IMAGING FINDINGS OF LOWER LIMB KLIPPEL-TREUNAUNAY SYNDROME

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PURPOSE

To report the imaging findings of Klippel-Trenaunay Syndrome(KTS), a rare congenital malformation characterized by the triad of capillary malformations, atypical varicosities or venous malformations, and bony or soft tissue hypertrophy usually affecting one extremity. In addition, management options are discussed.

MATERIALS & METHODS

We retrospectively reviewed the clinical characteristics and findings of 4 patients with KTS, including 3 female and 1 male patients, age range between 7 months to 7 years, who underwent assessment at Siriraj hospital, Mahidol University between January 2003 and July 2005. All patients presented with limb hypertrophy and port-wine stain. Imaging modalities including roentgenograms to detect limb length discrepancy, MRI and noninvasive arterial and venous evaluation (MRA and MRV) and imaging during percutaneous intervention are reviewed. Only 1 patient underwent leg angiography and intraarterial gelfoam embolization for subcutaneous hemorrhage.

RESULTS

All three features of KTS including capillary malformations (port-wine stains), varicosities or venous malformations and limb hypertrophy, were presented in all of our patients (100%). Extremity pain was presented in 1 patient (25%). Limb-length discrepancy was presented in 1 patient (25%). Atypical lateral vein was found in 1 patient (25%) without presentation of sciatic vein. No arteriovenous shunt was detected. Spontaneous cutaneous hemorrhages were presented in 2 patients (50%) which in one case needed angiography and intraarterial gelfoam embolization to stop bleeding. Direct Alcohol injection into abnormal superficial vein performed in all patients with successful obliteration of some venous pouches in the 2 patients.[1] The patients who fail to respond to alcoholic injection are those who have no cystic venous compartment. We also achieved to reduce extremity pain, which is occurred in one of our patient. In the literatures, the causes of pain in KTS occurred by many problems, but in these specific patients, the extremity pain is suggested to be due to growing pain. Ours intervention is performed with caution due to we realized that the condition may be worsen if we occluded the dilated superficial collateral vein which function as a normal vein drainage in association with deep vein hypoplasia.[2,3] Lifelong clinical follow-up is mandatory in this group of patients because the natural history of venous and lymphatic malformations is one of a progressive enlargement. Unfortunately our following-up period was not long enough, the long term therapeutic result was still uncertain.

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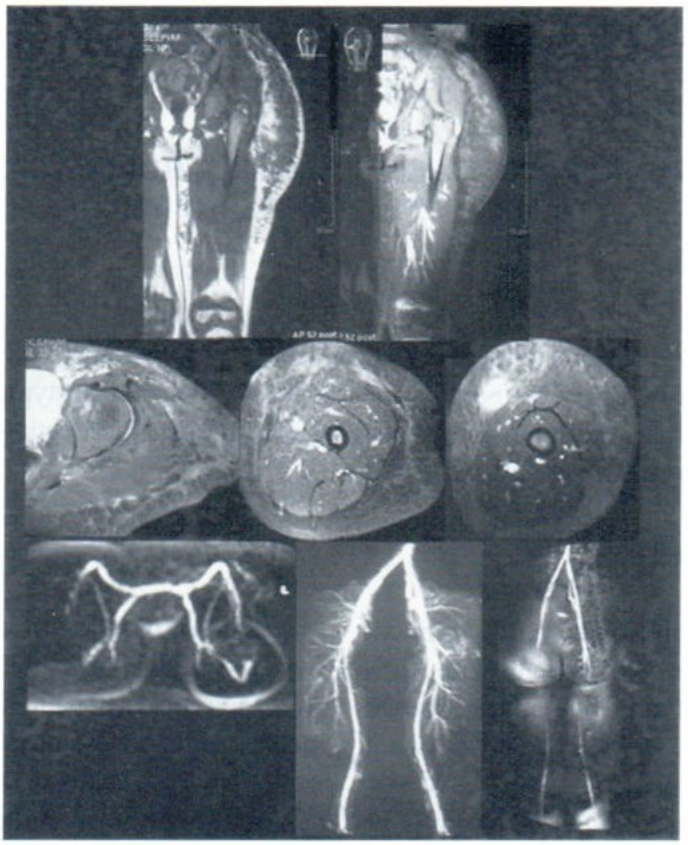


Fig. 1 A 3-year-old female patient presented with progressive enlargement of Lt. thigh and erythematous macule since birth. MRI and MRA Lt. leg showed a soft tissue mass lesion from the level of iliac brim to the above knee region with surpentine flow void structures. Heterogenous soft tissue and vascular enhancement was noted on post Gadolinium injection study. Relative minimal larger of Lt. leg arteries were observed.

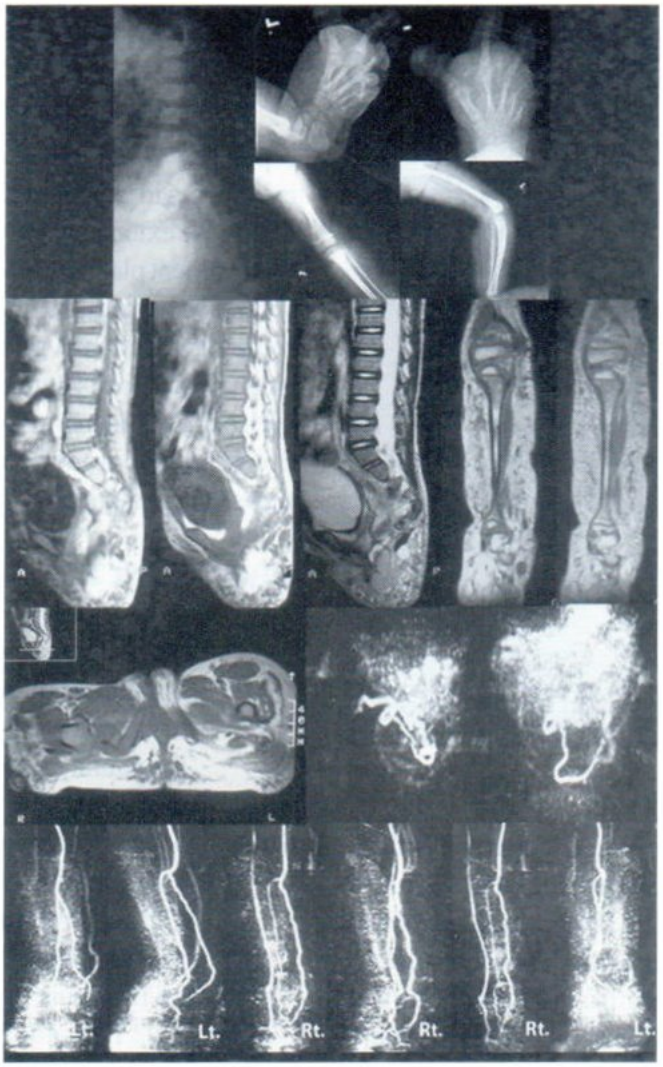
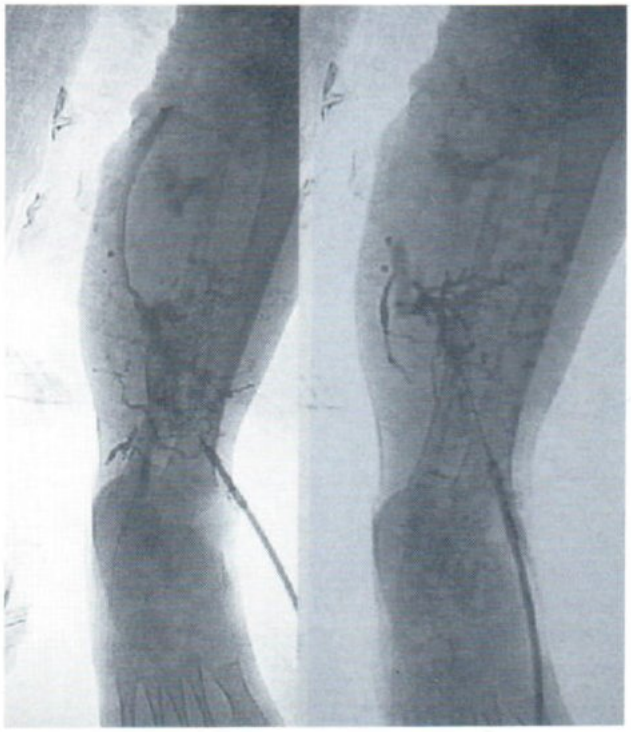


Fig. 2 A 7-month-old female patient presented with Lt. leg enlargement, bilateral club feet and both legs hemangioma with occasional subcutneous bleeding. Plain film of lower back, Lt. leg, Lt. foot, MRI and MRA of both legs showed asymmetry enlarged Lt. leg and deformed Lt. foot with subcutaneous soft tissue enlargement and multiple dilated flow void structures extended from Lt. buttock to Lt. foot. No internal pelvic organ involvement was observed. No arteriovenous fistula was detected. Rt. leg was also affected in a lesser degree.



3 (A) 3 (B)

Fig. 3 A 2-year-old male patient presented with Rt. leg enlargement and hemangioma with occasional bleeding since birth. Imaging during percutaneous intervention; (A) venography, (B) during alcoholic injection showed abnormal dilated veins with atypical lateral vein without presentation of sciatic vein. Intervention was performed with caution.

CONCLUSION

The multimodalities imaging approach especially MRI, MRA and MRV of the abnormal extremities in patient of KTS, allows detailed analysis of disease extension and provide crucial information for treatment planning. Even if most patients with KTS should be managed conservatively, occlusion either by direct injection of sclerosing agents such as direct alcohol injection or surgical resection of symptomatic varicosities or localized superficial venous malformations can give some benefit of relief symptoms for these group of patients.

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