatest challenges in managing KTS patients is the high risk of thromboembolic complications. Due to venous stasis in malformed and dilated vessels, patients are prone to thrombus formation, which can develop into life-threatening deep vein thrombosis (DVT) and pulmonary embolism (PE) [9].

Advances in imaging technology, particularly Computed Tomography Angiography (CTA) and Magnetic Resonance Imaging (MRI), have revolutionized how radiologists evaluate this syndrome. These modalities allow three-dimensional visualization of the entire vascular tree, assessment of deep vein patency, and identification of persistent embryonic vessels, such as the Lateral Marginal Vein of Servelle [9]. Furthermore, the understanding of associated lymphatic involvement in KTS is increasing, with many patients exhibiting lymph node swelling or cystic lymphangioma formation requiring specific intervention [6].

This case report is compiled to document an unusual clinical presentation of bilateral KTS in an adolescent male. Through in-depth analysis of imaging findings and correlation with the latest literature, this report aims to provide the medical community with valuable insights into a comprehensive diagnostic approach and complication prevention strategies for this rare vascular syndrome.

CASE REPORT

The patient, MKA, a 15-year-old male, was referred to the Department of Radiology with a history of chronic swelling of both lower extremities. Anamnesis revealed that this abnormality had been noticed since birth, where the toes of both limbs appeared abnormally spaced, although no obvious swelling was evident at that time. Swelling became visually apparent from the age of one year and progressed with age.

The patient's clinical course was marked by several episodes of exacerbation of pain. The patient reported that during an attack, the lower limbs would appear red, feel hot to the touch, and the soft tissue would become hard. Six months before this examination, the patient experienced an episode of severe pain in the knees and feet that rendered him unable to walk

for two days. In addition to the leg symptoms, the patient had a history of transient lumps appearing in various body parts, although these lumps had spontaneously regressed.

At present, the patient complained of a new lump in the left axilla that appeared one week prior to hospital admission. This lump grew rapidly, initially the size of a marble and developing to the size of a ping-pong ball within seven days. Clinically, the lump had ill-defined borders, was non-tender on palpation, with no skin ulceration, and was not accompanied by systemic symptoms such as fever or malaise. The patient had received initial treatment with anticoagulants and used compression stockings to aid edema management. A history of other systemic diseases such as hypertension, diabetes mellitus, heart disease, and asthma was denied. The patient also had no history of smoking, drug allergies, or tuberculosis treatment.

Physical and Supporting Examination

On physical examination, massive and symmetrical soft tissue swelling was found in both lower extremities, extending from the femoral region to the feet. No abnormal pulsations or bruits, which would indicate a high-flow arteriovenous fistula, were detected. The left axillary lump was soft and cystic on palpation, without signs of acute inflammation.

To comprehensively evaluate the vascular status, CTA and CTV of the bilateral lower extremities were performed on October 6, 2025, using intravenous contrast. The vascular assessment results are summarized in the following tables:

Table 2.1 Results of CTA, Arterial Phase, Bilateral Lower Extremities

Arterial Segment	Right Artery Findings			Left Artery Findings			Interpretation
Common	Diameter	±5.50	mm;	Diameter	±6.36	mm;	Normal
Femoral Artery	Patent lumen;	No		Patent lumen;	No		
	malformation/lesion			malformation/lesion			
Superficial Femoral Artery	Diameter	±4.56	mm;	Diameter	±4.73	mm;	Normal with Collaterals
	Collateralization (+)			Collateralization (+)			
Profunda Femoris Artery	Diameter	±3.83	mm;	Diameter	±3.61	mm;	Normal with Collaterals
	Collateralization (+)			Collateralization (+)			

Popliteal Artery	Diameter ± 5.42 mm; Proximal-distal segment normal	Diameter ± 5.23 mm; Minimal stenosis 0.7%	Normal
Anterior Tibial Artery	Diameter ± 1.79 mm; Normal architecture	Diameter ± 1.71 mm; Normal architecture	Normal
Posterior Tibial Artery	Diameter ± 1.91 mm; Smooth flow	Diameter ± 1.61 mm; Smooth flow	Normal
Dorsalis Pedis Artery	Diameter ± 1.20 mm; No thrombus	Diameter ± 1.28 mm; No thrombus	Normal

In the arterial phase, no evidence of a macroscopic arteriovenous fistula or early venous filling—the pathognomonic sign of Parkes Weber syndrome—was found [4]. The collateralization seen in the superficial and deep femoral arteries indicates an increased vascular demand in the hypertrophic soft tissue, but the main arterial lumens remain within normal limits without significant stenosis.

Table 2.2 Results of CTV, Venous Phase, Bilateral Lower Extremities

Venous Segment	Right Vein Findings	Left Vein Findings	Conclusion
Common Iliac Vein	Diameter ± 1.50 cm; Dilatation (+); Significant	Diameter ± 1.24 cm; Dilatation (+)	Pelvic Venous Ectasia
External Iliac Vein	Diameter ± 1.50 cm; Dilatation (+)	Diameter ± 1.67 cm; Dilatation (+)	Pelvic Venous Ectasia
Great Saphenous Vein	Diameter ± 4.45 mm; Collateral (+)	Diameter ± 3.56 mm; Tortuosity (+)	Superficial Malformation
Femoral Vein	Diameter ± 9.57 mm; Thrombus (+); Collateral (+)	Diameter ± 6.23 mm; Collateral (+)	Right DVT
Popliteal Vein	Diameter ± 8.51 mm; Dilatation (+); Collateral (+)	Diameter ± 1.16 cm; Dilatation (+); Collateral (+)	Deep Vein Ectasia

Small	Diameter	1.52	mm;	Diameter	1.32	mm;	Normal
Saphenous	Normal			Normal			
Vein							

The CTV findings were crucial, demonstrating massive systemic venous dilatation and collateralization in the bilateral deep venous systems. The presence of a thrombus in the right femoral vein provides an objective basis for the patient's complaints of pain and redness, diagnosed as deep vein thrombosis (DVT). Additionally, extensive soft tissue swelling from the femur to the bilateral pedis was observed on the accompanying abdominal and skeletal scanning.

Based on all clinical and radiological features, the patient was diagnosed with Klippel–Trénaunay syndrome with bilateral manifestations and DVT complication of the right femoral vein [1]. The patient's management was continued with oral anticoagulant therapy and conservative management using compression stockings to control venous insufficiency and prevent thrombus extension.

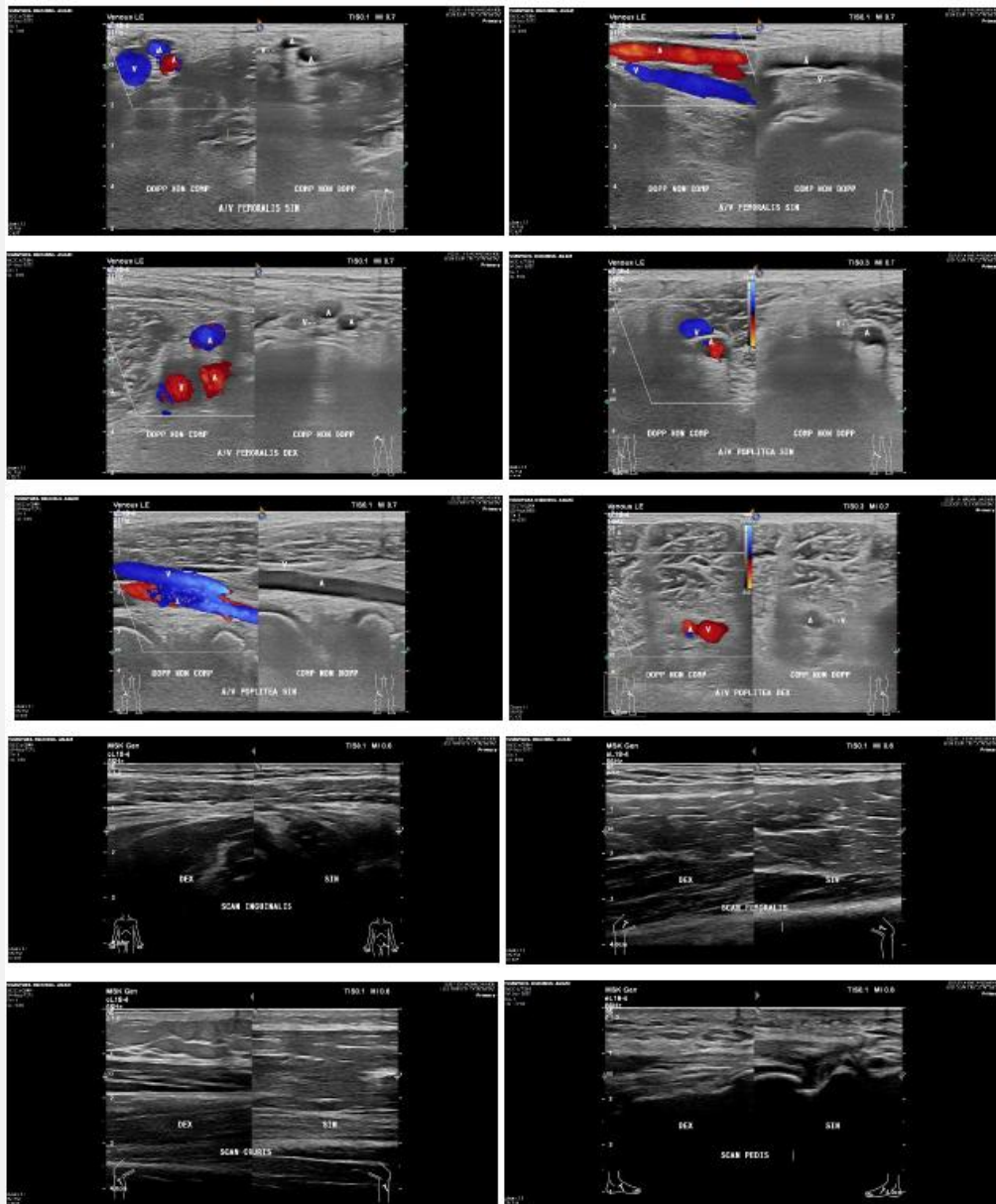


Figure 1. Ultrasound Results



Figure 2. Radiological Results of the Foot (AP/Lateral)

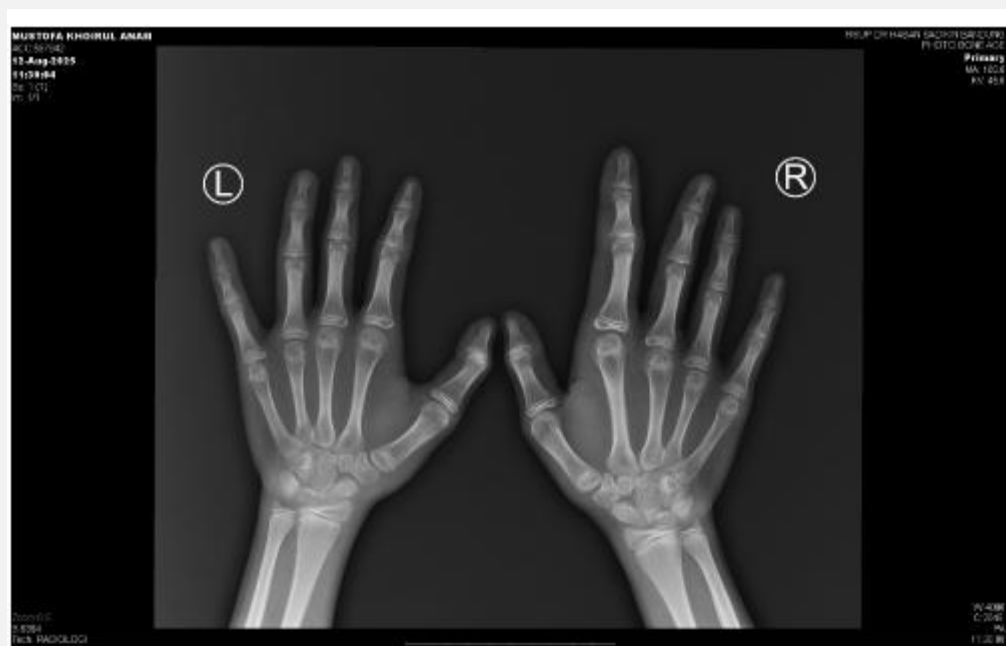


Figure 3. Radiological Results for Bone Age Assessment

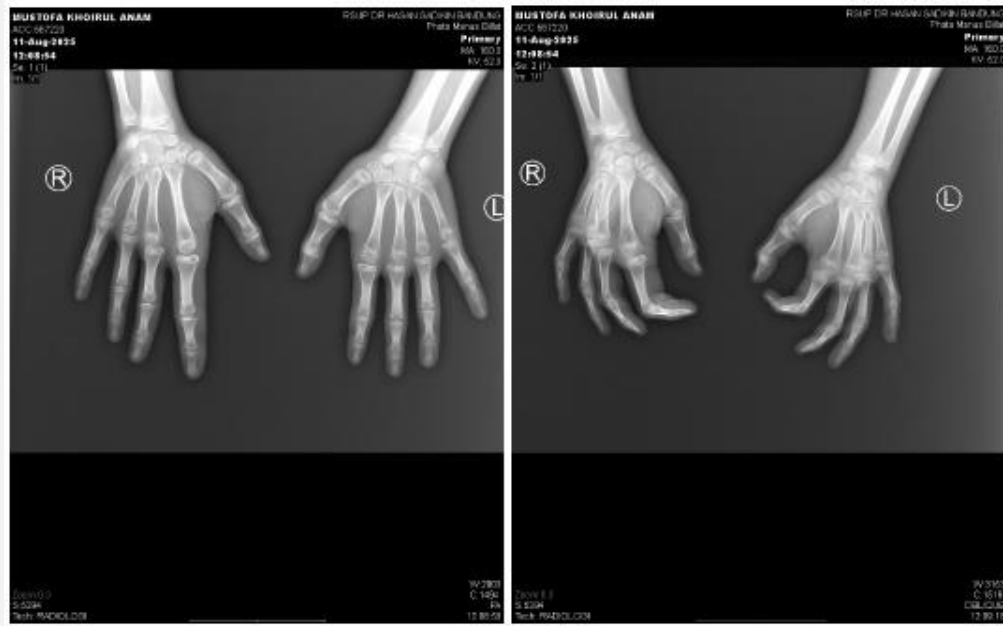


Figure 4. Radiological Results of the Hand

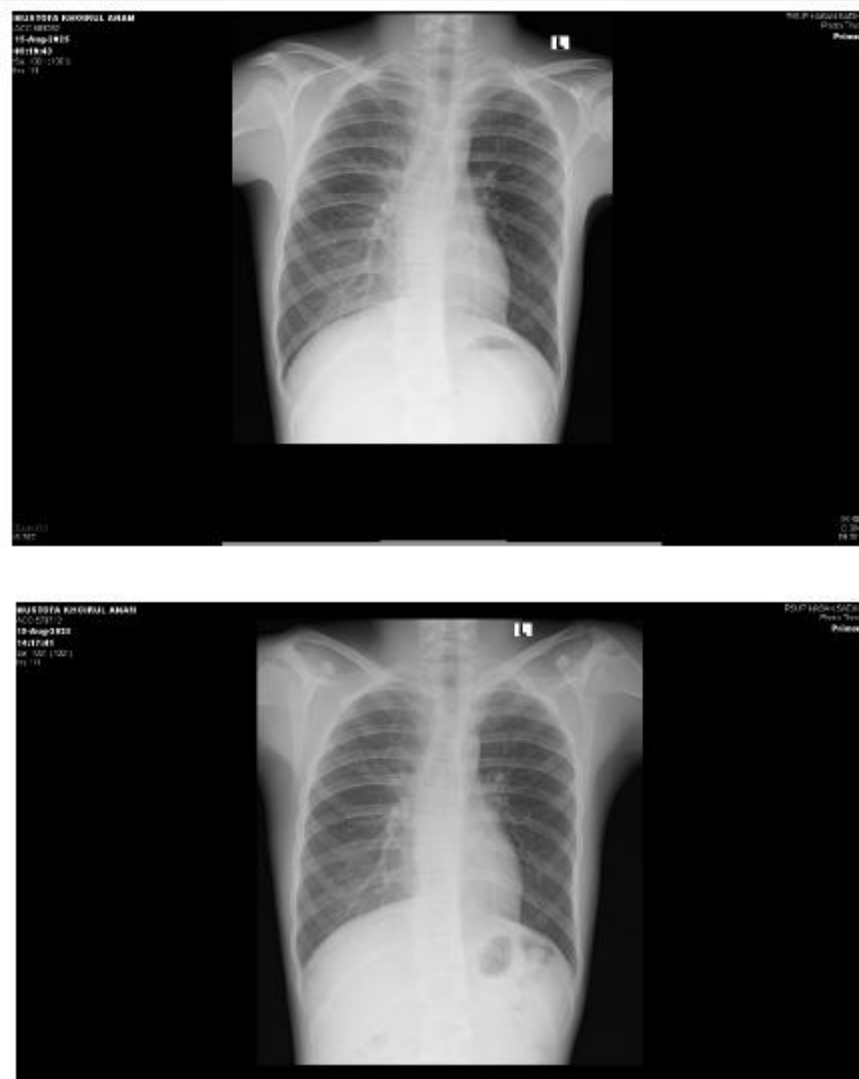






Figure 7. CTA Reconstruction Results (3D HD MIP)

DISCUSSION

The case of MKA provides a very clear illustration of the severe and diagnostically challenging clinical spectrum of Klippel–Trénaunay syndrome. Through an integrative analysis of the medical history, physical findings, and advanced imaging results, several important points can be discussed.

First, regarding the bilateral manifestation. Most literature states that KTS is a unilateral disorder in over 90% of cases [1]. Bilateral involvement of both lower limbs, as in this patient, is a rare variant and is often associated with a somatic genetic mutation occurring very early in embryonic development, thereby affecting mesodermal precursors on both sides of the body [8]. The phenomenon of "spaced toes" noticed since birth in both feet actually represents an early sign of symmetrical soft tissue and bone hypertrophy, which later developed into progressive massive edema.

Second, the radiological findings on CTV showed very significant venous dilatation in the bilateral deep venous systems, including the femoral vein reaching a diameter of 9.57 mm [14]. This is very interesting because in classic KTS, deep vein hypoplasia is often found. However, in this case, massive dilatation occurred, indicating severe chronic volume overload. The presence of a thrombus in the right femoral vein is direct evidence of DVT complication, an inherent risk in KTS patients with extensive venous dilatation [10]. The episodes of pain, redness, and heat reported by the patient were very likely manifestations of thrombophlebitis or recurrent subacute DVT episodes.

Third, the left axillary lump that grew rapidly within one week requires special attention. Given that KTS often involves mixed malformations (CLVM), a cystic lump in an area rich in lymph nodes such as the axilla is most likely a cystic lymphatic malformation or cystic lymphangioma (often called cystic hygroma) [12]. Rapid growth over a short period could be triggered by intracystic hemorrhage or low-grade infection causing sudden lymph fluid accumulation [6]. This explanation is consistent with the fact that KTS has a lymphatic component in up to 50% of patients [1].

Fourth, differentiation between KTS and Parkes Weber syndrome (PWS) was successfully achieved through CTA. The absence of macroscopic arteriovenous fistulas and relatively normal arterial diameters (without early venous filling) confirms that this malformation is purely low-flow [4]. This has major management implications: if the patient

were diagnosed with PWS, the main focus would be arterial embolization, whereas in KTS, the main focus is venous compression and anticoagulation [4].

The patient's current management with anticoagulant drugs and compression stockings is an appropriate step according to international medical standards [1]. However, given that the patient is only 15 years old and in an active growth phase, monitoring for bone length discrepancy is essential. If in the future a significant limb length difference is found that disrupts posture, epiphysiodesis should be considered before growth plate closure [2]. Furthermore, follow-up evaluation of the axillary mass with MRI is recommended to determine whether cyst sclerotherapy is needed if the mass continues to enlarge and interfere with upper extremity mobility or compress the brachial plexus nerves [45].

Overall, this case emphasizes the importance of clinical vigilance for thrombotic complications in KTS patients and the strategic value of vascular radiology imaging in mapping this multimodal disease.

CONCLUSION

Klippel-Trénaunay syndrome is a complex and progressive pathological entity requiring a multidisciplinary diagnostic and therapeutic approach. In this adolescent male patient, the diagnosis of KTS was established based on the classic triad manifesting bilaterally in the lower limbs, reinforced by radiological findings of massive venous dilatation and collateralization in the deep venous system. Detection of a thrombus in the right femoral vein by CTV confirms the high risk of thromboembolic complications in the KTS population and validates the need for adequate anticoagulant therapy.

CTA examination successfully excluded the differential diagnosis of Parkes Weber syndrome, ensuring that the current management strategy focusing on low-flow management is the safest and most effective path. The presence of a left axillary mass indicates progressive involvement of the lymphatic system (lymphangioma), which requires close monitoring using MRI modalities for future intervention planning. Patient education regarding the routine use of compression stockings and adherence to anticoagulant therapy is vital to prevent fatal complications such as pulmonary embolism and to maintain the patient's quality of life during the transition to adulthood.

REFERENCES

1. Klippel-Trenaunay-Weber Syndrome - StatPearls - NCBI Bookshelf - NIH. Available at: <https://www.ncbi.nlm.nih.gov/books/NBK558989/>
2. Klippel-Trenaunay Syndrome - PMC - NIH. Available at: <https://pmc.ncbi.nlm.nih.gov/articles/PMC3881214/>
3. Klippel-Trenaunay Syndrome | Applied Radiology. Available at: <https://appliedradiology.com/articles/klippel-trenaunay-syndrome>
4. Imaging Klippel-Trenaunay and Parkes Weber syndromes: A unilateral limb overgrowth case series. Available at: <https://sajr.org.za/index.php/sajr/article/download/3246/3946>
5. Imaging Klippel-Trenaunay and Parkes Weber syndromes: A unilateral limb overgrowth case series. Available at: https://scielo.org.za/scielo.php?script=sci_arttext&pid=S2078-67782025000100020
6. Klippel-Trenaunay syndrome - Symptoms and causes - Mayo Clinic. Available at: <https://www.mayoclinic.org/diseases-conditions/klippel-trenaunay/symptoms-causes/syc-20374152>
7. Klippel-Trénaunay syndrome - DermNet. Available at: <https://dermnetnz.org/topics/klippel-trenaunay-syndrome>
8. Case report: klippel-trenaunay syndrome. Available at: https://cdn.publisher.gn1.link/residenciapediatrica.com.br/pdf/en_v14n2e1039.pdf
9. Klippel-Trenaunay Syndrome Case - Applied Radiology. Available at: <https://appliedradiology.com/articles/klippel-trenaunay-syndrome-case>
10. Management: Clotting - K-T Support Group. Available at: <https://k-t.org/about-kt/management-clotting>
11. Multimodality imaging approach in a patient with Klippel-Trenaunay syndrome - PMC. Available at: <https://pmc.ncbi.nlm.nih.gov/articles/PMC6706670/>
12. Lymphatic Malformations - Dermatologic Disorders - Merck Manual Professional Edition. Available at: <https://www.merckmanuals.com/professional/dermatologic-disorders/benign-skin-tumors-growths-and-vascular-lesions/lymphatic-malformations>

13. An Atypical Case of Splenic Lymphangioma - Journal of Radiology Case Reports. Available
at: <https://www.radiologycases.com/index.php/radiologycases/article/view/5672>
14. Alwalid O, Makamure J, Cheng QG, Wu WJ, Yang C, Samran E, Han P, Liang HM. Radiological Aspect of Klippel-Trénaunay Syndrome: A Case Series With Review of Literature. Curr Med Sci. 2018 Oct;38(5):925-931.
15. (PDF) Imaging Klippel-Trenaunay and Parkes Weber syndromes: A unilateral limb overgrowth case series - ResearchGate. Available
at: <https://www.researchgate.net/publication/397758311>
16. Vaskularisasi Ekstremitas Atas dan Bawah | PDF - Scribd. Available
at: <https://id.scribd.com/presentation/438586112>
17. Vaskularisasi Pada Extremitas Inferior - Scribd. Available
at: <https://id.scribd.com/document/641350777>
18. Arteri Dan Vena Ekstremitas | PDF - Scribd. Available
at: <https://id.scribd.com/document/170021228>
19. Pembuluh Darah Ekstremitas Bawah | PDF - Scribd. Available
at: <https://id.scribd.com/presentation/490279159>
20. Vaskularisasi Extremitas | PDF - Scribd. Available
at: <https://id.scribd.com/document/530530057>
21. Vaskularisasi Ekstremitas Inferior | PDF - Scribd. Available
at: <https://id.scribd.com/doc/262930323>
22. Vaskularisasi Regio Femoris dan Vena | PDF - Scribd. Available
at: <https://id.scribd.com/document/485906171>
23. USG Vaskular Ekstremitas Bawah | PDF - Scribd. Available
at: <https://id.scribd.com/presentation/462234671>
24. Sistem Sirkulasi Ekstremitas Inferior | PDF - Scribd. Available
at: <https://id.scribd.com/document/467857265>
25. Venous Insufficiency - StatPearls - NCBI Bookshelf. Available
at: <https://www.ncbi.nlm.nih.gov/books/NBK430975/>
26. Varicose Veins - StatPearls - NCBI Bookshelf. Available
at: <https://www.ncbi.nlm.nih.gov/books/NBK470194/>

27. Memahami Sistem Limfatik dan Gangguan yang Dapat Terjadi - Alodokter. Available at: <https://www.alodokter.com/memahami-sistem-limfatik-dan-gangguan-yang-dapat-terjadi>
28. Anatomi Sistem Limfatik | PDF. Available at: <https://id.scribd.com/document/801494639>
29. Anatomi Sistem Limfatik - Umeds. Available at: <https://umeds.id/library-learning/concept-pages/anatomi-sistem-limfatik>
30. Anatomi Sistem Limfatik Manusia | PDF. Available at: <https://id.scribd.com/presentation/458797724>
31. sistem limfatis. Available at: <https://kedokteran.med.unhas.ac.id/wp-content/uploads/2016/10/SISTEM-LIMFATIS.pdf>
32. Nodus limfa inguinal - Wikipedia. Available at: https://id.wikipedia.org/wiki/Nodus_limfa_inguinal
33. SISTEM VASKULAR dan LIMFATIK.pdf. Available at: <https://repository.unimal.ac.id/5044/1/>
34. Kelenjar Getah Bening: Anatomi, Fungsi, dan Gangguannya - Hello Sehat. Available at: <https://hellosehat.com/sehat/informasi-kesehatan/kelenjar-getah-bening-adalah/>
35. Unusual cystic hygromas: Case report - PMC. Available at: <https://pmc.ncbi.nlm.nih.gov/articles/PMC10943632/>
36. Cystic Hygroma- A Case Report and its Embryological Basis - Longdom Publishing. Available at: <https://www.longdom.org/open-access/cystic-hygroma-a-case-report-and-its-embryological-basis-21777.html>
37. Cystic Hygroma - Longdom PDF. Available at: <https://www.longdom.org/open-access-pdfs/cystic-hygroma-a-case-report-and-its-embryological-basis-2161-0940.1000169.pdf>
38. Klippel-Trenaunay and Parkes-Weber syndromes: two case reports - PMC. Available at: <https://pmc.ncbi.nlm.nih.gov/articles/PMC5944310/>
39. Case Report: Klippel-Trénaunay-Weber syndrome - RBCP. Available at: <http://rbcp.org.br/details/2175/en-US/>
40. Klippel-Trenaunay and Parkes-Weber syndromes: two case reports - SciELO. Available at: <https://www.scielo.br/j/jvb/a/HzKLF8cgmSsrY7g5vpdxCnd/>

41. Clinical Practice Guidelines for Klippel-Trenaunay Syndrome (KTS) - K-T Support Group. Available at: <https://k-t.org/assets/images/content/BCH-Klippel-Trenaunay-Syndrome-Management-Guidelines-1-6-2016.pdf>
42. Imaging Klippel-Trenaunay and Parkes Weber syndromes - SAJR. Available at: <https://sajr.org.za/index.php/sajr/article/view/3246/3944>
43. (PDF) Clinical and imaging findings of klippel-trenaunay syndrome: A case report - ResearchGate. Available at: <https://www.researchgate.net/publication/358827706>
44. A Case of Klippel-Trenaunay Syndrome Complicated by Group A Streptococemia - PMC. Available at: <https://pmc.ncbi.nlm.nih.gov/articles/PMC10924929/>
45. Klippel-Trenaunay Syndrome in the Distal Part of the Unilateral Upper Limb - PMC. Available at: <https://pmc.ncbi.nlm.nih.gov/articles/PMC11930299/>
46. Deep vein thrombosis in the setting of Klippel-Trenaunay syndrome and sirolimus treatment. Available at: <https://pmc.ncbi.nlm.nih.gov/articles/PMC8358289/>
47. Compva: Klippel-Trénaunay syndrome - Compendium Vascular Anomalies. Available at: <https://www.compva.com/science/klippel-trenaunay-syndrome>
48. Clinical and Imaging Findings of Klippel-Trenaunay Syndrome: A Case Report - Neliti. Available at: <https://media.neliti.com/media/publications/412203>
49. Recurrent Venous Thromboembolism in Klippel-Trenaunay Syndrome Despite Warfarin - PMC. Available at: <https://pmc.ncbi.nlm.nih.gov/articles/PMC7205377/>
50. Klippel-Trenaunay syndrome - MedlinePlus. Available at: <https://medlineplus.gov/download/genetics/condition/klippel-trenaunay-syndrome.pdf>
51. Compression therapy in a patient with Klippel-Trénaunay syndrome. Available at: <https://journals.cambridgemedia.com.au/jwm/volume-24-number-3>
52. Current Understanding of Klippel-Trenaunay Syndrome: A Review Article. Available at: <https://scholarworks.gvsu.edu/cgi/viewcontent.cgi?article=1685>
53. (PDF) Klippel-Trenaunay Syndrome with Extensive Lymphangiomas - ResearchGate. Available at: <https://www.researchgate.net/publication/283333918>
54. Klippel-Trenaunay Syndrome (KTS): Symptoms and Treatment - Cleveland Clinic. Available at: <https://my.clevelandclinic.org/health/diseases/17152>
55. Klippel-Trenaunay Syndrome and Cellulitis - AOA. Available at: <https://aoao.org/2025/03/20/klippel-trenaunay-syndrome-and-cellulitis/>

56. Presentation and Management of Klippel-Trenaunay Syndrome - PMC. Available at: <https://pmc.ncbi.nlm.nih.gov/articles/PMC7282379/>
57. A Rare Case of Klippel-Trénaunay Syndrome - PMC. Available at: <https://pmc.ncbi.nlm.nih.gov/articles/PMC9645192/>
58. Parkes Weber or Klippel-Trenaunay syndrome? - PubMed. Available at: <https://pubmed.ncbi.nlm.nih.gov/15007616/>
59. Klippel-Trenaunay Syndrome (KTS) - Johns Hopkins Medicine. Available at: <https://www.hopkinsmedicine.org/health/conditions-and-diseases/klippel-trenaunay-syndrome>
60. Venous thromboembolism and prothrombotic parameters in Klippel-trenaunay syndrome - NJM. Available at: <https://www.njmonline.nl/getpdf.php?id=1314>
61. Klippel-Trenaunay syndrome with splenic cystic lymphangioma - Fetal Medicine Foundation. Available at: <https://fetalmedicine.org/abstracts/2024/var/pdf/abstracts/2024/06352.pdf>
62. What is the management plan for Klippel-Trenaunay syndrome? - Dr.Oracle. Available at: <https://www.droracle.ai/articles/638476>
63. Terapi Antikoagulan pada Anak - Sari Pediatri. Available at: <https://saripediatri.org/index.php/sari-pediatri/article/download/2003/pdf>
64. Klippel-Trénaunay Syndrome - Seattle Children's Hospital. Available at: <https://www.seattlechildrens.org/conditions/klippel-trenaunay-syndrome-kts/>
65. Recurrent Cystic Hygroma - Internet Scientific Publications. Available at: <https://ispub.com/IJS/3/1/11626>