Klippel-Trenaunay-Weber syndrome (KTWS) is a rare congenital disorder characterized by cutaneous hemangiomas, hypertrophy of bone and soft tissue, varicose veins, and clinically significant arteriovenous malformation (AVM) (1, 2, 3). AVMs associated with KTWS are located in the extremities, visceral organs, or spinal canal (1, 3). Most AVMs in the extremities are intramuscular or cutaneous. There is a paucity of data on AVMs in the suprapatellar fat pad associated with KTWS. We report imaging findings of a case with AVM in the suprapatellar fat pad of the right knee associated with KTWS.

**Case Report**

A 25-year-old man presented with a three-month-long history of painful swelling in the right knee. There was knee joint swelling but change in skin color or bruising. Aspirated fluid from the knee joint was bloody. There was no specific family history of congenital disease. Physical examination revealed a brown spot on the right leg, which was apparent at birth, and a cutaneous hemangioma in the right shin and ankle.

The right leg was slightly longer than the left on physical examination and the plain radiograph of lower extremity (Fig. 1). The right iliac and femoral artery was also greater in diameter than the left on femoral arteriogram (Fig. 2). A lower extremity venogram showed dilated and tortuous superficial varicose veins in the right ankle and leg (Fig. 3). There was no thrombosis in the venous system based on both venography and ultrasonography. A venous Doppler sonogram showed severe venous reflux in the right superficial femoral vein, as well as a popliteal vein due to valvular incompetence.

There were multiple dilated tortuous vascular channels in the suprapatellar fat pad on gray-scale and color Doppler sonograms (Fig. 4). We obtained an arterial Doppler spectral waveform within a vessel with spectral
analysis.

MR imaging showed numerous signal voids in the supra-patellar fat pad, in axial T1, sagittal proton density, and T2 weighted spin-echo images (Fig. 5). MR imaging also demonstrated large amounts of joint effusion and thick intrapatellar plica (Fig. 5).

A selective femoral arteriogram showed a dilated tortuous feeding artery and early enhancement of an enlarged draining vein and nidus, indicating AVM (Fig. 6).

Discussion

Klippel-Trenaunay syndrome is characterized by cutaneous hemangiomas, hypertrophy of bone and soft tissue, and varicose veins. When a clinically significant AVM is noted in addition to this triad, the syndrome is termed Klippel-Trenaunay-Weber syndrome (1-3).

Klippel-Trenaunay syndrome is usually unilateral and most frequently affects the lower extremities, but can affect the upper limb, trunk, and head (2, 4).

The cause of KTWS is still not clear. One hypothesis proposed an intrauterine insult during vascular differentiation, with subsequent invasion of the developing limb bud, and other proposed a congenital mesodermal ab-

Fig. 1. Plain anterior posterior radiograph of the leg shows that the right leg is longer than the left.

Fig. 2. Femoral arteriogram showing greater diameter of the right iliac (A) and femoral artery (B) than the left.

Fig. 3. Venogram in the distal leg shows diffuse dilatation of the deep venous system, including distal tibial vein. Note.- varicosity of the distal posterior tibial vein [arrows]
Fig. 4. A. Longitudinal US image at the suprapatellar area of the right knee demonstrates several anechoic tortuous vascular channels in and around the suprapatellar fat pad (asterisk), mainly projecting into the suprapatellar bursa.
B. The same scan of a color Doppler US image reveals numerous enlarged vessels.

Fig. 5. Axial T1 weighted SE image (A) (TR/TE, 700/12) and T2 weighted spin-echo image (B) (TR/TE, 2000/20, 2000/80) shows numerous signal voids (arrows) in the suprapatellar fat pad, mainly projecting into the distended suprapatellar bursa. There was large amount of joint effusion and thick intrapatellar plica in the right knee.

Fig. 6. A. Femoral angiogram shows an abnormal dilated feeding artery (arrows) originating from the right femoral artery.
B. Superselective arteriogram shows a dilated and tortuous feeding artery (arrows) and nidus (arrow heads) in the early arterial phase.
C. Superselective arteriogram shows early draining vein (arrows) with dilated and tortuous feeding artery and nidus in arterial phase.
normality (1, 4). Recently Berry et al proposed the involvement of a somatic mutation for a factor critical to vasculogenesis and angiogenesis in embryologic development (2).

Hemangioma in KTWS are usually located in the skin of the lower extremity, but it can extend deeper to subcutaneous tissue, muscle, bone, and visceral organs, which may lead to internal hemorrhage (3, 5).

Limb hypertrophy develops later in life. It is usually due to subcutaneous tissue hypertrophy, but can associate with bone hypertrophy, which results in leg length discrepancy. A leg length discrepancy of over 1.5 centimeters requires orthopedic correction (2). KTWS has several other osseous manifestations, such as syndactyly, polydactyly, and congenital hip dislocation (1).

A superficial varicosity of the affected limb is a characteristic finding in KTWS. KTWS patients often have deep venous malformations, such as aplasia, hypoplasia, duplication, or abnormal venous valve formation. There is also significant reflux in normal veins. Deep venous malformation is an important factor in deciding to operate on a superficial varicosity. An operation for superficial varicosities is contraindicated in patients with KTWS may be single or multifocal in the extremities, with KTWS may be single or multifocal in the extremities, visera or spinal canal (3). The AVMs associated with KTWS may be single or multifocal in the extremities, visceral organs, or spinal canal (3). The AVMs associated with KTWS may be single or multifocal in the extremities, or may be diffuse and involve the entire extremity and adjacent trunk (1, 3). AVMs in the extremities are usually intramuscular or cutaneous. This is the first report of KTWS-associated AVMs in the suprapatellar fat pad that caused painful swelling of the knee joint and hemarthrosis (1- 7).

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AVMs are usually diagnosed with multiple imaging modalities, which demonstrates a hypervascular lesion with a large feeding arterial vessel (7). Non-invasive imaging modalities such as MR imaging and Doppler sonography can be used for diagnosis of AVMs. On MR imaging, the AVMs appear as a tangle of multiple signal voids, usually without focal discrete soft-tissue mass. The lesions can also be associated with surrounding edema or fibrofatty stroma (7). Doppler sonography of AVMs shows a high velocity, low resistance waveform (6, 7). Arteriography demonstrates a dilated tortuous feeding artery, nidus, and an early draining vein (6, 7).

We confirmed the presence of AVMs by MR imaging, duplex ultrasonography, and angiography. MR imaging showed multiple signal voids around the suprapatellar fat pad, and a Doppler sonogram with spectral analysis demonstrated vessels with arterial waveform, high velocity, and a decreased resistive index in the knee joint. Femoral angiography showed a typical AVM in the knee joint.

Most complications of KTWS are related to the underlying vascular pathology, such as pain, bleeding, thrombosis, or pulmonary thromboembolism. Abnormal vessels in gut, kidney, or genitalia can cause severe bleeding (2).

In particular, the AVMs dilate progressively with age and can result in both local and systemic complications. Local complications are pain, bleeding, tissue ulceration, and impairment of limb function. AVMs can also cause cardiac overload, resulting in heart failure (6, 7). In our case, the chief complication of AVM was painful swelling of the knee joint. There was no color change or bruising, which is usually noted in extremity AVMs. We did not suspect vascular malformation after physical examination because of its location in the suprapatellar fat pad. Repeated aspirations of knee joint fluid during follow up were bloody, so we hypothesized that AVM caused a hemarthrosis.

Treatment of KTWS is conservative and symptomatic (1- 3). For AVMs, surgery is extremely difficult and total removal is rarely possible. Superselective transarterial embolization is the most effective treatment for AVMs (6, 7).

In summary, an AVM associated with KTWS, although rarely located in the suprapatellar fat pad, may cause joint pain and hemarthrosis.

References

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