



Case Report / Olgu Sunumu

Surgical treatment of Servelle-Martorell syndrome

Servelle-Martorell sendromunun cerrahi tedavisi

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ABSTRACT

Although the etiology and prognosis are not known precisely, Servelle-Martorell syndrome is often a venous, but rarely an arterial vascular malformation characterized by soft tissue hypertrophy and musculoskeletal hypotrophy of the affected extremity. In general, surgery is the choice in restricted conditions for the treatment of vascular malformations; however, it is unavoidable, if complications occur. These vascular malformations are often treated with medically using compression stockings and drugs. Herein, we present a 20-year-old male patient with Servelle-Martorell syndrome treated with surgically due to venous malformation.

Keywords: Angioosteohypotrophic syndrome; congenital venous malformation; phlebectatic osteohypoplastic angiodyplasia; Servelle-Martorell syndrome; surgical treatment; vascular malformation.

Servelle-Martorell syndrome (SMS) is a very rarely seen syndrome which can be confused with other syndromes, such as Klippel-Trenaunay syndrome (KTS), Parkes-Weber syndrome (PWS), and blue rubber bleb nevus syndrome (BRBNS) (Table 1).^[1] It is characterized by soft tissue hypertrophy and musculoskeletal hypotrophy of the affected extremity.^[2] It can be seen in a wide range of lesions extending from isolated subcutaneous vascular ectasia to large volume of lesions that infiltrate the skeleton and bone. These ectasia and aneurysmal enlargements can cause a monstrous appearance in the extremities. In some cases, the deep venous structure anomalies, abnormal venous locations, partial or complete absence of venous valves, deep venous hypoplasia or aplasia can be observed. The general treatment approach to SMS is conservative; however, surgery is partly recommended in case of complications due to the aneurysmal structure or severe shunts. The prognosis of the disease is not clearly known.

ÖZ

Servelle-Martorell sendromu, etyolojisi ve prognozu tam olarak bilinmemesine karşın, tutulan ekstremitenin yumuşak doku hipertrofisi ve kas-iskelet sistemi hipotrofisi ile karakterize, genellikle venöz, ancak nadiren arteriyel vasküler malformasyonudur. Genellikle vasküler malformasyonların tedavisinde cerrahi kısıtlı şartlarda uygulanmakla birlikte, komplikasyon gelişiminde kaçınılmazdır. Bu vasküler malformasyonlar sıklıkla kompresyon çorabı ve ilaçlar ile medikal olarak tedavi edilir. Bu yazıda, venöz malformasyon nedeniyle cerrahi olarak tedavi edilen 20 yaşında bir Servelle-Martorell sendromlu erkek olgu sunuldu.

Anahtar sözcükler: Anjiyoosteohipotrofik sendrom; konjenital venöz malformasyon; flebektatik osteohipoplastik anjiyodisplazi; Servelle-Martorell sendromu; cerrahi tedavi; vasküler malformasyon.

Herein, we present a 20-year-old male patient with SMS treated with surgically due to venous malformation.

CASE REPORT

A 20-year-old male patient was admitted to our clinic with a complaint of leg swelling, starting from the right foot and extending to the knee. The patient reported that the swelling was present only on the foot, when he was born, although it progressed to the knee until now. In his medical history, he was born vaginally on 32th weeks of pregnancy and his mother used an unknown painkiller (metamizole sodium) during pregnancy. On his physical examination, the right lower extremity was larger than the left lower extremity and venous dilatations extended to the anterior and posterior side of right lower extremity, particularly localized at the lateral side of the right lower extremity (Figure 1). The vascular lesions around the knee were partially thrombosed. No birthmark

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Table 1. Differential diagnosis of certain vascular malformations

Disease	Size difference between limbs	Skeleton-bone-muscle hypertrophy	Varicose veins	Dermal lesion/birthmark	Skin hypertrophy	Soft tissue hypertrophy	Arteriovenous malformation	Skeleton-bone-muscle hypotrophy	Ocular lesion
Servelle Martorell syndrome	+	-	+	-	+	+	+/-	+	-
Parkes Weber syndrome	+	+	+	+	+	+	+/-	-	-
Klippel-Trenaunay syndrome	+	+	+/-	+	+	+	+/-	-	+/-
Blue Rubber Bleb Nevus syndrome	-	-	-	+	-	-	+/-	-	-

was present in his body. The right lower extremity was shorter than the left lower extremity and there was limping due to bone-joint deformities. Complete blood count, biochemistry, and other laboratory parameters were normal. The right lower extremity venous Doppler ultrasonography showed no pathology in deep vein structures, saphenofemoral junction, and saphenous vein. Partial chronic thrombosis was seen in the enlarged venous segment in the lateral side of the right lower extremity and venous valves were native (Figure 2). Bilateral lower extremity artery Doppler ultrasonography showed no pathology. Lower extremity X-ray revealed hypoplastic and destructed right tibia and fibula caused by soft tissue hypertrophy and venous compression and the right fibula had a sharp curve in the middle section (Figure 2).

Bilateral lower extremity venous magnetic resonance angiography showed no pathology in the iliac veins, femoral veins, and saphenous veins. The lesion mainly affected the right lower extremity from just above the knee to the foot (Figure 3). To determine the origin of the venous filling, phlebography was performed which revealed independent venous dilatations. After the completion of preoperative evaluations, a written and informed consent was obtained from the patient, and he underwent operation under regional anesthesia. An incision was made from the lesions located on the foot which was marked preoperatively. Distal small venous lesions which was progressively expanding in proximal were removed by tying or/and cauterizing, gradually. The vascular lesions adhered to the tendons and outspread over the bone tissue were removed by cauterization (Figure 4). It was seen that the vascular lesions on the tibia were small, if they were located



Figure 1. (a) Preoperative appearance. (b) Postoperative appearance after three months.



Figure 2. (a) The arrow showing the saphenofemoral valve. (b) The arrow showing the notch on the right fibula.



Figure 3. Magnetic resonance venography showing common venous involvement in right leg.

under the fascia and those were greater, if they located over fascia. Soft tissue-weighted lesions were removed by cauterization and vascular lesions were tied with silk sutures. All of the hypertrophic soft tissue and venous dilatations were removed. No bleeding was observed during and following the operation. The patient was discharged on postoperative Day 2 without any complications. The appearance of the right lower extremity became too close to left lower extremity and there was no recurrence three months later after the operation (Figure 1).

DISCUSSION

There is a limited number of reports on SMS in the literature. According to the previous publications, this malformation is more common in the venous structure than in the arterial structures.^[1-3] However, no data reporting SMS due to arterial malformation are available in the literature. Previous cases were mainly reported from India.^[1,3-6] One case reported in Germany was also indeed from India.^[7] Based on these cases, the lesions were often located in the upper extremity. Vascular malformations are often less common in the population. Venous malformations are more commonly seen as KTS.^[8] Venous malformations are divided into two groups as truncular or extratruncular and low or fast flow based on the anatomical and flow characteristics,

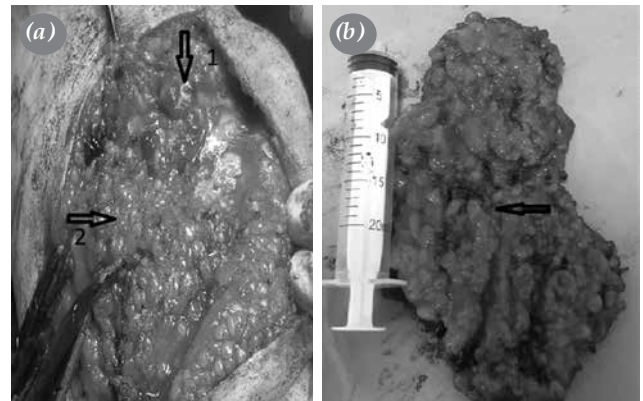


Figure 4. (a) (1) One of the malformed veins on the surgical site is seen. (2) Soft tissue hypertrophy is seen. (b) Tissue fragments of soft tissue and vascular malformation are seen.

respectively. According to this classification, SMS is classified as a low-flow, extratruncular lesion.^[2] It is commonly seen with extremity developmental disorders and with bone deformities, particularly related with bone-related vascular malformations. Gibbon and Pooley^[9] reported a SMS case with a pathological femoral shaft fracture. In our case, fibula would have been likely to be broken later over time. In addition, internal rotation of the patella was observed due to both soft tissue hypertrophy and vascular malformation. The posture disorder and limping were mainly due to SMS. Although guidelines suggest surgical treatment in case of secondary complications,^[2] surgery may be a smart choice in some cases before complications occur. However, the cases should be carefully selected, prepared, and evaluated by a multidisciplinary team including a radiologist, plastic surgeon, orthopedist, anesthesiologist, and pediatric surgeon. Angel et al.^[10] reported that they operated patients with the vascular malformation in pediatric age and successful results were obtained. No recurrence was seen in these cases. In another report, Lee et al.^[11] reported that the first goal of the surgical treatment should be the fixation of vascular malformation and second goal should be the correction of musculoskeletal or other system malformation. However, open vascular surgery and endovascular surgery has not been recommended routinely in vascular malformations due to the low vital risk, high recurrence rates, and possible high morbidity. In the literature, the largest series of SMS (34 SMS cases and 83 KTS cases) were reported by Paes and Vollmar.^[12] According to this report, symptoms were started before one year of age in 62% of the patients. Of the patients, 64% had malformations in the lower extremity and the remaining had malformation in the upper extremity,

and 67% of cases were males. The rate of lesions in the right and left sides was similar. All of the malformation originated from the venous structures. The patients inappropriate for surgery should be treated with elastic compression stock or bandage. An interdisciplinary approach is indispensable in the treatment and follow-up of vascular malformation with an unknown etiology and prognosis. Surgical treatment of this very rare malformation have few handicaps including the lack of clear vision of this disease, possible recurrence, inadequate surgical experience, lack of interdisciplinary centers for this operations and postoperative morbidity. However, avoiding surgery brings some additional risks, including gradually developing other system deformities due to malformations (i.e., skeletal and muscular system, digestive system, and neurological system) cosmetic disorders, stasis ulcers due to untreated lesions and psychological disorders (such as social isolation) shortly, the decision about intervening or not intervening in patient with vascular malformation requires a multifactorial and multidisciplinary approach and it must be evaluated by physicians and family of the patient together.

In conclusion, Servelle-Martorell syndrome is rarely seen and surgery is a rare option, and is not often performed in affected patients. It is recommended to follow-up medically, unless complication develops. If lesion-related complications develop, surgery should be performed following a multidisciplinary team decision and a meticulous preoperative preparation.

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