RESIDENT’S FORUM

Recurrent ulcers on a hypertrophic lower extremity since childhood

Case report

A 68-year-old woman with underlying hypertension came to our clinic for a progressively enlarging ulcer (10.5 × 6 cm) with occasionally active and projectile bleeding on her right dorsal foot for 5 months. The ulcer recurred several times on the same area since teenager years and resulted in indurated and fibrotic skin. Her right lower extremity became progressively hypertrophic with the presence of varicose veins since her childhood. At the time of her visit to our clinic, there were prominent varicose veins, port wine stains, and local warmth on the hypertrophic leg without thrills or bruits (Figure 1). The physical examination was otherwise normal. Laboratory examination revealed no thrombocytopenia or anemia. Bacterial isolation from the wound showed moderate growth of *Pseudomonas aeruginosa*, *Klebsiella pneumoniae*, and *Staphylococcus aureus*. Skin biopsy at the edge of the ulcer revealed extensive dermal sclerosis with deep ulceration and features of venous stasis and lymphedema. Image study by dynamic contrast-enhanced, multiphased magnetic resonance angiography of bilateral lower extremities showed increased vasculature with high-flow vascular lesions at the subcutaneous regions and muscles (Figure 2). Hypertrophy of the subcutaneous tissue and fibrous septa between muscle groups over the right leg and foot was also demonstrated.

Figure 1 The right lower extremity is hypertrophic, and there are prominent varicose veins on the affected limb. A large ulcer is present on the right dorsal foot, and there is port wine stain at the periphery of the ulcer.

Figure 2 Magnetic resonance angiography showed increased vasculature on the right lower extremity. The presence of high-flow arteriovenous malformation was evidenced by rapid opacification of the engorged vessels over the affected limb. The time interval between the three sequential pictures of (A), (B) and (C) is ~40–50 seconds.
Diagnosis

Parkes Weber syndrome.

Discussion

Parkes Weber syndrome (PWS) is characterized by a triad of port wine stain, varicose veins, and bony or soft tissue hypertrophy involving an extremity, plus an arteriovenous malformation (AVM) of the affected limb. Klippel-Trenaunay syndrome (KTS) also consists of the same triad but the vascular malformation is capillary, venous and/or lymphatic without arterial components. In other words, the vascular malformation in PWS is fast flow while that in KTS is slow flow. Due to their clinical similarities, the two syndromes had sometimes been considered together under the term Klippel-Trenaunay-Weber syndrome. The exact prevalence of PWS is unknown due to its rarity. Most cases, including the present case, are sporadic and without relevant family history. The pathogenesis is still not clear and proposed theories include intrauterine damage to the sympathetic ganglia or intermediolateral tract of the spinal cord leading to dilated microscopic arteriovenous anastomoses, and mesodermal defect during fetal development causing maintenance of microscopic arteriovenous communications. The association between KTS and variants of angiogenic factor gene AGGF1 had been reported.

The classic triad of PWS generally affects one single limb, especially the lower limb. Port wine stain is the most common feature with the earliest onset among the triad. Other clinical manifestations in PWS include lymphedema, spontaneous ulcerations with poor wound healing, and susceptibility to local infection over the affected limb as seen in our patients. The AVM in PWS also results in local warmth over the affected area. The high cardiac output associated with AVM may lead to heart failure. Our present case had mild exertional dyspnea after 50 years of age and left ventricular hypertrophy was revealed by chest roentgenogram and electrocardiogram, suggesting an earlier development of heart failure that was not further evaluated by echocardiography. The magnetic resonance imaging study is a powerful tool to reveal both soft tissue hypertrophy and the arterial components in the vascular malformation; therefore, it helps the definite diagnosis of PWS. The management of PWS is usually conservative and symptomatic. In the present case, wound care with 0.3% gentamicin ointment and bed rest resulted in almost complete healing of the ulcer over a 3-month period. Compression garments can be used for chronic venous insufficiency or lymphedema to stabilize the lesion and protect the skin. Vascular embolization immediately followed by surgical excision is so far the most promising surgical option for high-flow vascular malformations in PWS while surgery alone usually results in massive hemorrhage and rapid recurrence. Pulsed dye laser may be beneficial for port wine stain after repeated therapies.

In summary, recurrent ulceration over a hypertrophic lower limb in association with adjacent vascular malformations should arouse the clinical awareness of PWS or KTS. Clinical and radiologic surveys for the presence of arterial components in the vascular malformation help to establish the definite diagnosis of PWS that may be complicated by active bleeding and heart failure.

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