ISSN 1392–0995, ISSN 1648–9942 (online)
LIETUVOS CHIRURGIJA
Lithuanian Surgery
2005, 3(3), p. 184–191

Originalūs mokslo tiriamieji darbai

Diagnosis and treatment of predominantly venous congenital vascular malformations in the extremities: a survey of 50 cases

Sutrikusios galūnių kraujagyslių raidos, vyraujant venų patologijai, diagnostika ir gydymas: 50 ligonių tyrimas

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Background / Objective

The rate of congenital vascular malformations is 1.5%. The aim of the study was to analyze clinical manifestations of predominantly venous congenital vascular malformations, to determine indication criteria for invasive treatment and to evaluate its preliminary results.

Patients and methods

There were 50 patients under observation. Symptoms and signs were assessed and their significance in the establishing of congenital vascular malformation diagnosis was evaluated. The following imaging procedures were made: ultrasound duplex scan in 29, arteriography in 15, phlebography in 19, MRA in 3, CTA in 4 patients, lymphography in 1 patient. Surgical treatment consisted of phlebectomy, ligation of the Marginal Vein branches, excision of deep extratruncular venous formations. 29 patients underwent surgical treatment, 14 patients were treated conservatively. Congenital vascular malformations were apportioned according to Hamburg Classification.

Results

The truncular form of the disease was present in 30 patients. The extratruncular form was determined in 20 cases. Phlebectasia alone was a sign in 20 patients. It was combined with other marks in 21 patients. Port-wine stain manifested in 17 (77.3%) patients. In 10 patients phlebectasia and in 22 port-wine stain were noticed as birthmarks. Venostasis decreased after operations in all patients. Excision of extratruncular dysplastic veins resulted in pain alleviation. In 9 patients the disease was taken for simple varicose veins by general surgeons and in 2 patients port-wine stain was misdiagnosed and excised causing cosmetic defects.

Conclusions

The history, symptoms and signs are essential for making the diagnosis of predominantly venous malformations. The correct method of treatment can be chosen on the background of vascular imaging. Surgery was the method of choice in this disease with progressing symptoms.

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

Introduction

Congenital vascular malformation (CVM) remains the most complicated field in vascular surgery. The reasons for this are the rarity of the entity, variability of anatomy and of clinical manifestations as well as confusion in assessment of hemodinamic disturbances. The Hamburg classification facilitated significantly the clinician's work in defining the anatomical and pathophysiological type of CVM, in evaluating clinical signs and the findings presented by non-invasive and invasive diagnostic methods [1–4].This classification is presented in Table 1.

Predominantly venous congenital malformations (PVCM) make up about 70% of all CVM [5, 6]. Since the signs of this type of malformation come out later in life and bear some resemblance to those of primary varicosis, assessment of the whole complex of data, including the history, symptoms and physical marks, is substantial in making an early and correct diagnosis.

Physical examination can reveal the following manifestations: port-wine stain (naevus flammeus, nevus), phlebectasia, Marginal Vein, edema, the difference of extremities in length, ulceration, hypertrophy of soft tissues and bones [4–7]. It is still easier to make a preliminary diagnosis of PVCM when birthmarks exist [5, 8]. The most exposed birthmark is port-wine stain. It is present in somewhat more than a half of the cases of congenital arteriovenous fistulas [8]. But the rate of this sign in patients with PVCM is unknown. Port-wine stain is described as bluish red areas associated with cutaneous capillary dilatation [8]. The rate of other birthmarks in this type of malformation also remains obscure. The pecularities of phlebectasia characteristic of PVCM are most thoroughly described [8–12]. There are always reasons to suspect that phlebectasia is caused by CVM when it is noted early in life, particularly in cases of its unilateral or unusual location [5, 8, 13].

Nevertheless, the choice of treatment still continues to be a great problem. There are several indication criteria for surgical, radiological or combined procedures. For the most part they are also based on physical signs such as phlebectasia, progressive venostasis and hypertrophic changes in soft tissues and bones [14–16]. In patients with a hypertrophy not excee-

Table 1. Hamburg Classification (Belov, 1989)

Type	Form	
	Truncular	**Extratruncular**
Predominantly arterial malformations	Aplasia or Obstruction	Infiltrating
	Dilatation	Limited
Predominantly venous malformations	Aplasia Obstruction	Infiltrating
	Dilatation	Limited
Predominantly lymphatic malformations	Aplasia Obstruction	Infiltrating
	Dilatation	Limited
Predominantly as a-v fistulae characterized malformations	Deep a-v fistulae	Infiltrating
	Superficial a-v fistulas	Limited
Combined vascular Malformations	Arterial and venous without shunt Haemolymphatic with or without shunt	Infiltrating Haemolymphatic Limited haemolymphatic

ding 1 cm and capillary malformation being superficial or/and minimal varicosity, no treatment can be applied. These patients usually do well without any treatment [3, 4, 6, 13]. However, the experience of others in treating patients with CVM shows that operations in early age are more effective and therefore are recommended in spite of slight manifestations of the disease. It refers mostly to a-v fistulae with bone length difference [1, 2, 14, 16, 17]. The operation on the persistent marginal vein speaks also in support of this approach. The step-by-step resection of this vein stimulates the dilatation of the hypoplastic femoral vein ([14–16, 18]. Surgery in cases of PVCM distinguishes itself by a great variety of procedures. It ranges from exstirpation of decompensated veins and step-by-step resection of the Marginal Vein to removal of deep extratruncular infiltrating formations surrounding nerves and arteries [16].

A certain risk of complications and possible functional disorders after operations as well as cosmetic problems which can arise because of bad healing of surgical wounds are restrictive factors for indications to surgery [3, 4].

The aim of this study was to overview the patients with PVCM investigated in out- and inpatient clinic and present the symptoms and signs important for early diagnosis and serving as indication criteria for invasive diagnostic and treatment procedures.

Patients and methods

There were 50 patients under observation. Symptoms and signs were assessed and their significance in establishing congenital vascular malformation (CVM) diagnosis was evaluated. The age of the patients ranged from 8 months to 67 years, the mean age being 22.3 years. The observation period was 1 year for 16 patients, 1–5 years for 19 patients, 6–10 years for 5 patients, 10–20 years and more for10 patients. All CVM were apportioned according to Hamburg Classification. The following imaging procedures were made: ultrasound duplex scan in 29, arteriography in 15, phlebography in 19, MRA in 3 , CTA in 4 patients, lymphography in 1 patient. Indications for radical treatment were as follows: phlebectasias, progressive venostasis, trophic changes, length difference of the extremities and pain. Surgical treatment consisted of phlebectomy, ligation of the Marginal Vein branches, excision of deep extratruncular venous formations. Surgical treatment was applied to 29 patients, 21 patients were put on observation or treated by compression therapy.

Indications for radical treatment were as follows: phlebectasias, progressive venostasis, trophic changes, length difference of the extremities and pain. Surgical treatment consisted of phlebectomy, ligation of the marginal vein branches, extirpation of the marginal vein, excision of deep extratruncular venous formations.

Results

The clinical manifestations and the form of the disease determined by vascular imaging are presented in Table 2.

The truncular form of PVCM was present in 30 patients. Phlebectasia as a birthmark could be confirmed in 10 patients. It was seen in the first weeks after birth. The extratruncular form of phlebectasias was determined in 20 cases. Phlebectasia alone was a sign in 20 patients. It was combined with other marks in 21 patients. Port-wine stain was manifested in 17 (77.3%) of 21 patient with phlebectasia. Port-wine stain alone was present in 2 patients. In all patients this sign was noticed both by pediatricians and parents before the discharge of a newborn from an obstetrical unit.

Hypertrophy of soft tissue and bones was the second sign which was combined with phlebectasia and was observed in 11(52.4%) of 21 patients. Thick leg means only soft tissue hypertrophy not involving the bone and is supposed to be conditioned by venostasis showing up early in life. Hypotrophy of soft tissues and bones was characteristic of extratruncular infiltrating dysplasias.

Phlebectasia came into clinical attention in very different periods of life and rarely was considered to be a sign of CVM until manifestation of chronic venous insufficiency. The time of manifestation of this sign is shown in Table 3.

Thus, it is easy to explain that 9 patients with PCVM were taken as having simple varicose veins and

Table 2. Clinical manifestations and form of PVCM

Clinical manifestations	Predom. Venous N = 50	Truncular N = 30	Extratruncular N = 20
Phlebectasia alone	20	9	11
early in life	10	3	7
with atypical location	10	8	2
isolated phlebectasia	10	1	9
with ulceration	2	2	
without ulceration	18	7	11
Phlebectasia combined with	21	14	7
hypertrophy	4	2	2
port-wine stain	9	5	4
port-wine stain and extremity hypertrophy	6	6	
hypertrophy, port-wine stain and bruits	0	1	
hypotrophy and port-wine stain	2		1
Nevus flammeus alone	2	2	
lower extremity	2	2	
Thick leg	3	2	1
thick leg early in life	1	1	
thickening of the leg in the 2nd or 3rd decade	2	1	1
Hypotrophy alone	1	1	
Hypotrophy combined with port-wine stain	3	2	1

Table 3. Manifestation time and CVI in patients with sole phlebectasia

Age of patients at the moment of vascular surgeon's consultation	No symptoms of CVI	Swelling	Ulceration
From birth to 4 yrs	4	5	0
From 5 yrs until puberty	1	1	2
From the puberty on	5	3	3

were operated unsuccessfully by general surgeons. In addition to this, in 2 patients with port-wine stain hemangiomas were diagnosed and excised causing significant cosmetic defects.

The location of all phlebectasias was atypical. It is shown in Table 4.

The form of PVCM according to Hamburg Classification is presented in Table 5. It shows some morphological characteristic of this dysplasia, description of which was possible only by vascular imaging.

Aplasia and hypoplasia of deep veins were determined in 28 patients. In 17 of them Marginal Vein was present. Dilatation of the deep veins was observed in 2 patients both demonstrating avalvulia of these veins at the duplex scanning and phlebography. The infiltrating form of dysplasia was found in 14 patients. In seven of them only MRA and CTA enabled to localize venous dysplastic formations involving muscles and joints.

Operations were performed in 29 patients. Operations on Marginal Vein were performed in 8, excision of decompensated superficial veins and ligation of incompetent communicating veins in 3, removal of dysplastic venous formation in 16 and decompression of the left common iliac vein by interposition of arterial substitutes into the right iliac artery in 2 patients. In all patients alleviation of symptoms occurred: edema was decreased, pain disappeared, and ulcers healed. The rest 21 patients were put on observation and are under treatment by compression therapy.

Case reports

A 19-year-old girl presented to the out-patient clinic with edema of the left leg. The circumference of the left thigh and calf were by 2.5 cm larger than in the right leg. The swelling had shown up 3 years before and progressed gradually. Ultrasound duplex scan showed no pathology in the deep veins of the thigh and calf, but the left common iliac vein was seen as large as the inferior vena cava. The same findings were presented by phlebography. The operation was performed through a small midline incision. The right common iliac artery was mobilized and lifted by a tape. The size of the left iliac vein decreased significantly. A segment of large saphenous vein was harvested from the right groin. The right iliac artery was cut through and a 2.5 cm vein

Table 4. Location of phlebectasies

Region of location	Number of patients
Marginal vein	17
The thigh (not associated with any saphenous system)	15
Knee joint	16
Ankle	6
Dorsal surface of the foot	7
Upper extremity	3

Table 5. The form of PVCM according to Hamburg Classification

	Form of PVCM				
Type of dysplasia	Truncular	No. of patients	Extratruncular	No. of patients	Total
Predominantly venous	Aplasia Hypoplasia	28	Infiltrating	14	50
	Dilatation	2	Isolated	6	

interposition was made. As there were no signs of a spur, the further reconstructive procedure on the vein was declined. No stasis in the left iliac veins was observed by ultrasound postoperatively. The circumference of the left leg was diminishing gradually and came up to the size of the right leg within 6 months.

A 18-year-old girl was consulted in the out-patient clinic with the similar clinical signs as described in the first case. The swelling had appeared 2 years before. The ultrasound and phlebography data were speaking for a spur in the left iliac vein. An analogous surgical procedure was done, though there was some thickening of the vein wall at the inflow to the inferior vena cava. The interposition of a PTFE graft 2.5 cm in length and 8 mm in diameter was made, and the size of the left common iliac vein decreased. The postoperative ultrasound investigation revealed a slight stasis in the left iliac veins. The patient has been under observation for a period of 9 years. In spite of a successful release of the vein, she needs compression stocking and manual lymphatic drainage to suppress the oedema and maintain normal appearance of the leg.

A 23-years-old girl presented with marked edema of the entire left leg: 4 cm at the thigh and 3 cm at the calf. The edema was gradually increasing within the last four years. At the ultrasound investigation a tumor 6 × 4 cm in the left hypogastric area was displayed. The MRI revealed that the tumor was located at the confluence of internal and external iliac veins. The deep and superficial veins were normal by duplex scanning. At the operation the tumor was circumscribed and enclosed in a capsule. No compression of the pelvic veins was noted. An intraoperative biopsy showed a lymphangioma. The diagnosis was confirmed by postoperative cytological examination. As the edema maintained, a lymphography was made and the obstruction of the pelvic and femoral lymphatic vessels were detected. The further management consisted of permanent elastic support and manual lymph drainage.

A 12-year-old girl was consulted because of pain in the left leg, which was shorter by 1.3 cm than the right one. A port-wine stain was observed on the thigh. No edema was present. Ultrasound duplex scanning showed intramuscular cavernous dysplasia infiltrating adductor muscles in the lower part of the thigh with hypodynamic flow. Phlebography showed axial and intramuscular dysplasia on the lower leg and thigh. The posterior tibial trunk was enlarged and valveless. MRA confirmed the findings and revealed that the intramuscular infiltration was very close to the sciatic nerve and could come into relation with the common peroneal nerve. The girl was operated on and the dysplastic formation contacting with the nerves was removed. The pain disappeared. Still, a suspicion of hypodynamic a-v fistulae remained. As this form of a-v shunts doesn't need selective embolization, the patient was put on observation.

An analogous operation was performed for a 14-year-old girl who presented with pain in the region of knee joint and in the lower thigh. The adductor magnus, vastus medialis and adductor longus muscles were involved in the cavernous hemangiomas. The partial resection of the muscles was performed and the function returned to normal after a short rehabilitation. The girl also became free of pain. Intracutaneous teleangiectasias and small subcutaneous phlebectasias were treated by sclerotherapy.

A 32-year-old man was admitted because of a 40 × 50 cm stain in the left flank (Fig.1) spreading onto the anterior abdominal and the lower chest surface. Varicose veins were seen in this area. They were painful by palpation. The stain was present from birth. He had been operated on at the age of 2 months for the first time and from that moment on underwent 21 operations which were performed by pediatric and general surgeons. An unsuccessful attempt was made to excise the stain which was taken for hemangioma. The patient was first seen in the University Centre of Vascular Surgery at the age of 32 years. A plain X-ray film showed multiple phleboliths. Massive dysplastic veins situated in the abdominal wall and going deep into the muscles and the pleural cavity were detected by ultrasound scanning. CTA showed a very large perforating vein connecting dysplastic formations in the abdominal and chest wall with phlebectasias covering the diaphragm, spreading to the posterior mediastinum and into the paravertebral area. This vein was draining into *v. azygos*. The extratruncular form of PVCM was determined and the dysplastic veins were removed at the operation from the abdominal and chest wall ligating perforating veins, thus disconnecting the communication between the dysplastic veins in the muscles and intrathoracic venous formations. The congestion in the abdominal and thoracic wall and the stain were significantly reduced.

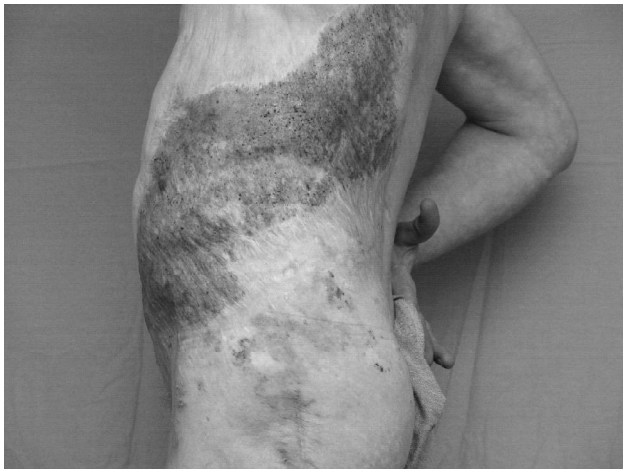

Fig. 1

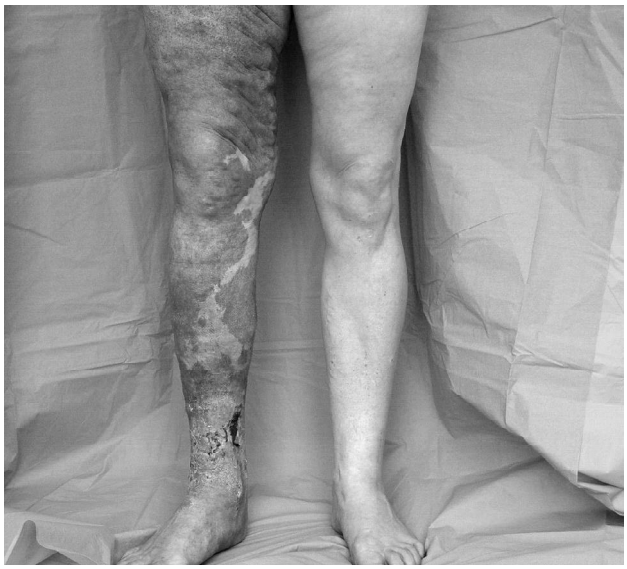

Fig. 2

A 54-year-old female with a trophic non-healing and bleeding ulcer 3 × 5 cm and severe pain was admitted to the Centre. Large varicose veins involving both the thigh and leg were situated atypically and not associated with any saphenous trunks. The history showed that she had had a small nevus of portwein stain type on the dorsal surface of the right foot early in the childhood. The nevus enlarged during the first pregnancy and started to spread to the leg and thigh. Varicose veins appeared almost at the same time and were progressing very fast (Fig. 2). A phlebectomy was performed by a general surgeon who took this pathology for simple varicose veins. The varicosities progressed even faster after this operation. Soon after this operation two trophic ulcers appeared. The first one was localized typically, the other one was situated on the dorsal surface of the foot. The one on the foot reached 5–6 cm in diameter. The ultrasound duplex scanning and phlebography revealed an incompetent Marginal Vein and several perforating veins with reflux. The Marginal Vein was entirely removed and the perforating veins were ligated at the operation. The ulcers healed within the period of 3 months and the pain disappeared.

Discussion

The analysis of symptoms and signs in patients with PVCM revealed the most frequent manifestations of this malformation. They were as follows: atypically

situated phlebectasia, Marginal Vein, soft tissue and bone hypertrophia and port-wine stain (*naevus flammeus*). The rarest ones were soft tissue and bone hypotrophy and port-wine stain as single marks. Phlebectasia as an only sign was most frequent among the signs manifesting separately. There is no doubt that the diagnosis of CVM can be made on the background of physical examination. The question arises whether the external marks alone can reliably serve for picking out PVCM from the whole complex of other types of CVM. Though the most frequent, phlebectasia turned out to be a misleading sign, especially in cases when it had come out as a single mark or later in life. There were 9 patients in whom phlebectasias were taken for simple varicose veins, though the history and atypical location of dilated veins could put the general surgeon on the right track in making the correct diagnosis. The second very important sign of PVCM was the presence of a Marginal Vein which was situated on the lateral aspect of the thigh or leg and was detectable in the standing position. This sign is not pathognomonic for PVCM. It is observed also in patients with congenital arteriovenous fistulas [4, 10–12]. Thus, in patients manifesting Marginal Vein additional tests for arteriovenous fistulas should be carried out to confirm or reject this pathology.

From this study it has become clear that the success in diagnostic is handicapped by the lack of knowledge of the procedures thatwhich constitute an adequate physical examination of a patient. Too little information about the consequences of the disease is also of importance. All patients who have the signs resembling CVM should be consulted by vascular surgeon for evaluation of hemodynamic disturbances and for finding out the type and the form of the disease according to Hamburg Classification. Most of the symptoms and signs detected by physical examination alone are of great prognostic value. Such signs as increasing venostasis, progressing elongation of an extremity, scoliosis and pain predict significant disability and give an apprehension of the necessity to treat these patients by invasive methods. This is supported by the results of other clinicians who have a large experience in the field.

Though the diagnosis of CVM offers no difficulties, in most cases the choice of a treatment method is impossible without a special angiologic investigation. The method of treatment could be chosen only after establishing the type and form of the dysplasia [1, 3, 4, 16]. It means that vascular imaging should be made in all patients with CVM signs. The ultrasound duplex scanning is the first method to be used. It was performed in all 29 operated on patients and showed the pathology presented in Table 5. Phlebography helped in determining the segments of hypoplasia or aplasia of the deep veins showing the site of avalvular zones. It is also of great value in patients with infiltrative forms of venous dysplasia situated subfascially and involving muscles and joints. Phlebography supplemented the ultrasound investigation in 19 patients. Nevertheless, it failed to localize the dysplastic formations in 7 patients in whom CTA and MRA were needed.

The present analysis showed that port-wine stain can be taken for hemangiomas. It is one of the most unpleasant mistakes for patients, because it is fraught with irretrievable harm from the cosmetic standpoint. There were 2 patients who were misdiagnosed and operated on unsuccessfully, trying to excise the nevus without any efforts to determine the underlying pathology.

It is generally accepted that venous decompensation, trophic changes, increasing extremity length difference and pain are the main indications for applying the invasive methods in patients with CVM and in particular in those with PVCM [2, 4, 14, 15]. These criteria were followed while selecting the surgical method of treatment.

Conclusions

The physical examination alone offers an excellent possibility to differentiate congenital vascular malformations from acquired vascular diseases and to urge a specialist to further investigations by vascular imaging. The most informative diagnostic methods were ultrasound duplex scanning and phlebography supplemented with CT and MR angiography. Such clinical signs as phlebectasia or port-wine stain can be misdiagnosed, thus making a harm for a patient with CVM. Surgery was the method of choice in CVM with progressing symptoms. In patients with minimal hemodynamic disturbances, compression therapy is sufficient to prevent the progressing of the disablement.

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