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Surgical management of vascular anomalies in extremities: a case series



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ABSTRACT

Introduction: Vascular malformations and vascular tumors are the two categories of vascular anomalies in the ISSVA categorization system. Although surgery has historically been the primary method of treating vascular abnormalities, it is increasingly frequently combined with other procedures. Thus, the purpose of this study is to outline the several surgical approaches for treating vascular abnormalities in the extremities.

Case: In this series study revealed six patiens with vascular malformation who underwent surgical treatment. The locations of the lesion more frequently in lower extremity than upper extremity. Few cases need reconstruction using flap or skin graft. Three cases were recurrent hemangiomas which underwent surgical excision before this study. In the follow-up, there were no signs of tumor recurrence after the secondary surgical excision. Four patients had combined therapy with scleroting agent.

Conclusion: Hence, surgical management is constantly the treatment of coice for vascular anomalies. Combined therapy with scleroting agent may use in the case which unable to do expansive excision.

Keywords: ehemangiomas, vascular malformations, vascular anomalies.

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INTRODUCTION

Vascular malformations and vascular cancers, which are still mysteries in contemporary medicine, are categorized as vascular abnormalities. Hemangiomas are regarded as distinct vascular tumors from vascular malformations in the ISSVA categorization system of vascular anomalies.¹ Vascular anomalies are primarily affecting the pediatric group however can abide problematic in adulthood.²

Insideny of vascular anomalies occur in 1-10/100000 people. The most frequently localization are cephalic, followed by trunk, lower and upper extremities.³ Patients with vascular anomalies commonly complained pain, disfigurement or functional impairment. Clinical history and physical examination are two important components of an accurate clinical assessment that should form the basis of the diagnostic strategy for vascular abnormalities.¹

A variety of imaging techniques can be used to identify vascular anomalies related to lesion size, flow characteristics, and connectivity to neighboring

structures.⁴ It is now possible to diagnose vascular irregularities more easily and comprehend available treatment choices, such as medication, surgery, and intervention. Historically, surgery has been a key component in managing vascular abnormalities; however, these days, it is frequently combined with other procedures to achieve the desired result and enhance quality of life.^{2,5} Therefore, this study aims to present the various surgical management of vascular anomalies in extremities

CASES

Six patients with vascular malformations who received surgical therapy were identified in this series investigation. The features of the patients are listed in Table 1. Out of the six patients, four were found to have hemangiomas. Lesions were more commonly found in the lower extremities than the upper extremities. While age has different features, gender numbers are the same. Of the four hemangioma patients in this study who received additional surgical treatment, two experienced recurrences. Few patients require skin grafts or flap

reconstruction. Four out of the six patients that received scleroting agent therapy in combination.

Patient-1

A woman, age 21, reported having a lump on her left hand. The lump had been there since birth and was surgically removed when the child turned four, but it has now returned to the same location. The mass was soft, compressible, and free of any bruit or thrill. It was found on the left hand's medial palmar side. A subcutaneous soft tissue tumor with unclear borders and calcifications was seen on MSCT angiography, which raised the possibility that it was a hemangioma. An 8 x 6 cm mass that had penetrated the muscles and fascia was surgically removed, leaving a large defect on the medial palmar side. An abdominal flap was used to rebuild the defect at the same time. After 21 postoperative days, the flap was separated to allow enough time for neovascularization. Good clinical outcomes were seen at the one-month and one-year follow-up, and there were no clinical indications of a tumor recurrence.

Table 1. Patient characteristics

Num	Age (Years)	Gender	Diagnosis	Location	Management	Combined Therapy
1	21	Female	Recurrent Hemangioma	Left Hand (Manus)	Surgical excision with abdominal flap reconstruction	-
2	19	Male	Arteriovenous Malformations	Right foot (Pedis)	Surgical excision	1% polidocanol
3	20	Female	Recurrent Hemangioma	Right femur	Surgical excision	
4	50	Male	Venous Malformation	Left arm (brachial-antebrachial)	Surgical excision with Full thickness skin graft	1% polidocanol
5	6	Female	Hemangioma	Left foot (Pedis)	Surgical excision	1% polidocanol
6	23	Male	Recurrent Hemangioma	Left Cruris	Surgical excision	1% polidocanol

Patient-2

A male patient, 19, reported a sore mass on the medial plantar aspect of his right foot. The growth had become larger over the course of two weeks. The patient stated that a tiny, painless lump had been present since birth. Upon physical examination, a big bluish, soft, non-compressible, pulsatile mass measuring $\pm 8 \times 5$ cm was palpable. There was more localized warmth and erythema in the cutaneous stain. The patient's residual musculoskeletal and neurovascular exams were within normal bounds. After doing magnetic resonance imaging (MRI) of the foot, it was shown that the dorsalis pedis and arcuatus arteries receive vascularization from the posterior distal tibialis pedis, while the plantar medial, plantar lateral, and arcus plantaris arteries receive vascularization from the tibialis anterior. Nidus or puntat, a hyperintense granule in the center, and a pedis with a draining vein were also visible on the MRI. 1% polidocanol was injected into the mass that could not be excised and was therefore positioned in the muscular component in order to integrate surgical therapy of the malformation with the excision of the AVM in the subcutaneous part.

Patient-3

A female patient, age 21, complained of a tumor in her right femur. Six years ago, the mass—which had been there from infancy—was surgically removed. Following surgery, a histopathological

**Figure 1.** Clinical pictures before operative management**Figure 2.** Imaging of Vascular Malformations

assessment revealed hemangioma-like features. 20% of the bulk was still there after the initial surgery and it grew back. The previous two months have been particularly painful for the mass. Upon physical examination, a firm, ill-defined mass measuring 15 by 10 cm was discovered in the medial region of the

right femur. It was also more pigmented. There was no auditory bruit, no tangible thrill, and no pulsation. A subcutaneous lymphangioma was suggested by the multiloculated cystic mass found in the first third proximally of the right femur during the CT angiography evaluation. The patient had surgery to remove the

lump from a large area of their body. After anatomical pathology was investigated, a hemangioma was suggested as the mass.

Patient-4

A 50-year-old male came with a painless left arm mass that appeared from birth. At first, the size of mass was approximately 1-2 centimeters in length, and increased gradually over the years. He experienced an extreme pain when he was a teenager and forty. Physical examination showed a 27-centimeter mass on the lower left arm with blue-ish appearance, undefined border and decreasing its size when the arm was raised. The mass was soft, compressible with no thrill or pulsatile. CT angiography of the patient revealed low-flow vascular malformation that supports venous malformation in left arm region, with multiple phleboliths and normal arteries. Surgical resection was performed for completely resectable lesions. Incision through the mass exposed vascularized mass with undefined border and multiple phlebolith. The tumour was infiltrated into the subfascia, tendon, muscle, and cutaneous nerve. The wound defect after resection was reconstructed with full-thickness skin graft. In follow-up patient was given 1% polidocanol to reduce recurrency.

Patient-5

A 6-year-old girl admitted with a left foot mass which larger in time. The mass appeared since infant and painless. The mass was located on the dorsum of the left foot. The mass was solid with no thrill, no pulsation and no bruit. Ultrasonography showed soft tissue tumor with size 1,38x2 centimeter which had color flow. CT angiography revealed a subcutaneous soft tissue mass with pool-contrast which connecting to the vascular and calcifications which suggested a hemangioma. Surgical management was performed and found vascularated mass infiltrated to fascia, tendon, musculus and bone surface. Radical excision was held and continued with polidocanol injection in the lesion after the excision. In follow-up after surgery, patient tolerated well, without any complication.

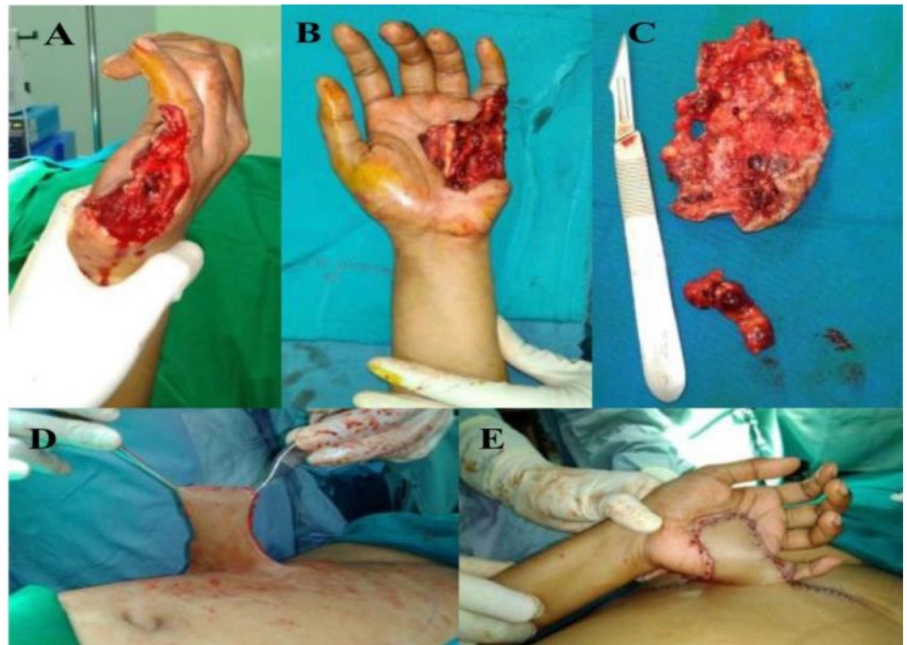


Figure 3. Intra-operative procedure of Hemangiomas (A-C) A mass infiltrating the fascia and muscle was excised. (D-E) The defect was reconstructed with an abdominal flap

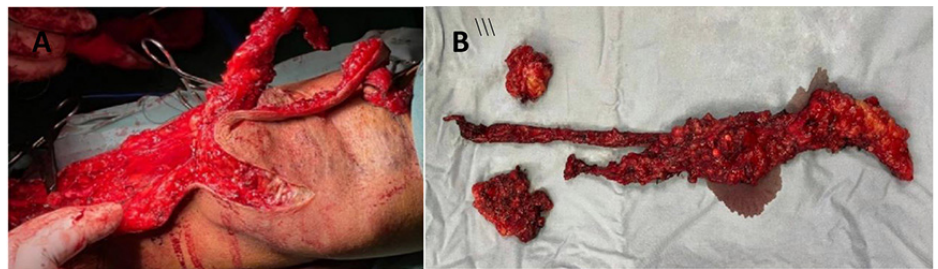


Figure 4. A. Tumor infiltrates into subfascial and muscle, B. Tumor after resection



Figure 5. Post-operative pictures

Patient-6

A-23-year-old male presented a painful mass in the left foot for last two months. The mass was appeared since he was twelve and was repeated excision three times when he was 20th, 21th and 22th. Physical examination was found a mass in the posterior of left cruris with size 25 x 13 x 4 cm, solid, without pulsation and thrill. CT angiography revealed a mass in the one third left cruris that consist of vascular components which impressed a hemangioma. The patient underwent open surgical and found a mass in the posterior of posterior tibial muscles. The mass was excision and continued with 1% polidocanol injection on the lesion area. There was no complain and complication after surgery. In a-year follow-up, there was no sign of reccurency.

DISCUSSION

Mulliken and Glowacki identified vascular anomalies by employing radiography, electron microscopy, and histochemistry to distinguish between two main types of vascular anomalies: tumors and malformations. Endothelial hyperplasia is a feature of vascular tumors, which can vary from frequent infantile hemangiomas to rare malignant neoplasms or uncommon lesions of borderline malignancy. Structural anomalies or inborn defects in vascular development are known as vascular malformations. There are two types of vascular malformations: low flow malformation and high flow malformation.⁶

Clinical history and physical examination data should be included in an accurate clinical assessment that forms the basis of the algorithm used to diagnose vascular abnormalities. Although a complete or clear history is not always accessible, most cases of vascular tumors and abnormalities can be distinguished from one another based on clinical history. Specifically, the distinct mass lesion growth pattern often allows for the distinguishing.¹

The timing of their clinical manifestation, their growth patterns, and the characteristics of their endothelial lining's proliferation in the cell culture are crucial distinctions between hemangiomas and vascular malformations. Even though

Table 2. Classification ISSVA of Vascular Anomalies⁶

Type of Tumor	Classification of Vascular Anomalies
Vascular tumors	Infantile Hemangioma
	Hemangiomaendotheliomas
	Miscellaneous
	Angiosarcoma
Vascular malformations	Low-flow
	• Lymphatic malformation (LM)
	• Capillary malformation (CM)
	• Venous malformation (VM)
	High-flow
	• Arterial malformation (AM)
	• Arteriovenous malformation (AVM)

they might not be visible, vascular abnormalities are always present from birth as inborn defects. These aberrations consistently increase in proportion to systemic expansion. Hemangiomas, in contrast to vascular abnormalities, usually do not exist at birth; instead, they develop later in infancy and expand disproportionately to the growth.¹ Hemangiomas grow rapidly, then undergo a plateau phase and finally undergo involution, but represent wide variation in the rate, duration, and degree of growth and spontaneous tumor regression.⁷

In this case we report six cases of vascular anomalies include four cases of hemangiomas, a venous malformation case and an arteriovenous malformation case. In physical examination, hemangiomas, VM, and AVM has own characteristics. Hemangiomas have red appearance if it involves the superficial dermis and blue-ish or normal appearance if it occupies the deep dermis. The tumor shrinks during the involuting phase of a hemangioma, which starts at around one year of age. The tumor becomes less tense, the vivid color disappears, and the skin pales in the lesion's center. Most children finish their involution by the time they are 3.5 years old. Remaining skin damage, discoloration, telangiectasias, fibrofatty tissue, scarring, or superfluous skin affect 50% of patients.⁶

Because of the increased blood flow in AVM, the lesions may feel warm, pulse, and thrill. They may also appear blue. AVMs develop naturally throughout

childhood, although they can expand quickly due to thrombosis, infection, or hormone stimulation. Expansion can cause local steal phenomena and skin ischemia, which can cause pain, ulceration, destruction of soft tissue and bone, bleeding, and functional impairment. Treatment for these symptoms is required. Next, VMs have a soft, bluish enlargement that is compressible and usually painful. Occasionally, palpable phleboliths without pulse and thrill are present. Although VMs often grow slowly, many lesions may be able to invade widely.^{6,8}

A number of non-invasive imaging methods can be used to characterize vascular anomalies, offering details on the size, flow characteristics, and relationship of the lesion to nearby structures. Computed tomography (CT) enhanced with contrast medium and CT angiography not only aid in the evaluation of bone lesions and phleboliths but also furnish data regarding vascular anatomy, thrombosis, enhancement, calcification, and involvement of surrounding structures.⁹

In the management of vascular anomalies, surgical treatment has long been a major choice, however its role has been replaced with other options including medical and interventional. Surgical treatment in hemangiomas is unnecessary unless indications are compelling. In cases of intractable pain, rapid tumor growth, ulceration, bleeding, and cosmetic or functional impairment, surgical treatment may be considered. The actual timing of

surgery is controversial. In the literature explain that the ideal time for operative management is between 3 and 4 years of age. After 3 years of age, hemangioma will no longer involute significantly.⁶ Surgical excision may be preferred option in Venous Malformations. Surgical excision without any combination therapy can result in excellent outcomes with minimal risk of recurrence, if the lesion is localized. Extensive Venous Malformations are generally not acceptable to resection or are only partially resectable and need pre-operative sclerotherapy. The mainstays management of AVMs are embolization, sclerotherapy, surgical resection, and reconstruction.⁶

In order to reduce the likelihood of recurrence, the surgical goal is total resection, as opposed to phased resection, which is relevant to slow-flow vascular malformations. In certain instances, concurrent intraoperative sclerotherapy has shown effective in treating hemangiomas, an arteriovenous malformation, and significant venous malformations at the moment of surgical resection. The benefits of intra-operative sclerotherapy for the patient are a single operation, lower costs, and more convenience. For the surgeon, intra-operative sclerotherapy gives good delineation and stable hemostasis.¹⁰ Other study reveal intra-operative sclerotherapy allowed surgeons to safely resect large Vascular Malformations with better visualization of dissection and minimizing blood loss.¹¹

Two of cases used reconstruction technique to closed the defects. Patient-1 underwent abdominal free flap to reconstructed the resultant defect. The resultant defect of patient-4 was closed using full-thickness skin graft. The removal of vascular abnormalities on the extremities can be difficult since it is important to remove the tumor as much as possible without compromising normal function. The challenge of soft tissue regeneration for major lesions persists because of the requirement to preserve

their aesthetic appeal while restoring function. Various rebuilding strategies are available based on the patient's overall health as well as the local circumstances surrounding the donor site and wound. Axial pattern flap, free flap, and random pattern flap are a few often employed methods.¹²

CONCLUSION

Surgical management is mainstay the treatment of choice for vascular anomalies. Excision of vascular malformations on the extremities is challenging due to the need to maximize tumor eradication while minimizing damage to normal function. Combined therapy with sclerotherapy agent may use to localize the tumor in the case which unable to do expansive excision. Some surgeries are resulting wound defect which need comprehensive management involving multiple surgical and medical departments.

ETHICAL CONSIDERATION

The patient gave written, informed consent for the publication of this report and any related photos.

CONFLICT OF INTEREST

We have no conflicts of interest to declare.

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AUTHOR CONTRIBUTION

All authors contributed equally to this study.

REFERENCES

1. Lee BB, Antignani PL, Baraldini V, Baumgartner I, Berlien P, Blei F, et al. ISVI-IUA Consensus Document Diagnostic Guidelines of Vascular Anomalies [Internet]. Vol. 34, Epub. 2015. Available from: <https://pubmed.ncbi.nlm.nih.gov/25284469/>
2. Handra-Luca A, Montgomery E. Vascular malformations and hemangiolymphangiomas of the gastrointestinal tract: morphological features and clinical impact [Internet]. Vol. 4, Int J Clin Exp Pathol. 2011. Available from: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3127065/>
3. Judd TM, Thornberry L. When Arteriovenous Malformation Arises In The Foot [Internet]. 2016. Available from: <https://www.hmpgloballearningnetwork.com/site/podiatry/when-arteriovenous-malformation-arises-foot>
4. Noshier JL, Murillo PG, Liszewski M, Gendel V, Gribbin CE. Vascular anomalies: A pictorial review of nomenclature, diagnosis and treatment. World J Radiol. 2014;6(9):677–92. Available from: <https://pubmed.ncbi.nlm.nih.gov/25276311>
5. Dasgupta R. Surgical Management of Vascular anomalies. Vol. 2, Current Otorhinolaryngology Reports. Springer Science and Business Media B.V.; 2014. p. 285–91.
6. Cronenwett JL, Johnston KW. Rutherford's vascular surgery. 2014.
7. Drolet BA, Trenor CC, Brandão LR, Chiu YE, Chun RH, Dasgupta R, et al. Consensus-Derived Practice Standards Plan for Complicated Kaposiform Hemangioendothelioma. J Pediatr. 2013;163(1):285–91. Available from: <http://dx.doi.org/10.1016/j.jpeds.2013.03.080>
8. Donnelly LF, Adams DM, Bisset GS. Vascular malformations and hemangiomas: A practical approach in a multidisciplinary clinic. American Journal of Roentgenology. 2000;174(3):597–608.
9. Hyodoh H, Hori M, Akiba H, Tamakawa M, Hyodoh K, Hareyama M. Peripheral Vascular Malformations: Imaging, Treatment Approaches, and Therapeutic Issues. RadioGraphics. 2005;25(suppl_1):S159–71. Available from: <http://dx.doi.org/10.1148/rp.25si055509>
10. MacArthur CJ, Nesbit G. Simultaneous intra-operative sclerotherapy and surgical resection of cervicofacial venous malformations. Int J Pediatr Otorhinolaryngol. 2019;118:143–6. Available from: <http://dx.doi.org/10.1016/j.ijporl.2018.12.017>
11. Gray RL, Ortiz RA, Bastidas N. Combined Surgery and Intraoperative Sclerotherapy for Vascular Malformations of the Head/Neck. Ann Plast Surg. 2018;80(4):S156–7. Available from: <http://dx.doi.org/10.1097/sap.0000000000001366>
12. Chen H, Chen B, Li B, Luo X, Wu H, Zhang C, et al. Gastrodin Promotes the Survival of Random-Pattern Skin Flaps via Autophagy Flux Stimulation. Oxid Med Cell Longev. 2021;2021:6611668. Available from: <https://pubmed.ncbi.nlm.nih.gov/33505583>



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