CASE REPORT

Distinctive features of stump volume change in a fresh lower limb amputee with Parkes-Weber syndrome

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SUMMARY

A 23-year-old man with Parkes-Weber syndrome suffered ischaemia on the toes of his left foot with overactive high-shunting. The symptoms were refractory and persistent, and trans-tibial amputation was finally performed; all detectable active lesions that had been involved by arteriovenous shunts were removed. Marked stump swelling was found a couple of times postoperatively when compression treatment was absent. Despite excessive enlargement, conventional compression treatment using elastic bandages was effective in suppressing the enlargement rapidly and in controlling the stump volume. Additionally, 8 months postoperatively, stump enlargement has not recurred using silicone liner as compressive measures. We speculated that marked stump swelling may have been caused by postoperative temporal venous congestion due to the residual varicosities in the stump. Excessive volume change of the stump confused us and stagnated prosthetic rehabilitation. Recognition of such clinical characteristics will facilitate smooth and favourable prosthetic rehabilitation in fresh amputees with Parkes-Weber syndrome.

BACKGROUND

Parkes-Weber syndrome is a rare combined vascular malformation defined by the presence of port-wine stain (cutaneous capillary malformations), arteriovenous malformations with multiple arteriovenous fistulas and limb enlargement.¹ Patients show significant high-flow high-shunt haemodynamics due to arteriovenous fistulas and clinically distinctive complications, such as cutaneous ischaemia and cardiac insufficiency.^{1–2} This condition also results in poor prognosis for limb viability.^{2–4} Limb amputation plays an important role in cases in which the limbs are severely involved and/or in which the condition is life-threatening.^{4–6}

Stump volume may be markedly changeable due to the abnormal haemodynamics. Parkes-Weber syndrome causes many difficulties in daily prosthetic fitting, which is a major concern for amputees.^{6–7} However, few previous studies have discussed stump volume changeability. We discuss the changeability of stump volume in Parkes-Weber syndrome based on clinical experience in a patient undergoing trans-tibial amputation.

CASE PRESENTATION

A 23-year-old man presented with refractory ischaemia on the toes of his left foot, and finally underwent trans-tibial amputation.

He was diagnosed with Parkes-Weber syndrome during childhood. The only symptom was limb discrepancy characterised by bony over-length of the left leg. The right leg was lengthened at the age of 5 years. The patient suffered no serious sequelae, but superficial varicosities and port-wine stain with clearly defined borders gradually appeared on the left foot and crus (figure 1).

At 22 years of age, local bleeding occurred on the toes of his left foot, recurring many times. Ischaemia also appeared around the same region, which resulted in skin ulceration and intractable pain in the toes (figure 2). Conservative treatment, including repetitive embolisation, failed to manage these conditions for over 6 months. Ischaemic pain disturbed his daily activities, including work and sleep. Radiographic examinations revealed a remarkable arteriovenous fistula at the ankle

Figure 1 Superficial varicosities and port-wine stain with clearly defined borders on the lower left leg.

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Figure 2 Skin ulceration on the toes of the left foot.

draining into an ectatic venous sac, and the presence of deep varicosities—the peroneal vein and the posterior tibial vein were markedly dilated, which suggested that occult arteriovenous shunts were also latent along the same veins (figure 3).

DIFFERENTIAL DIAGNOSIS

Clinically, Klippel-Trénaunay syndrome is a very similar condition, which is also a combined vascular malformation defined by the presence of port-wine stain, lymphaticovenous malformations and limb hypertrophy.^{1–3 7} However, Parkes-Weber syndrome can be definitely differentiated by the presence of arteriovenous fistulas, which are always absent in Klippel-Trénaunay syndrome.^{1–3 7} The two conditions show quite different haemodynamics: Klippel-Trénaunay syndrome is a pure low-flow condition, whereas Parkes-Weber syndrome shows a significant high-flow condition.^{1 2 7}

TREATMENT

Left trans-tibial amputation was performed to remove all detectable active lesions: remarkable arteriovenous fistula at the ankle, questionable varicosities (especially peroneal vein) and the whole of the port-wine stain. The fish-mouth method (equal anterior and posterior flaps) was applied for stump plasty. The muscles were sewn over the cut end of the bone and the skin was sutured at the end of the stump. The length of the stump was 12 cm. Severe stump pain and phantom limb pain presented postoperatively. In the early period, the pain severity was rated around 70 with the visual analogue scale (VAS) and taking oral Pregabalin and Duloxetine was mildly effective. The pain diminished to 10 with the VAS 6 weeks postoperatively and the patient took no painkillers after that. The surgical wound was meticulously followed up to avoid wound breakdown.⁶ Despite taking 3 weeks to remove all the stitches, slight bleeding from the wound scar continued. Soft dressing using elastic bandages, the aim of which was to promote maturity of the stump by

conducting uniform compression, was postponed until the bleeding had stopped.

At 4 months postoperatively, compression treatment using elastic bandages was started with soft dressing. The maximum girth of the stump was 29 cm at the start. After starting the daily compression treatment, the stump girth reduced rapidly, reaching 24 cm (ie, a total reduction of 5 cm was obtained within 1 month). Thereafter, the stump girth was maintained under daily compression treatment.

At 6 months postoperatively, a custom modular prosthetic device was made. A total contact socket system using a silicone liner was applied to give uniform pressure on the whole of the stump.⁷ It took 1 month to make the prosthesis, during which time the patient neglected daily compression treatment of the stump.

At 7 months postoperatively, the patient began to report dullness and numbness in the whole stump. On the first day of trying the prosthesis, he was unable to wear it even for several minutes due to intolerable pain when the stump was in the socket. Both fullness and arterial pulsation were determined by palpation on the whole stump (video 1). Careful measurement indicated that the stump had increased in girth by 5 cm compared with that 1 month previously, and the patient was instructed to immediately resume compression treatment. A reduction of girth by 5 cm was obtained within 2 weeks after resumption of compression treatment. The symptoms of the stump (dullness, numbness, fullness and arterial pulsation) had also disappeared. The patient was able to wear the socket without any complaints and to walk well immediately.

OUTCOME AND FOLLOW-UP

The patient was recommended to continue compression treatment whenever the prosthesis was off. However, instead of heeding our instructions, he has spent daily life wearing the prosthesis and/or silicone liner at all times except when sleeping. Two and a half years have passed, and the stump has developed no problematic symptoms and no difficulties regarding socket fitness have been encountered. The patient has been able to continue his ordinary social activities, including working.

DISCUSSION

Robertson⁸ reported 28 patients with Parkes-Weber syndrome including 6 cardiac enlargements, 5 significant leg length discrepancies and 3 required amputations. Our patient had suffered an axial overgrowth of the leg in childhood and an ischaemic condition of the leg as an adult, but he fortunately has not presented any signs of cardiac impairment as yet.

It is difficult to decide on the level of amputation for limbs with vascular malformations.⁷ Patients may function better after amputation than they did before,^{5 7} but total excision of detectable active lesions does not always result in perfect remission of the disease morbidity, and therefore the amputated limb must also be continuously and carefully followed up after amputation.

Stump volume may vary with haemodynamic changes in the vascular content.⁷ Although Parkes-Weber syndrome is a combined vascular malformation, in principle, an ischaemic condition (high-shunt haemodynamics) may be more dominant than a congestive condition (low-flow haemodynamics); that is, the incidence of swelling of the affected limb may be low in Parkes-Weber syndrome. Although our patient had not suffered limb swelling without compression treatment before amputation, excessive stump swelling presented a couple of times postoperatively when compression treatment was absent. However, despite excessive enlargement, standard compression treatment

Figure 3 CT angiography of the lower limbs (anterior view), which shows markedly dilated vessels in the left leg and blood flow defect in the left distal foot.

Video 1 Arterial pulsation of the stump was felt easily on the thumbs when the stump markedly enlarged.

of wrapping the stump with elastic bandages was effective, suppressing the swelling rapidly and controlling girth. Wearing silicone liner also seemed to be effective in controlling stump

volume 8 months post-operatively. The success of these measures suggested that the massive stump swelling was due to venous insufficiency.¹⁻⁷ Preoperative radiological examinations revealed the presence of deep vein varicosities in the affected limb. It was likely that the venous congestive condition became more dominant in the stump after removal of the distal active high-shunt lesion and that the remaining varicosities of the stump caused the distinctive feature of stump volume change. Although we missed performing postoperative imaging studies to address venous dilation on the stump, postoperative physical findings, such as fullness and arterial pulsation of the stump, may also suggest the presence of blood flow congestion.

Despite the seriousness of stump swelling in prosthetic rehabilitation, there have been few reports regarding the associated prosthetic difficulties in patients with Parkes-Weber syndrome. In 1977, Letts⁶ reported 11 patients with “Klippel-Trénaunay syndrome with arteriovenous malformation,” all of whom should have been classified as having ‘Parkes-Weber syndrome’. According to the report, the difficulty of prosthetic fitting was because of recurrent stump swelling in four leg amputees, suggesting that such difficulty may be encountered only

initially, and that the swelling usually subsides within 2 years with the remainder of the amputated limb decreasing in size. The clinical course of our patient was substantially consistent with this description. It is unclear why excessive swelling tendency of the stump may have subsided over time. We speculated that dilation of the venous system occurred secondary to the high-pressure due to the high-flow high-shunt condition (ie, venous hypertension) before amputation, and removal of the high-flow condition from the patient may have promoted restoration of the secondary venous dilation over time.

The majority of clinicians may be confused with the excessive change of the stump girth in fresh amputees with Parkes-Weber syndrome when they first encounter it. However, the clinical course of our patient suggests that standard compression treatment is applicable even though the stump swelling is marked,

and that the excessive condition of the stump tends to subside over time. Recognition of such clinical characteristics may be essential to manage proper and favourable prosthetic rehabilitation in fresh amputees with Parkes-Weber syndrome.

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Contributors TY conceived and designed this work, and representatively wrote the paper. AT and HT planned and performed the surgical treatment (amputation), collected the preoperative clinical and radiological data of the patient and discussed the topic. TY and SY managed the prosthetic rehabilitation, collected the postoperative course about stump girth change and discussed the proper rehabilitative management. TY and AT followed up on the patient to date and collected his middle-term course after amputation.

Competing interests None.

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Learning points

- ▶ Initially, in lower limb amputees with Parkes-Weber syndrome, the stump may swell markedly under the situation without compression treatment.
- ▶ Despite marked enlargement, standard compression treatment for stump swelling may be effective in suppressing it.
- ▶ Venous congestion due to remaining varicosities of the stump is likely a cause of such excessive changeability of the stump volume.
- ▶ Such a condition after amputation may last for months and may subside over time.

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