Introduction: Klippel Trenaunay Syndrome (KTS) is a rare, congenital malformation. Several theories have been postulated to describe its pathogenesis. However, the exact etiology is not known. It’s characterized by a triad of (1) haemangioma due to capillary malformation, (2) bone and soft tissue hypertrophy, and (3) varicose veins. Interestingly, lipodystrophy rather than hypertrophy of the involved limbs had been described in some cases. The clinical presentation of this syndrome is variable ranging from minimal disease to severe presentation such as significant cosmetic disfiguring, life threatening bleeding and embolism. The KTS is classified according to severity. This is important step in away to educate patients, to predict prognosis and to set treatments, especially in severe cases. Physicians should not only be attentive to the physical aspects but also to the psychological and social aspects of KTS.

Case presentation: A 16-year-old boy presented with multiple port-wine stain type vascular anomalies and varicose veins. Also, there was a marked deforming enlargement of his right foot, right knee and number of his left toes with, striking gigantism of both lower limbs. This is consistent with clinical presentation of KTS. Furthermore, there were additional features matching myotonic dystrophy outlook; face showed a remarkable lipodystropy with bilateral upper limbs wasting and atrophy. This could represent the Inverse- Klippel Trenaunay Syndrome. According to our knowledge, this had never been reported to be associated with Klippel Trenaunay Syndrome.

Conclusion: Klippel Trenaunay and Inverse Klippel Trenaunay Syndrome can be seen in the same patient.
varicose veins were present over the medial aspect of his leg and over the trunk anteriorly and posteriorly (Figure 3). Patient’s face showed gross lipodystrophy, wasting of sternomastoid muscles and upper arms muscles, bilaterally (figure 4A-4D). There was no motor weakness despite the noticeable muscular atrophy. Neurological examination showed no cranial nerves abnormalities. There was no scoliosis, skeletal 
or spinal anomalies.

Laboratory investigations revealed very low hemoglobin of 3.8 g/dl (normal range 13.5-18 g/dl), low haematocrit of 13.1% (normal range 38.8-50%), low mean cell volume of 56.8 fl (normal value 76-96 fl), Low mean cell hemoglobin of 16.4 pg (normal value 27-32 pg), and low mean cell hemoglobin concentration of 29.0 g/dl (normal range 30-36 g/dl). There was a profound iron deficiency with an iron level of 23 mcg/dl (normal value 6-170 mcg/dl). Other laboratory results including electrolytes, liver function, renal function, and coagulation parameters were normal. Urine analysis showed no haematuria and stool showed no occult blood. Musculoskeletal x-rays revealed no 
distorting bony overgrowth (hyperostosis).
On the basis of the above triad of port wine stain, limb hypertrophy and varicosity diagnosis of Klippel Trenaunay Syndrome was made. The patient was treated conservatively by elastic compression stockings, systemic antibiotic, daily dressing of an ulcer region and blood transfusion. With this conservative treatment his bleeding ulcer diminished spontaneously, his blood level increased and his ulcer started to heal. Patient was giving an appointment to the surgical outpatient clinic for regular dressing of his ulcer and has been advised to keep his limb elevated whenever possible.

Discussion

The constellation of soft-tissue hypertrophy, varicose veins and a cutaneous hemangiomatous lesion is characteristic of Klippel Trenaunay Syndrome.

The history of the patient revealed a marked interval progression of soft-tissue hypertrophy which occurred with his growth. The patient and his parent stated that the sizes of his limbs were growing excessively with time, a feature that is reported in KTS.

Gingivitis is a result of bone elongation or circumferential soft-tissue hypertrophy [9]. Girls are more affected than boys. The left side is more involved than the right. This preferential of left side involvement suggest some mechanical factor responsible [10]. The condition may involve half of the body and give rise to true hemi-hypertrophy (bilateral and contra lateral involvement occurs). Gigantism of the whole limb may be produced or hypertrophy may be confined to part of the extremity [10]. In this patient, the gigantism involved different parts of both lower limbs with more prominent hypertrophy changes in the right lower limb (right knee and right foot). Left leg showed less hypertrophy changes that confined to the lower end of the left leg and the third, fourth and fifth left toe.

Danarti et al. reported that Klippel Trenaunay Syndrome is defined by a coexistence of nevus flammeus and overgrowth of one or more limbs [4]. They reported that, deficient growth of an affected limb may likewise be noted. Hence, a term inverse Klippel Trenaunay Syndrome has been proposed by them. They collected from the literatures a number of cases of Klippel Trenaunay Syndrome associated with deficient growth such as shortening or hypoplastic muscle mass of the affected extremity. They proposed that some patients may carry compound heterozygotes 'plus' and a 'minus' allele at the responsible gene locus, and the post zygotic recombination may give rise to two different cell clones that are homozygous for either allele [4].

Features of Klippel Trenaunay Syndrome have been reported in myotonic dystrophy patient [11]. Myotonic dystrophy: an autosomal dominant disorder, is the commonest muscular dystrophy in adults [12]. The clinical severity and age of presentation are extremely variable [12]. The mean age at onset of myotonic dystrophy lies between 20 and 25 years [13]. Absence of clinical myotonia in the adult myotonic dystrophy patients vaies from 10% in one study to 45.8% in another study [14]. Our patients showed external features of myotonic dystrophy: facial muscle lipodystrophy and wasting of the neck sternomastoid and upper limbs muscles.

To our knowledge, this is the first case having bilateral wasting of both upper limbs and having a myotonic dystrophy like face with presence of a typical features of Klippel Trenaunay Syndrome. Apart from the outlook of myotonic dystrophy, the patient has no other feature of myotonic dystrophy. Our patient may be having a combined Klippel Trenaunay-inverse Klippel Trenaunay Syndrome or he has an incomplete presentation of myotonic dystrophy and KTS.

Literatures Review

Historic overview

Long time ago, Virchow and both Hebra and Kaposi described the condition as elephantiasis telangectodes [10]. In 1900, the famous paper of two French physicians "Maurice Klippel and Paul Trenaunay" about angiodysplastic disorder came out. Where they described two patients with haemangiomatoses lesions of the skin associated with asymmetric soft tissue and bone hypertrophy, and coined the term "naevus variqueux osteohypertrophique" [15]. This mesodermal abnormality syndrome characterized by clinical trial of (1) port-wine stain due to capillary malformation, (2) bone and soft tissue hypertrophy; and (3) varicose veins.

Etiology

The precise etiology is unknown. The KTS syndrome is generally thought to occur sporadically [16]. However, in some cases, clinical manifestations of the syndrome have been found in family members, suggesting an autosomal dominant inheritance [16]. The presumed pathogenetic pathway is that of mesodermal developmental derangement, leading to maintenance of microscopic arteriovenous communications in the limb bud, with consequent development of the naevas, superficial varices, and limb hypertrophy [17]. Several hypotheses regarding cause and pathogenesis in KTS exist, but none explains the full characteristics of KTS. Hypotheses include an alteration in vascular remodeling, perhaps at the level of altered angioiopetin-2 antagonism [18]. Servelle’s theory stated that there is a primary obstruction of the venous system resulting in venous hypertension and thus development of abnormal venous pathways and tissue overgrowth [19]. Also, maintenance of microscopic arteriovenous communication in the limb bud vein had been proposed [17]. Others suggested alteration of the balance between angiogenesis and vasculogenesis, a process that is controlled by numerous genes [20].

Clinical presentation

The classic clinical triad includes varicose veins, cutaneous capillary malformations, and tissue hypertrophy that usually involve the extremities. On the other hand, In some subsets of patients only 2 of the 3 classic findings are present [21]. The clinical presentation of KTS is variable [22]. It ranges from port wine stains and few varicose veins causing cosmetic deformity to severe disability associated with massive limb overgrowths, chronic pain syndrome, skin infections and arthritis [23]. Thrombo-embolism and life-threatening pelvic or recurrent rectal bleeding as a result of venous malformations had been reported [24].

Deep venous system anomalies are reported to be present in 8% to 18% of patients with KTS [25]. However, others reported prominent superficial varicose veins are present in a majority of patients with Klippel Trenaunay Syndrome [9,26]. In over two-thirds of patients, a characteristic incompetent lateral venous channel arises near the ankle and extends a variable distance up the extremity to the infrainguinal or pelvic deep venous system [27]. This venous malformation frequently present as persistent embryonic veins, of which the lateral marginal vein (the vein of Servelle) has been the most typical finding, which found in 68-80% of patients [28].

Deep vein anomalies range from venous hypoplasia to frank aneurysm and valve hypoplasia to avalulcia. The prevalence of deep venous aplasia or hypoplasia, as detected with venographic techniques (ascending venography and varicography), ranges from 18% [27] to
Lymphatic malformations have also been common in up to 70% of cases [1], that include primary lymphhaedema, cystic hygroma or lymphangiectasia associated with reflux of chyle [29].

Varicose veins and venous malformations can involve abdominal and pelvic organs. Rarely, patients with KTS can have intraosseous vascular malformations [1]. Genitourinary system involvement includes the penis, scrotum, vagina, vulva, and bladder. Bleeding from the vascular malformations can present as haematomata, haematuria, rectal bleeding, intracerebral or intraspinal haemorrhage. Cha et al. reported that GI tract involvement may be more common in KTS than previously believed because most of cases remain unrecognized without overt symptoms [30].

Although Klippel Trenaunay Syndrome generally involves only one of the lower extremities, bilateral involvement, upper extremity involvement, single limb involvement (in 80-85% of cases) or extension into the trunk may occur [9]. The lower limb is the site of malformation in approximately 95% of patients [18,31]. The bony abnormalities may affect all bones in an extremity or limited to one or two bones. The enlargement of the extremity consists of bone elongation or circumferential soft-tissue growth of the affected limb [9,27]. Hypertrophy is caused by local hyperemia and secondary venous stasis [32] which are attributed to arteriovenous shunting and venous anomaly [33].

Bone and soft tissue hypertrophy may be evident at birth or may become evident as the patient grows [34]. The cutaneous vascular lesion is generally a capillary malformation and usually involves the enlarged limb, although involvement of the whole side of the body or of the contralateral limb may be seen [9,27]. In addition, other bone deformities can be seen such as macrodactyly, syndactyly, split hand deformity, phalangeal agenesis and hip joint dislocation [35]. Soft tissue hypertrophy may be limited to a localized mass on the back or chest, or it can be diffuse involving an entire arm or leg [29].

The capillary hemangioma or port-wine stain usually presents first [31]. This hemangioma has a distinct, linear border that respects the midline and is often increase on the lateral aspect of the limb. It is typically of the nevus flammeus type, but cavernous hemangiomas with port wine stain or lymphangiomatosis may also occur [31]. Hemangioma depth is variable. It may be limited to the skin or extend deeper to subcutaneous tissue, muscle, bone, and visceral organs, which may lead to internal hemorrhage that may manifest as haematuria or haematochezia. Large cutaneous lesions may sequestrate platelets leading to Kasabach-Merritt syndrome, a type of consumptive coagulopathy [36]. Interestingly, cases of epidural haemangioma and angiomylipoma have been reported to occur at the same segmental level as cutaneous haemangioma in KTS syndrome [37].

Complications

Klippel Trenaunay Syndrome is associated with both local and systemic complications. Local complications include Extremity pain, stasis dermatitis, cellulitis, ulceration and spontaneous cutaneous hemorrhage. Chronic venous insufficiency, coagulopathy, thrombosis or thrombophlebitis are commonly encountered in KTS [9,27,32]. Gangrene can occur as a result of thrombophlebitis. Other systemic complications include consumptive coagulopathy and congestive cardiac failure. Coexisting ipsilateral angiomatosis may be exemplified by neurovascular anomalies such as cerebral arteriovenous fistulae [38], and spinal cord arteriovenous malformations [39]. Abdominal viscera can be affected by ipsilateral angiomatosis that involve the colon [34], and the urinary tract [40] with resulting rectal bleeding and haematuria. Intrathoracic manifestations include increased liability to pulmonary embolism [27], pulmonary vein varicosities [33] and lymphangectatic sclerosis, that give rise to pleural and pericardial effusions [41]. Renal system involvement might occur with a renovascular hypertension [42]. Clinical sequelae of the lymphatic component of the syndrome include lymphangitis, cutaneous lymphatic vesicles, lymphorrhrea, or mass effect from macrocystic portions of lymphatic malformations [43].

Diagnosis

Diagnosis is essentially a clinical one. Proper physical examinations of the limbs and other part of the body are important. Work-up of the lesion may involve noninvasive imaging like Doppler ultrasound scanning of the venous system of the leg to establish patency, incompetence, thrombosis, arteriovenous shunting and malformations and hypoplasia anomalies. Clinical examination and ultrasound can rule out diagnosis of KTS [44]. Other non invasive techniques include standard radiography to measure limb length, and magnetic resonance imaging (MRI) to look for bone, fat, muscle hypertrophy and lymphedema can be utilized. Contrast venography and computed tomography scans can be used to evaluate deep venous system and collateral, especially when ablation of dilated superficial embryonic vein is under consideration [3]. Finally, lymphoscintigraphy can be used to evaluate the lymphatic system.

Management

Although most patients do well without treatment, management of this syndrome includes careful diagnosis, prevention and treatment of complications [45]. There are some absolute indications of treatment such as haemorrhage, infections, acute thromboembolism and refractory ulcers. Generally, the management of this syndrome can be divided into medical and surgical interventions [32]. Multidisciplinary management approach for KTS is warranted [3]. The paediatrician, internist, phlebologist, orthopaedic, plastic and vascular surgeons, interventional radiologist, cardiologist, vascular internist and a physical therapy physician can all be involved in KTS patient’s management [1].

Management is largely conservative and the extent of diagnostic evaluation is determined by the planned treatment. Stockings or pneumatic compression is the hallmark of conservative management. It’s indicated for chronic venous insufficiency, lymphedema, recurrent cellulitis, and recurrent bleeding. Percutaneous sclerosis of localized venous malformations or superficial venous varicosities may be indicated in some patients [46]. Patient with severe chronic venous insufficiency, disturbing cosmetic appearance, and complications of haemangioma, may benefit from surgical treatment. Techniques for ablation of superficial veins and malformations are individualized and may include sclerotherapy with alcohol or foam, that showed a good success but can cause nerve injury and cutaneous damage [47]. Foam sclerotherapy, with sodium tetra decyl sulphate and polidocanol, has been shown to be of a low risk and effective treatment for superficial venous disease [48,49]. Patient’s cosmetic satisfaction with multiple session foam sclerotherapy has been reported in about 71.5% of treated patients [50]. Others treatment approaches includes endovenous thermal ablation, surgical stripping and phlebectomy. Local wound care, compression dressings, special orthopaedic footwear and lifestyle modification may also be required to manage activities of daily living and improve the function of the limb [51]. An amputation may be required for bone overgrowth with non-healing de-cubitus ulcers, for recurrent bleeding from haemangiomas or when the large size of the
limb had interfered with the daily activities [32]. Care must be taken because the condition of patients with Klippel-Trenaunay Syndrome may worsen if intervention is performed on dilated superficial collateral veins associated with deep vein hypoplasia. Imaging before vascular interventions must confirm venous anatomy and deep venous drainage [26].

Regarding limb hypertrophy, heel inserts are generally sufficient for limb discrepancies of 1.5 cm or less. For greater discrepancies, orthopedic surgery may be considered. For overgrowth of one limb epiphyseodesis and for severe arthritis total knee arthroplasty has shown good results. [23]. In addition, excision of soft-tissue hypertrophy and epiphyseodesis can be used to control leg length discrepancy [26–28].

Vascular abnormalities are congenital and thus do not respond to agents used in the treatment of haemangiomas, such as prednisone and interferon-α [32]. Nowadays, pulsed-dye laser treatments can lighten the superficial haemangioma component [52]. However, the deeper vascular malformations are often inadequately treated. Their large size is also a factor that hinders rate of clearing with laser treatment. Second-generation pulsed-dye lasers have long pulsed widths, which, along with the dynamic cooling device, allows for safe use at higher frequencies and greater clinical improvement. Also, the treating clinician may consider the use of laser treatments with the 595-nm V-beam long-pulsed-dye laser for a better clinical improvement of the skin lesions [32].

Cellulitis and thrombophlebitis can be managed with analgesics, antibiotics, and corticosteroids. In patients with a history of recurrent cellulitis, intermittent or prophylactic antibiotics may be considered [1].

Anticoagulant therapy is indicated in acute thrombosis and prophylactically prior to surgical procedures [22]. Intraoperative use of tourniquet will decrease bleeding and selective use of an inferior vena cava filter will prevent pulmonary embolism [3].

Gastrointestinal and urogenital involvement is not low as was previously thought and may be seen in about 20 % of cases [27,53,54]. Persistent haematocheza, haematuria, and vaginal and oesophageal bleeding are considered indications for surgical intervention. Those patients usually require endoscopic cauterization, but sometimes refractory bleeding may require colonic resections [55,56]. Finally, microsurgery, endovascular embolization, and, more recently, stereotactic radiosurgery are used for the treatment of the spinal vascular malformation [21].

Conclusion

It is important to recognize that Klippel-Trenaunay Syndrome is a unique that requires a comprehensive multidisciplinary management approach for a better patient care. Lifelong clinical follow-up is mandatory in such patient as the natural history of the various organs involvement is one of progressive deterioration.

Consent

Written informed consent was obtained from the patient and his family for publication of this case report and any accompanying images.

Authors’ Contributions

HS wrote the manuscript and compiled the figures. AI edited the manuscript. All authors analyzed and interpreted the patient data. All authors read and approved the final manuscript.

References

1. Zea MI, Hanif M, Habib M, Ansari A (2009) Klippel-Trenaunay Syndrome: a case report with brief review of literature. J Dermatol Case Rep 3: 56-59.

2. Fait G, Daniel Y, Kupferminc MJ, Gull I, Peyser MR, et al. (1996) Klippel- Trenaunay-Weber syndrome associated with fetal growth restriction. Hum Reprod 11: 2544-2545.

3. Giovizcpi P, Driscoll DJ (2007) Klippel-Trenaunay syndrome: current management. Phlebology 22: 291-298.

4. Danarti R, König A, Bittar M, Happel R (2007) Inverse Klippel-Trenaunay syndrome: review of cases showing deficient growth. Dermatology 214: 130-132.

5. Oduber CE, Khemlani K, Sillevis Smitt JH, Hennekam RC, van der Horst CM (2010) Baseline Quality of Life in patients with Klippel-Trenaunay syndrome. J Plast Reconstr Aesthet Surg 63: 603-609.

6. Mazaeyer E, Enjolras O, Laurian C, Houdart E, Drouet L (2002) Coagulation abnormalities associated with extensive venous malformations of the limbs: differentiation from Kasabach-Merritt syndrome. Clin Lab Haematol 24: 243-251.

7. Alomari AI (2009) A truly unusual overgrowth syndrome: an alternative diagnosis to Klippel-Trenaunay-Weber syndrome. Intern Med 48: 493-494.

8. Weber FP, A.f.i.c.w.h.o.l.a.h. BJQ (1970) Angioma formation in connection with hypertrophy of limbs and hemihypertrophy. Br J Dermatol 19: 231-235.

9. Gioviczki P, Stanson AW, Stickler GB, Johnson CM, Toomey BJ, et al. (1991) Klippel-Trenaunay syndrome: the risks and benefits of vascular interventions. Surgery 110: 469-479.

10. Moynahan EJ (1961) Naevoid hypertrophy of the lower limbs, with gigantism of digits (Klippel-Weber-Trenaunay syndrome). Proc R Soc Med 54: 695-696.

11. Gourie-Devi M, Mehta BC (1982) Association of Klippel-Trenaunay-Weber syndrome with myotonic dystrophy. J Neurol Neurosurg Psychiatry 45: 1074-1075.

12. Lotz BP, van der Meyden CH (1985) Myotonic dystrophy. Part II. A clinical study of 96 patients. S Afr Med J 67: 815-817.

13. Harper PS (1979) Myotonic dystrophy. Philadelphia: WB Saunders, p. 31-33.

14. Pryse-Phillips W, Johnson GJ, Larsen B (1982) Incomplete manifestations of myotonic dystrophy in a large kinship in Labrador. Ann Neurol 11: 582-591.

15. Klipple M, P Trenaunay (1900) Du naevus variqex osteohypertrophique. Arch Gen Med. 77: 641-672.

16. Aelvoet GE, Jorens PG, Roelen LM (1992) Genetic aspects of the Klippel- Trenaunay syndrome. Br J Dermatol 126: 603-607.

17. Baskerville PA, Ackroyd JS, Browse NL (1985) The etiology of the Klippel- Trenaunay syndrome. Part II. A clinical study of 96 patients. S Afr Med J 67: 815-817.

18. Baskerville PA, Ackroyd JS, Browse NL (1985) The etiology of the Klippel- Trenaunay syndrome. Ann Surg 202: 624-627.

19. Berry SA, Peterson C, Mize W, Bloom K, Zachary C, et al. (1998) Klippel- Trenaunay Syndrome. Am J Med Genet 79: 319-326.

20. Servede M, Babiliot J (1980) Deep vein malformations in the Klippel-Trenaunay Syndrome. Phlebologie 33: 31-36.

21. Tian XL, Kadaba R, You SA, Liu M, Timur AA, et al. (2004) Identification of an angiogenic factor that when mutated causes susceptibility to Klippel-Trenaunay syndrome. Nature 427: 640-645.

22. Rohany M, Shaibani A, Arafat O, Walker MT, Russell EJ, et al. (2007) Spinal arteriovenous malformations associated with Klippel-Trenaunay-Weber syndrome: a literature search and report of two cases. AJNR Am J Neuroradiol 28: 584-589.

23. Lee A, Driscoll D, Giovizcpi P, Clay R, Shaughnessy W, et al. (2005) Evaluation and management of pain in patients with Klippel-Trenaunay syndrome: a review. Pediatrics 115: 744-749.

24. Jacob AG, Driscoll DJ, Shaughnessy WJ, Stanson AW, Clay RP, et al. (1998) Klippel-Trenaunay syndrome: spectrum and management. Mayo Clin Proc 73: 28-36.
25. Redondo P, Bastarrrika G, Aguado L, Martinez-Cuesta A, Sierra A, et al. (2009) Foot or hand malformations related to deep vein system anomalies of the lower limb in Klippel-Trénaunay syndrome. J Am Acad Dermatol 61: 621-628.

26. Thomas ML, Macle GB (1974) Phlebography in the Klippel-Trenaunay syndrome. Acta Radiol Diagn (Stockh) 15: 43-56.

27. Baskerville PA, Ackroyd JS, Lea Thomas M, Browse NL (1985) The Klippel-Trenaunay syndrome: clinical, radiological and haemodynamic features and management. Br J Surg 72: 232-236.

28. Kim YW, Lee BB, Cho JH, Do YS, Kim DI, et al. (2007) Haemodynamic and clinical assessment of lateral marginal vein excision in patients with a predominantly venous malformation of the lower extremity. Eur J Vasc Endovasc Surg 33: 122-127.

29. Servelle M (1985) Klippel and Trénaunay's syndrome. 768 operated cases. Ann Surg 201: 365-373.

30. Cha SH, Romeo MA, Neutze JA (2005) Visceral manifestations of Klippel-Trénaunay syndrome. Radiographics 25: 1694-1697.

31. al-Salman MM (1997) Klippel-Trénaunay syndrome: clinical features, complications, and management. Surg Today 27: 735-740.

32. Jh MHR (2003) Klippel-Trenaunay syndrome. Dermatol Online J 9: 31.

33. Owens DW, Garcia E, Pierce RR, Castrow FF 2nd (1973) Klippel-Trenaunay-Weber syndrome with pulmonary vein varicosity. Arch Dermatol 108: 111-113.

34. Ghaemrezaei GG, Kangarloo H, Volberg F, Meyers MA (1976) Diffuse cavernous hemangioma of the colon in the Klippel-Trenaunay syndrome. Radiology 118: 673-678.

35. McGroty BJ, Amadio PC (1993) Klippel-Trenaunay syndrome: orthopaedic considerations. Orthop Rev 22: 41-50.

36. Aronoff DM, Roshon M (1998) Severe hemorrhage complicating the Klippel-Trenaunay-Weber syndrome. South Med J 91: 1073-1075.

37. Alexander MJ, Grossi PM, Spetzler RF, McDougall CG (2002) Extradural thoracic arteriovenous malformation in a patient with Klippel-Trenaunay-Weber syndrome: case report. Neurosurgery 51: 1275-1278.

38. Oyesiku NM, Gahm NH, Goldman RL (1988) Cerebral arteriovenous fistula in the Klippel-Trenaunay-Weber syndrome. Dev Med Child Neurol 30: 245-246.

39. Djindjian M, Djindjian R, Huth M, Rey A, Houdart R (1977) Spinal cord arteriovenous malformations and the Klippel-Trenaunay-Weber syndrome. Surg Neurol 8: 229-237.

40. Fligelstone LJ, Campbell F, Ray DK, Rees RW (1994) The Klippel-Trenaunay syndrome: a rare cause of hematuria requiring nephrectomy. J Urol 151: 404-405.

41. Joshi M, Cole S, Knibbs D, Diana D (1992) Pulmonary abnormalities in Klippel-Trenaunay syndrome: A histologic, ultrastructural, and immunocytochemical study. Chest 102: 1274-1277.

42. Samuel M, Spitz L (1995) Klippel-Trenaunay syndrome: clinical features, complications and management in children. Br J Surg 82: 757-761.

43. James CA, Allison JW, Waner M (1999) Pediatric case of the day. Klippel-Trénaunay syndrome. Radiographics 19: 1093-1096.

44. Samimi M, Lorette G (2010) Klippel-Trenaunay syndrome. Presse Med 39: 487-494.

45. Jolobe OM (1996) Klippel-Trenaunay syndrome. Postgrad Med J 72: 347-348.

46. Laor T, Burrows PE, Hoffer FA (1996) Magnetic resonance venography of congenital vascular malformations of the extremities. Pediatr Radiol 26: 371-380.

47. Burrows PE, Mason KP (2004) Percutaneous treatment of low flow vascular malformations. J Vasc Interv Radiol 15: 431-445.

48. Cabrera J, Cabrera J Jr, Garcia-Olmedo MA, Redondo P (2003) Treatment of venous malformations with sclerant in microfoam form. Arch Dermatol 139: 1409-1416.

49. Smith PC (2006) Chronic venous disease treated by ultrasound guided foam sclerotherapy. Eur J Vasc Endovasc Surg 32: 577-583.

50. Nitecki S, Bass A (2007) Ultrasound-guided foam sclerotherapy in patients with Klippel-Trenaunay syndrome. Isr Med Assoc J 9: 72-75.

51. Lee BB, Do YS, Byun HS, Choo IW, Kim DI, et al. (2003) Advanced management of venous malformation with ethanol sclerotherapy: mid-term results. J Vasc Surg 37: 533-538.

52. Yamauchi PS, Soriano TT, Lask GP (2000) Treatment of port wine stains using the pulsed-dye laser at 585 nm with the dynamic cooling device. J Cutan Laser Ther 2: 33-36.

53. Wilson CL, Song LM, Chua H, Ferrara M, Devine RM, et al. (2001) Bleeding from cavernous angiomatosis of the rectum in Klippel-Trenaunay syndrome: report of three cases and literature review. Am J Gastroenterol 96: 2783-2788.

54. Furness PD 3rd, Barqawi AZ, Bisignani G, Decker RM (2001) Klippel-Trénaunay syndrome: 2 case reports and a review of genitourinary manifestations. J Urol 166: 1418-1420.

55. Wilson CL, Song LM, Chua H, Ferrara M, Devine RM, et al. (2001) Bleeding from cavernous angiomatosis of the rectum in Klippel-Trenaunay syndrome: report of three cases and literature review. Am J Gastroenterol 96: 2783-2788.

56. Husmann DA, Rathburn SR, Driscoll DJ (2007) Klippel-Trenaunay syndrome: Incidence and treatment of genitourinary sequelae. J Urol 177: 1244-1249.