CASE REPORT

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Endovascular laser ablation management of pediatric Klippel trenaunay syndrome: a case report

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ABSTRACT

Introduction: Klippel trenaunay syndrome (KTS) is a rare and complex congenital vascular anomaly as a combination of vascular capillary malformations (port wine stain), venous malformations (VM) and excessive extremity growth with or without lymphatic malformations. The incident KTS is estimated to be 2 to 5 per 100,000 and found in the male gender without distinguishing race. This syndrome is rare, with a low incidence. Klippel trenaunay syndrome uses conservative management with the use of compression stockings. Other KTS management, such as minimally invasive intervention with endovascular laser ablation (EVLA) therapy, is rarely used due to the rare number of diseases. Thus, this study aims to perform the management of KTS in pediatrics.

Case description: We report 11-year-old girls complaining of pain and swelling on the right leg when standing long and doing activities. Physical examination appears to show a purplish-red birthmark that arises on the surface of the skin. Ultrasound and CT scan angiography with contrast in the lower region extremities showed hypertrophy accompanied by peripheral branches in the cutaneous layer. Patients are performed endovascular laser ablation using general anesthesia. After endovascular laser ablation, the patient was treated for 2 days without any complaints.

Conclusion: KTS is a rare, complex and varied syndrome that has not been found yet. KTS only performs symptomatic treatment depending on clinical and severity. Endovascular laser ablation is a minimally invasive intervention management that must be considered and has a fairly low risk and complication, especially in pediatrics.

Keywords: Klippel trenaunay syndrome, endovascular laser ablation, pediatric.
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INTRODUCTION

Klippel-trenaunay syndrome (KTS) is a combination of vascular malformations whose flow slows (capillaries, lymphatics, and veins) to the extremities with excess growth.[1,2] This syndrome is rare, with a low incidence.[3] The incidence of KTS is estimated to be 2-5 per 100,000 people, with males being the most commonly affected regardless of race.[4] Several cases have been documented in Korea. Sung et al. reported 19 patients diagnosed with KTS, 9 (47%) male and 10 (53%) female. The mean follow-up period was 4.1 years (7 months-9 years).[3]

Klippel-trenaunay syndrome is characterized by congenital malformations most commonly found in the lower or upper extremities of the unilaterally and rarely seen in the body trunk, neck, or head.[1,2] KTS has three symptoms: varicose vein malformations (VMs), capillary malformations (port wine stain), soft tissue, and hypertrophy.[4,5] KTS has clinically varied depending on severity, ranging from having only capillary hemangiomas or varicose veins to having severe deformity in limbs resulting in disability. Vascular malformations may be seen from birth, but accelerated growth of veins and hypertrophy of the extremities will be seen during growth.[4] It also has a specific clinical and radiological examination that distinguishes it from other complex anomalies of the vascular system.[6,7]

The etiology of KTS is currently under investigation. Some experts argue that Klippel- trenaunay syndrome is a complex prototype abnormality with excess growth disorders associated with vascular anomalies. Somatic mutations in the phosphatidylinositol-4-5-bisphosphate 3 kinase catalytic subunit (PIK3CA) gene has been associated with Klippel trenaunay syndrome's pathogenesis. This causes phosphatidylinositol-3-kinase (PI3K)/ protein kinase activation and cell expansion via mTORC2 dysregulation. Mutations arise throughout the embryological stage of development, resulting in the features seen in this disorder.[8]

Klippel-trenaunay syndrome uses conservative management with the use of compression stockings. Pain management in KTS uses anti-inflammatory drugs to relieve pain. Other KTS management, such as minimally invasive intervention with endovascular laser ablation (EVLA) therapy, is rarely used due to the rare number of diseases. EVLA therapy is more satisfying than open surgery. EVLA has evolved and proven safe and effective in adults for varicose vein management related to superficial vein incompetence.[4,6,7]

CASE DESCRIPTION

A young girl of 11 years complained of pain in her right leg. Long periods of standing and doing activities cause pain. Physical examination reveals a purplish-red birthmark on the skin's surface on the right foot from the outer part of the thigh to the calf. When the right foot stands, the left foot appears swollen. There were no complaints about the birthmark. The previous history with similar signs and symptoms was denied. Family history was denied. The history of the first operation in 2019 in the area because there were lumps under normal circumstances, complete vaccines and immunizations, heart abnormalities, and other abnormalities do not exist. The history of other diseases was denied. Before underwent the physical examination, informed consent was obtained from the patient for the publication of this report and any accompanying image.

According to the physical examination, the patient's blood pressure was 116/73 mmHg, pulse rate was 93 beats per minute, respiratory rate was 20 beats per minute, and temperature was 36.6 ^{0}C. The weight and height of the patient were 28 kg and 120 cm. Many spots of various nummular and lenticular sizes were found in the inferior extremities of lateral femoral dexterity, including spider vein, hyperemia, and port wine stain, which spread to the anterior genu region until the lateral cruris (Figure 1).

An ultrasound-supported examination found abnormal veins with a size of approximately 6 cm to 7 cm (figure 2). CT scan angiography with contrasts regional lower extremities showed left extremity hypertrophy, superficial femoral and tibial vein peripheral branch dilation, and tortuous dilatation of superficial veins in the cutaneous/subcutaneous region of the femur and cruris sinistra with an arteriovenous shunt (Figure 3).

Endovascular laser ablation procedures were performed under general anesthesia with vital signs monitoring. The patient was positioned in Trendelenburg upside-down to ensure that the vein was full of blood and thus easier to puncture. The target vein was accessed percutaneously by micropuncture with ultrasound guidelines. Once the target vein was

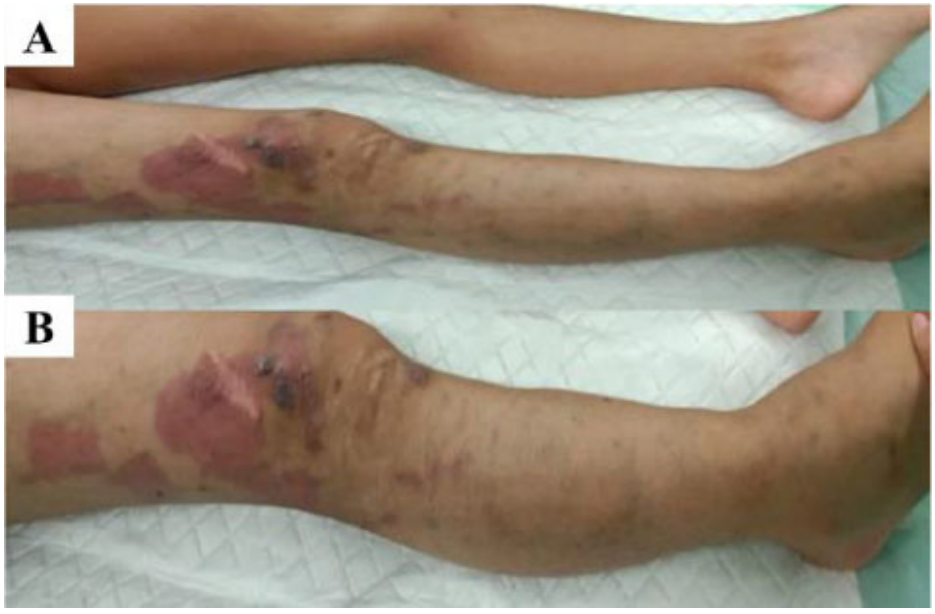

Figure 1. Port wine stain varies in size lateral femoral regions, genu regions and lateral cruris regions. A) Lowe extremity comparison, B) Right lower extremity.

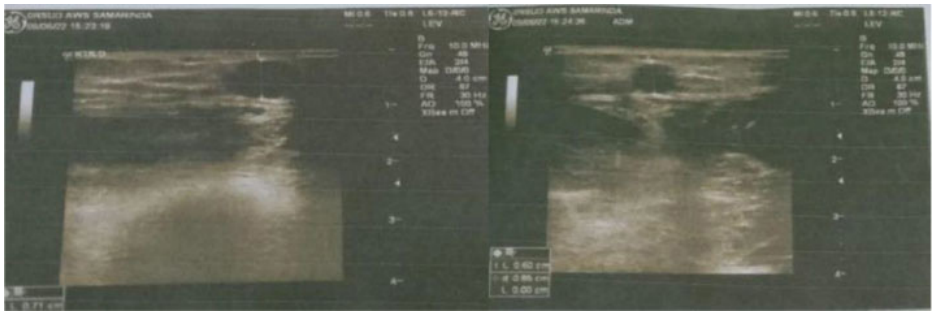

Figure 2. USG vena was the right inferior extremity.

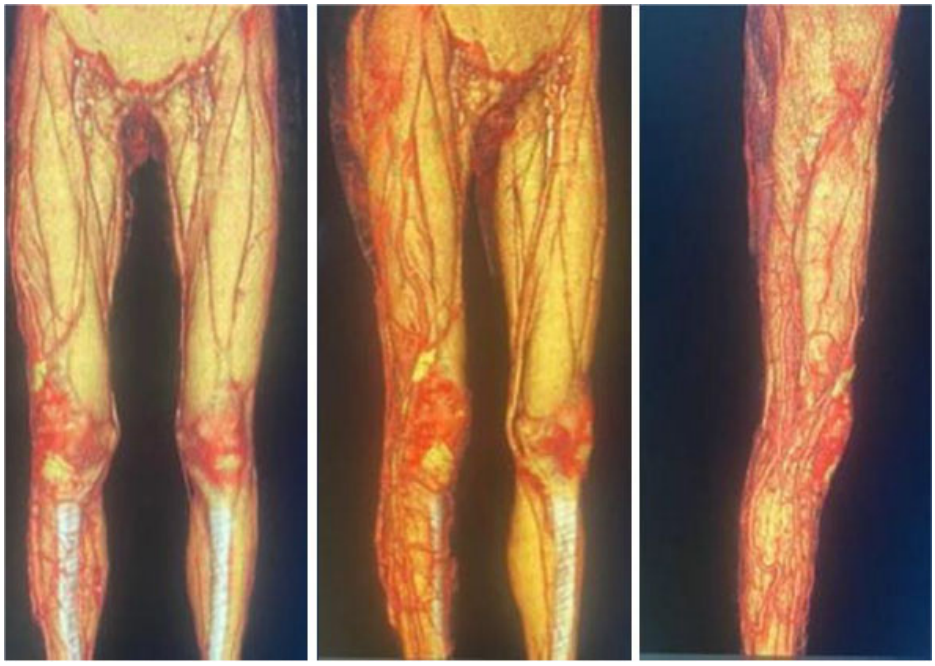

Figure 3. CT Scan angiography of inferior extremity. Hypertrophy inferior extremities and dilatation branches a. superficial femoral and peripheral a. tibialis dextra.

obtained under the ultrasound guide, the J-tip guide wire of 0.0035 inches passed past the hollow needle into the vein lumen. The needle was then removed, and a small incision was made at the guidewire entry point to allow the passage of a 4f laser sheath with a dilator on the wire. The guide wire and freelance dilator were replaced with a laser fiber guided by ultrasound through the sheath to the femoral junction saphenous. Laser fibers make sure the end remains covered in gloves. The procedure can be performed using tumescent fluid under ultrasound direction with multiple needle injections using a foot pump system. The tumescent liquid used in Sodium Chloride Solution is 0.9% at 40°C without local anesthesia. An 18-G needle was used to insert tumescent fluid in a circular fashion until the liquid enveloped the perivenous. The tumescent fluid keeps blood from leaking from swollen veins. The patient's position must be changed back to Trendelenburg during the tumescent administration from distal to proximal so that the blood was not trapped. Tumescent fluid was given more in the femoral junction saphenous area so that the site is compressed better and avoids the expansion of thrombolytic phenomena into the femoral vein or the presence of bubbles passing through the heart.

The patient's position was almost certainly Trendelenburg. The laser setting was adjusted by the type of wavelength and fiber type. This maneuver was associated with the administration of tumescent fluid, which helps to increase the surface area of contact between the laser end and the venous wall and to reduce the amount of blood that can absorb even a small portion of the energy that induces clump formation. Then the laser fiber was pulled back slowly. After the withdrawal to the end of the catheter, evaluated with ultrasound to see the absence of flow and reduced compression in the treated vein.

After EVLA therapy, the patient was advised to wear elastic stockings on the treated limbs for two weeks. The patient was treated for two days in the room with analgesic and antibiotic medications. Patients have no complaints after EVLA therapy.

DISCUSSION

Klippel-trenaunay syndrome is a rare congenital disorder first reported in 1900 by Maurice Klippel and Paul Trenaunay. Klippel-trenaunay syndrome is a rare and complex congenital vascular anomaly defined by the International Society for the Study of Vascular Anomalies (ISSVA) as a combination of vascular capillary malformations (port wine stain), venous malformations (VM), and excessive growth of extremities with or without lymphatic malformations.[2] KTS needs to be diagnosed precisely and early so that it can be distinguished from similar conditions such as Parker Weber Syndrome (PWS), lymphatic filariasis, Beckwith- Wiedemann Syndrome, Proteus Syndrome, Russell-Silver Syndrome, Maffuci Syndrome, Child Syndrome (Congenital Hemidisplasia with Ichthyosiform erythroderma and limb defects, Neurofibromatosis type 1 (NF1), and Triploid Syndrome.[4,7]

This case report found three clinical symptoms of KTS: port wine stain, soft tissue hypertrophy, and varices. Based on Audra A's research Noel, etc., all patients studied (100%) had varicose veins, 65% had port wine stains, and 90% had extremity hypertrophy. Other systemic abnormalities associated with KTS include central nervous system abnormalities with neurological symptoms, gastrointestinal bleeding, and genitourinary lesions.[5,7,9] Another case report from Indonesia reported that the 1,5 years old boy with KTS had enlargement of the veins and complaint of an enlarged left leg accompanied by a sign born on the left thigh. From birth, the left leg looks bigger than the right leg, and there is a birthmark in the form of flat purplish-red patches on the left thighs. Then appear purplish red nodules on the red spot.[10] Similar signs and symptoms were also found in this case report.

According to the KTS Boston Children's Hospital guideline, there is suspicion in the gene PI3K and mTOR, but the gene is still being investigated further[11]. It is said that the cause is due to mesodermal developmental abnormalities that result in increased size and number of veins, leading to increased blood flow in the affected extremities. This opinion is supported by measurements of increased blood flow in the affected extremities, primarily extremities with capillary malformations, although no arteriovenous fistula is detected. Hypothesized mesodermal abnormalities can be regulated by angiogenesis and vasculogenesis, such as vascular endothelial growth factor (VEGF). However, the effect of such hemodynamic factors on extremity hypertrophy is not directly related and is less significant than previously thought. Thus, it hypothesizes that soft tissue and bone hypertrophy are not the direct causes of the static vein in patients KTS.[6,7,11]

Klippel-trenaunay syndrome lacks a definite treatment consensus due to an unknown etiology that is still being investigated. It involves diverse structures and various problems patients face, from abnormal walking posture to psychological disorders and age group differences to the disease. A multidisciplinary team approach is essential for the proper management of KTS patients. The KTS treatment is only symptomatic and does not improve people's quality of life.[4,12,13]

The basic management of KTS is conservative, which only reduces symptoms. Conservative management recommends wearing elastic compression stockings.[13] Elastic stockings help protect the extremities and minimize the trauma that causes bleeding in large superficial malformations. Elastic stockings are also useful for reducing the symptoms of venous insufficiency and lymphedema. But does not affect the size of the extremity.[4,13] Elastic stitching is rarely recommended in pediatrics because of the discomfort it causes in children. Elastic stockings can be combined with other therapies such as lifting legs, physiotherapy, lifestyle modifications, and drug use. Treatments such as analgesics, antibiotics, and corticosteroids are used for patients with cellulitis and thrombophlebitis.[2,4,12]

In surgical management, the success rate depends on supporting examination to see the extent of involvement of blood vessels or related lesions and how the severity of lesions is caused.[2,4] Surgery at KTS is a challenge because the risk of symptoms can be recurrent. KTS, in this case, we use surgery with minimally invasive, i.e., with endovascular laser

ablation with a chance of recurrence and complications of minimum.[11,14] According to the Sung et al. study, most patients received conservative and compression therapy. Meanwhile, one patient received vein stripping and stab phlebectomy, and three samples got an addition to medicine, namely sclero-TX. After receiving the treatment, the patient was followed up for 7 – 108 months.[3] In this case report, the patient underwent endovascular laser ablation procedures. After EVLA therapy, the patient was advised to wear elastic stockings on the treated limbs for two weeks. The patient was treated for two days in the room with analgesic and antibiotic medications.

Endovascular laser ablation has evolved and proven safe and effective in adults for varicose vein management related to superficial vein incompetence. Endovascular laser ablation is rarely reported or is not the main management strategy in pediatric anomaly vascular cases, especially in Klippel-trenaunay syndrome.[1,5,14] Previous research on endovascular laser ablation in KTS cases showed feasibility and safety for pediatric patients. The study explains that of the eight pediatric patients for whom KTS conducted endovascular laser ablation management, seven showed improvements in pain complaints and swelling. In some cases, wearing compression or stocking underwear for 6 weeks after an endovascular laser ablation procedure is advised. Stockings reduce pain and prevent the progressive expansion of vein lesions. In pediatric cases, stockings are only used 5 days after endovascular laser ablation. Pediatric stockings are a challenge because children are uncomfortable wearing such clothes.[1,5,14]

The main complications of this procedure, such as burns, nerve injuries, arteriovenous fistulas, thrombosis due to endothermal heat, and deep vein thrombosis, are rare. Other complications, such as bruising on the maintained lower extremities and swelling after this procedure, can usually occur. Based on previous studies, about 24% of patients experienced bruises on the treated extremities, and 90% of post-endovascular laser ablation swelling occurred. The main complications of KTS are not present in some published pediatric cases. KTS management in pediatrics, currently with low risk and complications, is endovascular laser ablation therapy. Endovascular laser ablation therapy aims to reduce symptoms so that people with KTS can perform activities and improve their quality of life, especially in children.[9,11,14,8,10,12] Fortunately, in this study, the patients have no complaints after EVLA therapy.

CONCLUSION

Klippel-trenaunay syndrome is a rare, complex, and varied syndrome that has not been found yet. Therefore, KTS has yet to have a definite consensus on proper treatment or management plans. An endovascular laser ablation procedure was performed in this study, and wear elastic stockings on the treated limbs for two weeks. The patient was treated for two days in the room with analgesic and antibiotic medications. Patients have no complaints after EVLA therapy.

DISCLOSURE

Author Contribution

All authors contributed to the manuscript preparation until the manuscript was published.

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None.

Conflict of Interest

We have no conflict of interest to declare

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