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Surgery of venous malformations in the extremities

Galūnių venų raidos defektų chirurginis gydymas

Vytautas Triponis, Birutė Vaišnytė

Vilnius City University Hospital, Vascular Surgery Center, Antakalnio str. 57, LT-10207 Vilnius, Lithuania
E-mail: vytautas.triponis@mf.vu.lt

Vilniaus miesto universitetinės ligoninės Kraujagyslių chirurgijos centras, Antakalnio g. 57, LT-10207 Vilnius
El. paštas: vytautas.triponis@mf.vu.lt

Background / objective

The diagnosis of venous malformations (VM) should be made as early in life as possible. Most of symptoms and signs of this type of vascular dysplasias are taken for manifestation of simple varicose veins or hemangiomas. Therefore surgical treatment can be inadequate and cause further deterioration of peripheral circulation in the extremities. The aim of this study was to elucidate the diagnostic possibilities to avoid mistakes in evaluating the etiology of vascularization disorders and in choosing the right method of treatment.

Patients and methods

The histories of the disease of 32 patients operated on for venous malformations in Vilnius University Centre of Vascular Surgery were analyzed. The methods of treatment performed by general surgeons and the treatment results obtained after establishment of the cause of vascular disturbances were assessed. The following diagnostic procedures were made: ultrasound duplex scan in 32, arteriography in 15, phlebography in 30, MRA in 6, CTA in 4 patients. Surgical treatment consisted of phlebectomy, exstirpation of the marginal vein or ligation of its branches and excision of deep extratruncular venous formations.

Results

Oedema had 26, ulcers 2, and pain was present in 22 patients. There were 17 patients with truncular and 15 patients with extratruncular VM. Twelve patients were operated on by general surgeons without suggesting congenital vascular malformation (CVM). These operations did not improve the peripheral circulation. Stasis decreased or disappeared in 17 (65.4%), ulcers healed in 2(100%), pain alleviation was achieved in 16 (72.7%) patients after surgery performed in the Centre.

Conclusions

The correct method of treatment was chosen on the background of vascular imaging. Operations resulted in palliation or vanishing of symptoms in most patients.

Key words: vascular malformation, symptoms and signs, vascular imaging, surgical treatment

Įvadas/tikslas

Venų raidos defektų diagnozė turi būti nustatyta kuo ankstesniu gyvenimo laikotarpiu. Daugelis šios ligos simptomų ir požymių gali būti priskirti paprasčiausiai venų varikozei arba hemangiomai. Todėl chirurginis gydymas gali būti nevisavertis ir sukelti tolesnį galūnių kraujotakos blogėjimą. Šio tyrimo tikslas buvo parodyti tikslios diagnozės svarbą, kad būtų išvengta klaidų vertinant kraujotakos sutrikimų kilmę ir parenkant tinkamą gydymo metodą.

Ligoniai ir metodai

Išanalizuoti anamnezės duomenys 32 ligonių, operuotų Vilniaus universiteto kraujagyslių chirurgijos centre nuo galūnių venų raidos sutrikimų. Buvo įvertinti operacijų, kurias atliko bendrosios chirurgijos specialistai, ir operacijų, atliktų nustačius kraujotakos sutrikimų kilmę, rezultatai. Atlikti šie diagnostikos veiksmai: ultragarso dvigubas skenavimas (32), arteriografija (15), flebografija (30), magnetinis branduolių rezonansas (6), kompiuterinė tomografija (4 ligoniams). Chirurginį gydymą sudarė flebektomija, marginalinės venos ekstirpacija arba jos šakų perrišimas, giliųjų nekamieninių veninių darinių pašalinimas.

Rezultatai

Edema buvo 26, opos 2, skausmai 22 ligoniams. Kamieniniai defektai buvo 17, o nekamieniniai – 15 ligonių. Dvylika ligonių buvo operuoti bendrosios chirurgijos specialistų nenustačius, kad kraujotakos sutrikimų priežastis – įgimta kraujagyslių liga. Šios operacijos nepagerino periferinės kraujotakos. Stazė sumažėjo arba išnyko 17 (65,4%) ligonių, operuotų Kraujagyslių chirurgijos centre, opos užgijo 2 (100%), skausmas praėjo16 (72,7%) ligonių.

Išvados

Tinkamas gydymo metodas buvo parinktas remiantis kraujagyslių vaizdo metodais. Operacijos palengvino arba visai pašalino ligos simptomus daugeliui ligonių.

Reikšminiai žodžiai: kraujagyslių raidos defektai, simptomai ir požymiai, kraujagyslių vaizdo metodai, chirurginis gydymas

Introduction

Congenital vascular malformations (CVMs) belong to the rarest vascular diseases. VMs make up 70% of all vascular dysplasias in the extremities [1, 2]. In spite of progress in the diagnosis and treatment of this entity, determination of malformation anatomy and assessment of hemodynamic disturbances remain a challenging field of medicine. The diagnostic mistakes are associated with application of incorrect treatment methods and with further deterioration of peripheral circulation. This can lead to significant inability of patients due to progressive oedema, exulceration and gangrene. The Hamburg Classification facilitates evaluation of clinical findings and definition of anatomical and pathophysiological type of vascular malformation [3–5].

CVM can be suspected on the background of physical examination in the majority of cases by external manifestations. It reveals the most characteristic CVM signs: nevus, hyperthermia, phlebectasia, oedema, extremity length difference, ischemia and ulceration [3–5]. Nevertheless, only vascular imaging methods can define the type of malformation and clear up anatomical peculiarities and pathophysiological mechanisms of haemodynamic disturbances which serve as indication criteria to surgery and determine the kind of therapy [6].

Surgical treatment of VM is still considered to be the best method of treatment [2, 6, 7]. It can be combined with embolization or sclerotherapy [7–9]. Pure surgery is especially effective in the treatment of

truncular and extratruncular forms of the disease such as venous aneurysms and marginal vein [4, 7, 10, 11]. Surgical procedures are aimed at reducing the haemodynamic impact of vascular defects. Therefore they have to be given priority in the management of VM. The restorative operations such as veno–veno bypasses are rarely applicable. If they are available ablative surgery removing vascular defects can be combined with shunting operations [12, 13]. Ablative surgery includes excision of dysplastic blood vessels. One of them is the marginal embryonic vein which is very characteristic of patients with VM. This vein can play also a positive role in the haemodynamics. Removal of the entire marginal vein can lead to dramatic consequences. Therefore the exact imaging of this vein is an indispensable condition [14, 15].

Some authors are convinced that complete resection of VMs should be limited to an isolated superficial lesion without extension into the contiguous structure in order to minimize surgical morbidity. They claim that extensive lesion beyond the deep fascia with the involvement of muscle, tendon or bone or extending into the pelvis or gluteal region from the extremity is not ideal for excisional surgery. They recommend embolization therapy or embolo-sclerotherapy as the first line of treatment in these cases [1, 6, 9, 16, 17].

Patients and methods

There were 32 patients operated on in Vilnius University Centre of Vascular Surgery in the period 1983–2005. The age of patients ranged between 7 and 57 years, mean 14.3 years. There were 9 male and 23 female patients in this series. The age of the patients distributed by years as follows: from 1 to 5 – 3; from 6 to 10 – 3; from 10 to 20 – 15; over 20 – 11.

The observation period was from 1 year to 17 years, the mean duration of follow-up being 3.9± 3.8 years. All VM were apportioned according to Hamburg Classification. The following imaging procedures were made: ultrasound duplex scan in 28, arteriography in 15, phlebography in 19, MRA in 6, CTA in 4. Indications for radical treatment were as follows: phlebectasias, progressive venostasis, trophic changes, length difference of the extremities and pain.

The treatment methods used after the anatomical and functional diagnosis were as follows: exstirpation of phlebectasias saving the venous drainage system in 6, removal of the marginal vein in 9, exstirpation of deep subfascial extratruncular dysplastic veins in 17 patients. Operations performed at non-vascular institutions without awareness of the nature of the disease comprised 2 phlebectomies and 10 excisions of imaginary hemangiomas.

Indications for operations were decompensation of the superficial venous system in 28, increasing oedema in 20, leg ulcer in 2 and pain in 22 patients. There were 7 patients with bone and soft tissue hypertrophy present together with phlebectasia. In these patients, surgery of dysplastic veins was done in the hope of diminishing the excess growth of an extremity.

Results of operations were evaluated by following criteria: decreasing oedema, healing of ulcers, disappearance of pain. Evaluation of clinical changes was done during follow-up every 3 months by registering the volume of the extremities, photographing and measuring ulcer size and patient's description of pain. The improvement of peripheral circulation was estimated by the following criteria: pain and stasis disappeared or decreased, ulcers healed.

There were operations performed in other institutions without awareness of the disease nature. The results of operations were evaluated according to the above named criteria.

Results

On the background of clinical findings and as a result of vascular imaging, the following forms of venous dysplasias were determined: of 32 patients, phlebectasia alone was present in 18 (56.5%), in 10 (55.6%) of them the location of dilated veins was atypical. Marginal vein was present in 9 patients. Phlebectasia was combined with other marks of CVM in 14 patients. Ulcers appeared in 2 patients after phlebectomies made by non-vascular surgeons at the institutions of general surgery. Twenty-two of 32 (68.8%) patients had symptoms and signs characteristic of CMS.

Ultrasound, angiography, CTA and MRA revealed the following pathology: in 17 patients with truncular

changes a hypoplasia and aplasia of the deep veins and in primary valve incompetence. Venous extratruncular changes were determined in 15 patients. Four patients had deep dysplastic venous formation infiltrating subfascial structures: in 3 muscles and tendons were involved, in 1 the mass of dysplastic veins came into relation with the sciatic and common peroneal nerve. In 3 patients, dysplasia was located in the lower extremity and in 1 it was observed in the forearm and upper arm. In 1 patient, CTA revealed the dysplastic veins located in the chest wall coming deep into the pleural cavity. Dilatation of the anterior tibial vein and aplasia of the posterior tibial veins were present in 1, hypoplasia and aplasia of the deep veins were diagnosed in 13, primary valve incompetence was diagnosed in 4 cases.

In 9 patients, CVM had been taken for simple varicose veins by non-vascular specialists. In 2 patients nevus and in 8 patients cutaneous manifestation of truncular and extratruncular VM were misdiagnosed and excised as a superficial hemangioma, causing cosmetic defects. In 2 patients, trophic changes appeared and progressed rapidly after phlebectomies.

The results of operations performed at the non-vascular institutions were as follows: stasis increased in 3, ulcers appeared in 2, pain increased in 6 patients. Cosmetic defects after excising cutaneous and subcutaneous dysplastic veins were present in 2 patients.

Case reports

A 23-year-old patient was referred to the Centre in 2005 for phlebectasias of the lower extremities which were noticed at the puberty. Phlebectomy was peformed by general surgeons at the age of 16, causing increasing oedema and ulceration. The phlebectasias progressed, the legs took a typical postthrombotic view: discoloration, lipodermatosclerosis and exulceration on the inner aspect of the ankle. Phlebectasias came out in the suprapubic and hypogastric region as well as on the chest wall. No flow was detected by ultrasound in the inferior vena cava. MRA confirmed the absence of the vena cava beginning from the pelvic veins and up to the right atrium. Only collateral circulation through the lumbar venous system was seen. Aplasia of the inferior vena cava was ascertained. It was clear that further phlebectomies could impair the trophic of the skin in the legs. Reconstructive surgery was not possible, and the compression therapy was recommended.

A 24-year-old patient was consulted for large phlebectasias and superficial thrombophlebitis. The left leg was longer by 4 cm. The superficial veins were most phlebectatic on the whole leg, with significant oedema and bleeding keratoses in the lower part of the leg. *Vena marginalis* was seen strongly enlarged starting at the ankle and submerging into the depth of gluteal subfascial tissues. Dopler investigation showed the reflux through the whole length of the marginal vein. The patient was consulted by many physicians from the age of 6 years, but only compression therapy was advised. The latter was misapplied. Critical decompensation of venous system urged the patient to apply for help to the Centre. Aplasia of the femoral vein was determined by ultrasound scan and the anatomy of veins was specified by phlebography. The patient was operated on and part of decompensated superficial veins was removed, the perforating veins were ligated, some areas of keratosis were excised. The marginal vein remained as the only drainage system of the extremity. A compession stocking CCII was recommended and further step-by-step exstirpation of dilated veins foreseen. The oedema and pain disappeared after the operation.

Table 1. Results of operations performed at the Centre of Vascular Surgery

Symptoms and signs	Total number of operated patients	Peripheral circulation improved N, %	No changes N, %	Worsened N, %
Stasis/edema	26	17 (65.4)	8 (30.8)	1 (3.3)
Ulcer	2	2 (100)	0	0
Pain	22	16 (72.7)	5 (22.7)	0

A 16-year-old girl was admitted to the Centre because of discolouration and phlebectasias of the right forearm. Discoloration spots appeared at the age of 2 years. At the age of 5 years she was operated on by children's surgeons and the most prominent phlebectatic veins in the forearm were removed. After this operation only slight progression of the disease was observed. The outburst of the symptoms and signs of a vascular disease were noticed at the age of 14 years: varicose veins were spreading very fast overall the right arm, the pain was increasing because of the expanding thrombophlebitic process. The phlebography revealed extratruncular dysplastic veins connected with the deep vein system by valveless perforating veins pumping the blood to the subcutaneous venous system and overloading it. The operation was divided into three stages to assure the better healing of the wound in the area of disturbed circulation.

Discussion

The analysis of the results of 32 operations in patients with predominantly venous vascular malformations has shown that 22 patients have had symptoms and signs characteristic of congenital vascular disease which is not encountered in cases of aquired venous pathology. They were: atypically localized and isolated phlebectasias, the marginal embryonic vein, *nevus*, extremity length difference, bone and soft tissue hyperthrophy. Nevertheless, there were 12 operations in the analyzed material which had been performed without the right diagnosis. The operations had been performed taking the pathology for primary varicose veins or for haemangiomas.

The literature tells that the named symptoms and signs serve for diagnosis making at the early age if they come out after birth [3, 5, 6, 12]. These patients should be put on observation or operated on if there are indications for operations: progressive venous insufficiency, exulceration, pain and asymmetry in growth of the extremities. A more complex problem arises when these signs become prominent in puberty or even later, especially if the only sign is phlebectasia. A physician can take these signs for clinical manifestation of chronic venous insufficiency, particularly for primary varicose veins. Twenty patients had CVM birthmarks. The analysis of all these cases showed that general surgeons and general practitioners did not estimate properly the meaning of the birthmarks and missed the possibility to establish the diagnosis of CVM by referring the patients to vascular centers. There had been 12 operations performed for imaginary varicose veins or hemangiomas, and in 20 patients the treatment was applied after a consultation of vascular surgeon. Operations for varicose veins caused worsening of venous outflow in the operated on extremities. Irreversible changes and a great harm can be done especially by exstirpation of the entire marginal vein without determination of its origin and the point of its inflow to the deep vein system. The exstirpation of this vein, which can mimic a laterally situated great saphenous vein, causes serious disturbances of venous blood drainage. The removal of this vein can result in a thorough disaster if there is a hypoplasia or aplasia of the deep veins. It is well known from the literature that the marginal vein or embryonic lateral vein is accompanied by dysplasia of the deep venous system [3, 11]. The analysis of 20 patients who asked for medical advice in a primary care unit at the age of puberty or later revealed that not all of them were referred to a vascular surgeon. The treatment delay can result in increasing lymphostasis and soft tissue changes which eventually become irreversible.

A lesser mischief is made by operations for imaginary hemangiomas which are only a peak of an iceberg, hiding deep extratruncular or truncular dysplasias. Such operations can eventuate at the worst in significant cosmetic defects and continuing or progressing symptoms of pressure to the surrounding anatomic structures (nerves), venous insufficiency, bone and soft tissue hypertrophy or hypotrophy.

The anatomic and functional diagnosis was made by evaluating symptoms and signs and the results of vascular imaging. Ultrasound scanning was the first method of preliminary disclosure of anatomic and functional disturbances in the deep venous system. The most informative diagnostic methods were angiography, MRA and CTA, especially in deep extratruncular dysplasias. Surgery was the method of choice in VM in patients with venous decompensation, leg ulcer and pain. The decompensation of the

superficial venous system was an indication for phlebectomy, exstirpation of the marginal vein or ligation of its branches saving the drainage system which can be detected by vascular imaging methods. The patients with deep infiltrating dysplasia were operated on if the pathology caused pain or disturbed the function of the extremity. Operations in patients with CVM without awareness of anatomical and functional disturbances can lead to deterioration of peripheral circulation and cause inability.

Conclusions

A correct method of treatment can be chosen on the background of vascular imaging. The operations resulted in the palliation or vanishing of symptoms in most patients.

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