CASE REPORT

Tissue expansion in the management of Klippel–Trénaunay syndrome in pediatric patients

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Summary Klippel–Trénaunay syndrome (KTS) is a rare congenital disorder that consists of mixed vascular malformations. The management of KTS is challenging and multidimensional. Debulking or excisional surgery may be performed in certain patients with KTS. Tissue expansion is a widely used reconstruction technique in plastic surgery and facilitates the reconstruction of skin and soft tissue defects after the excision of large vascular anomalies. In children with lower extremity KTS, large areas of the involved skin and soft tissue can be removed safely and reconstructed by using expanded tissue. In this paper, we report two cases of pediatric lower limb KTS with large cutaneous involvement and substantial soft tissue hypertrophy around the knee. The placement of a tissue expander was planned preoperatively. For 18 weeks postimplantation, the expander was overinflated to 3–4 times its capacity. Partial excision of the malformed tissue around the knee was subsequently performed. The resultant defect was reconstructed using the expanded tissue. Both patients were postoperatively inspected regularly at our outpatient clinic. Neither patient had surgical or tissue expander-related complications. One year after surgery, there was no evidence of local recurrence. Both children retained full range of knee joint motion without limitations in activities of daily living. Tissue expansion is an effective and reliable method of reconstruction. It may be considered in addition to surgical excision in patients with KTS with substantial cutaneous involvement around the joints. Long-term management and close follow ups are necessary for achieving satisfactory outcomes.

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1. Introduction

Klippel–Trénaunay syndrome (KTS) is a rare congenital disorder that was first described in 1900 by French physicians Maurice Klippel and Paul Trénaunay. The disease is characterized by mixed vascular anomalies composed of lymphatic, capillary, and venous malformations. Klippel–Trénaunay syndrome differs from Parkes Weber syndrome in that it does not entail substantial arteriovenous shunting. The classic triad features of KTS are the following: (1) port-wine stains (i.e., capillary malformations); (2) soft tissue and bone hypertrophy on association with lymphatic malformations; and (3) abnormal varicosities (i.e., venous malformations).

The management of KTS is difficult because of its progressive nature and wide range of disease severity. Treatment requires a multimodal approach comprising conservative therapy, sclerotherapy, laser therapy, and surgery. Each of these approaches has a specific role in the management of various disease components. Performing surgery is controversial, although resection and stripping of engorged veins may be considered for patients with prominent and painful varicosities. Partial excision of the malformed tissue can be performed in certain patients. In this paper, we report two patients with pediatric lower limb KTS in whom tissue expanders were initially applied, followed by partial excisional surgery. Adequate tissue expansions were achieved and facilitated immediate reconstruction of the skin and soft tissue around the knee joint.

2. Case reports

2.1. Case 1

A 5-year-old boy presented to our clinic with a congenital vascular lesion in his left lower limb. For several years, the lesion had progressively grown and his left thigh had become considerably larger than his right thigh. He could walk without discomfort, although he had an abnormal gait. The anterior left thigh and knee exhibited a large erythematous patch with prominent veins; an additional smaller patch was on the proximal thigh (Fig. 1). He also had soft tissue hypertrophy. On the basis of these typical findings, the child was diagnosed as having KTS. In addition, magnetic resonance imaging (MRI) revealed hyperintense signals on T2-weighted images around the anterior and lateral aspects of the left thigh and knee, which measured approximately 20 cm × 8 cm × 2.6 cm (Fig. 2).

To address the large cutaneous lesion, partial excisional surgery with immediate reconstruction was planned. Under general anesthesia, a circular-shaped tissue expander (100 mL) with an internal port system was placed at the superior medial aspect of the knee and adjacent to the vascular lesion (Fig. 3A). Wound drainage was not applied. Gentle compression was instead applied by an elastic bandage. Following satisfactory wound healing and suture removal, inflation was initiated 2 weeks postoperatively. Weekly instillation was performed for the next 18 weeks to a total volume of 410 mL. The tissue expander was removed after 3 weeks. In addition, the large vascular anomaly was partially excised, and the defect was immediately reconstructed using the expanded tissue as a rotational flap (Fig. 3B). The patient’s postoperative course was uneventful. All sutures were removed by the 19th day. The pathology report described a mixed-type vascular malformation consisting of a disorganized mixture of various vascular channels with arterial, venous, ambiguous, capillary, and lymphatic features. Fatty changes and hypertrophy were also noted. Further surgical excision of other small areas of cutaneous lesions, scar revision, and split-thickness skin grafts were performed. Three years after his initial presentation, the child walks with a normal gait and enjoys playing football (Fig. 4).

2.2. Case 2

A 1-year-old boy presented to our clinic with a congenital vascular lesion on his left lateral thigh. He was diagnosed as having KTS on the basis of the clinical features. The boy was managed initially with compression therapy and 10 sessions of sclerotherapy. However, the size of his cutaneous lesion slowly increased. By the age of 3 years, superficial red-brown crusting lesions were visible. This indicated that the capillary and underlying lymphatic malformations had progressed. In addition, the area over his left lower limb exhibited superficial engorged lateral veins. The child received stripping and excisional biopsy of the skin lesions under general anesthesia.

We subsequently began to plan to excise the large cutaneous lesion and underlying malformed soft tissues. We applied a rectangular tissue expander (140 mL) using a similar approach as used in Case 1. Following our protocol, postsurgical inflation was achieved over a course of 18 weeks to a total volume of 420 mL (Fig. 5). After 3 weeks, surgery was performed to remove the tissue expander. At the same time, the malformed skin and soft tissues were
partially excised. The resultant defect, which measured 8.3 cm × 6.0 cm, was reconstructed using the expanded tissue as the rotational flap. Pathological examinations revealed vascular malformation composed of mildly ectatic thin- and thick-walled vascular channels with focal hemorrhages in the superficial and deep dermis. There was no evidence of malignancy. The reconstructed area healed without complication (Fig. 6).

3. Discussion

Klippel–Trénaunay syndrome occurs in all ethnic groups with an equal frequency worldwide. There are no known direct hereditary factors and its etiology remains unclear. Unilateral lower limbs are predominantly affected. Most patients exhibit the classical triad: (1) port-wine stains, (2)
soft tissue and bone hypertrophy, and (3) abnormal varicosities. A definitive diagnosis can be made based on identifying two of the three aforementioned features. In most patients, at least one abnormal finding is noted shortly after birth, whereas the remaining features usually become evident as the child ages. To further investigate KTS, MRI, a noninvasive and nonionizing tool, is typically performed to provide detailed multiplanar images of soft tissue in the affected area. The depth and extent of the malformed tissues and its relationship with the surrounding structures can be clearly illustrated. For all our KTS patients, MRI is the standard imaging tool used to assess the extent of the disease and to conduct preoperative planning.

Multiple treatment modalities are typically used in managing KTS, depending on the clinical presentation of each patient. Fig. 7 shows the simplified treatment protocol that we have used in managing children with lower extremity KTS. In most patients, early conservative management facilitates the control of local symptoms such as discomfort from limb swelling. Compressive therapy reduces local discomfort, limits limb swelling, and minimizes localized intravascular coagulation. We advise all of our patients to wear custom-made compression garments 24 hours per day, except when taking a shower. Sclerotherapy with intravascular or intraliesional sodium tetradecyl sulfate or ethanolamine oleate is useful for inducing inflammation and the subsequent obliteration of abnormal varicosities in venous malformations. Multiple sessions are necessary for different lengths of time (from months to years) to achieve disease control. Certain patients benefit from the surgical stripping off of prominent veins and varicosities in the legs and thighs. Pulsed dye laser treatment is indicated for port-wine stains, but it is only useful in superficial diseases. It may be used as an adjunct for the superficial components of deeper lesions after a successful course of sclerotherapy in capillary malformations.

In patients with KTS that affects the limbs, lymphatic malformations can cause substantial limb discrepancy because of associated bone and soft tissue overgrowth. In addition, lymphatic malformations can be problematic because of recurrent infections. Furthermore, the skin can...
be affected and become fragile and form small vesicles that often ulcerate or bleed. Sclerotherapy has limited success because lymphatic malformations in KTS are microcystic. Performing surgery remains controversial; therefore, conservative management is widely advocated. However, debulking surgery and excision of the involved skin and soft tissue should be considered for certain patients. For postexcisional defects, skin grafts are typically sufficient for reconstruction of the lower extremities. For defects close to the joint, we recommend reconstruction with a local flap by using the expanded skin and soft tissue to preserve joint function.

In 1957, Neumann first reported expansion of the skin to reconstruct a traumatic ear defect. Using a collapsed rubber balloon and an external port, the periauricular skin was serially expanded for 4 months without extrusion or infection. Expansion of the skin and soft tissue may be employed to increase the size of full-thickness skin grafts, local or regional flaps, or distant or free flaps prior to transfer. Tissue expansion has been used to reconstruct tissue in various clinical problems such as for the secondary reconstruction of burn scars, large congenital nevi, and tumor ablation. Reconstruction using the locally expanded tissue enables the resurfacing of large anatomic areas with skin of similar color, quality, and texture.

The application of tissue expanders in children has been widely discussed, and numerous relevant studies have been published. We adopted a tissue expander with a low profile and an internal port system to minimize the risk of infection and exposure. The internal port should be placed away from the expander to avoid accidental puncture of the expander when accessed. The accessibility of the injection port requires careful consideration. In pediatric patients, an easy-access port site facilitates an efficient filling process and helps reduce distress in children and parents.

The shape and size of the expander used are typically based on the estimated dimensions of the defect and the configuration of the surrounding normal skin. Because of

Figure 7 Simplified version of our treatment protocol that we use in managing pediatric lower extremity Klippel–Trénaunay syndrome (KTS). Multiple treatment modalities are used. They can be used simultaneously, depending on the disease severity and clinical presentation of the individual patient.
anatomical restrictions and the limited surface area at the site of placement, we selected the largest possible expanders that could be applied in the children. It is crucial to consider flap advancement and rotation, the subsequent expansion, and donor site morbidity. During placement, care should be taken to avoid damaging the expander. After adequate surgical wound healing, expansion can begin 2–3 weeks postimplantation.

Overinflation of tissue expanders beyond the manufacturer’s recommended capacity seems to be the norm in clinical practice. In one clinical study, the expander was inflated to 3.5 times the manufacturer’s stated capacity without any complications. We have routinely overinflated the tissue expander to 3–4 times the recommended capacity. At the end of the final expansion, we ensured that the expander had sufficient volume for the apical circumference of the dome of skin overlying the expander to be 2–3 times the width of the defect. Removal of the tissue expander and definitive surgery were delayed for 2–3 weeks to ensure that the expanded skin flap was pliable and viable. Previous studies have described various methods for planning tissue expansion.

We did not observe perioperative or expansion-related complications such as infection, expander leakage, premature deflation, and ischemic necrosis of the overlying skin. However, other studies have reported complication rates ranging from 13% to 40%. Surgeons must be aware that patients younger than 7 years tend to exhibit considerably more complications than patients of other ages. This may be associated with the small amount of tissue available for expansion. In addition, preschool children are less likely to cooperate and may accidentally traumatize their expander sites. Complications in the pediatric population tend to occur in the extremities, especially in the lower extremities. We do not place multiple expanders in the extremities because of the high rate of complications. Subsequent expansions can instead be repeated if necessary. Because vascular anomalies such as KTS progress as a child ages, we strongly recommend early management to achieve satisfactory disease control. In general, if a child can safely receive general anesthesia, surgery should be considered for certain patients, regardless of their young age.

4. Conclusion

The management of KTS is challenging and multidimensional. Tissue expansion is a valuable and effective reconstruction method. In this report, both children with lower extremity KTS exhibited large areas of malformed tissue, which were excised. The area around the knee was reconstructed successfully by using the expanded tissue. Both children subsequently continued to receive sclerotherapy for residual venous and capillary malformations. One year after surgery, no local recurrence had occurred. The two cases and their early results that we presented in this paper represent a very small part of our work. Long-term treatment and follow-ups are necessary to achieve ideal final results. Our work on vascular malformations is on-going. In the future, we hope to present a larger series to guide the management of KTS.

References


