TWENTY YEAR REVIEW OF TREATMENT MANAGEMENT OF VENOUS MALFORMATIONS

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ABSTRACT

Venous malformations (VMs) are the most common of all vascular malformations and they belong to a group of "slow-flow" malformations. The aim of the present study was to analyze retrospectively the outcome of all patients with venous malformations hospitalized and treated at the Department of Pediatric and Adolescent Surgery in Graz over the 20 years period. Twenty five patients were included into this study. Data of gender distribution, diagnosis at time of discharge, results of conducted imaging, information concerning medical history and clinical symptoms as well as invasive and non invasive treatment during the course of disease including the duration of hospitalization and the follow-up have been evaluated. The locations of the venous malformations were in the head and neck in 16%, trunk in 16%, upper extremities in 24% and lower extremities in 44%. The main imaging techniques performed were magnetic resonance imaging with angiography (MRI-MRA) in 51% of patients and ultrasonography (US) in 33% of patients. The preferred therapies in the present study were complete resection in 68%, partial resection 8%, sclerotherapy 8%, lasertherapy 8%, non-operative therapy 8%, or combinations between them. The complexity of the diagnosis and treatment options of venous malformations needs a close interdisciplinary cooperation of experts from different disciplines, to provide the best possible care for these patients. Through the collaboration of experts from different fields an exact diagnosis and best treatment options can be found.

Keywords: venous malformations, surgical resection, sclerotherapy, lasertherapy

INTRODUCTION

Venous malformations (VM) are the most common of all vascular malformations and they belong to a group of "slow-flow" malformations. The main locations of VM are head and neck (40%), extremities (40%) and trunk (20%) and with a proportional gender ratio [1]. VM are usually asymptomatic (depend from size and location) as pain, ulceration, cosmetically deforming, bleeding, abnormal bone growth, fractures, etc. They most commonly involve skin, subcutis and mucosa but they may also infiltrate bone, muscle and other organs [1,2]. Usually they are blue gray in color and grow slowly over time. They can be small and varicose or large and extensive, involving the extremities, face or trunk, but may also involve visceral organs. Common complication of venous malformations is Phlebothrombosis, causing pain, swelling and stiffness of joints and muscles. The location and size is important for the clinical pathology. Venous malformations in the craniofacial region may cause obstructive ocular and breathing complications such as exophthalmia and apnea. Superficial lesions may be cosmetically deforming. The lesions that involve muscle or bone may lose function and result with pathologic fractures [1,2,3].

Diagnostic of VMs

However, the ultrasound cannot penetrate bones or air, so we cannot get more information about deep pathological lesions in the thorax or in the gastrointestinal tract. Doppler Ultrasound can give qualitative data to define the type of vascular malformation and furthermore enables the distinction from Hemangioma’s, by obtaining blood flow characteristics [2,4]. In venous malformations it is very important to detect segmental and long refluxes in the deep and superficial vein systems. Ultrasound can be extremely useful during sclerotherapy to follow the distribution of the sclerosing agents. MRI is more sensitive than CT in detecting venous malformations and their imaging
manifestations shows: other internal body structure, fat infiltration and dilated veins associated soft-tissue masses. MRI can provide important results about the quality of flow in the vessels showing a detailed vascular anatomy. Presence or absence of flow voids can differentiate high - flow from slow – flow lesions. MRI is the leading imaging modality in the diagnosis of vascular malformations. The main information is gained from a combinations of T1-weighted, fat-saturated T2- weighted and gradient-echo flow-weighted MR images [5,6].

Treatment of VMs

For successful treatment of patients with congenital vascular malformations, it is important to remove the vascular malformations or the hemodynamic disturbance caused by them. The removal of vascular malformations may be or may not be radical, therefore it is necessary to create a clear treatment plan and define a goal for each treatment step. From all different treatment options the least invasive and simplest should be selected [2]. Based on the type of vascular malformation, treatment may include laser therapy, sclerotherapy, embolization and/or surgical resection. These procedures can be performed as a single treatment modality or in combination with each other [7].

The techniques of laser application in vascular malformations aim at destroying the pathologic vascular structure. In capillary malformations, laser treatments are the first choice of therapy, whereas in venous and lymphatic malformations laser therapy is a supplement to surgical excision and sclerotherapy [2,8]. Lasers can be used for photoagulation, vaporization or excision procedures, like Nd:YAG 1064 nm laser (neodymium- yttrium- aluminum – garnet), KTP laser 532 nm (potassium–titanium–phosphate), diode laser 800– 980 nm, pulsed dye lasers 585 and 595 nm, argon laser 514 nm and carbon dioxide (CO2) laser 10,600 nm [9].

Sclerotherapy is a form of nonsurgical intervention, used to treat venous malformations by injecting substances such as ethanol, polidocanol and sodium tetradeeyl sulphate into the malformed vessels. Injection of a sclerosing agent (liquid) into the area of venous deformity produces a chemical irritation of the endothelial cells. Then a thrombus is produced and the vein is finally transformed into a fibrous thread that ultimately disappears [2]. Surgical treatment is a common method to treat venous malformations. Often surgery can be applied alone as the only procedure, however it can also be applied together with sclerotherapy. The best option for venous malformations is treating them with surgical resection [2,12]. The venous malformations may be difficult to treat by surgical resection when they are located deep and involve vital structures, such as in craniofacial region, pelvis or other visceral organs.

Venous malformations of the limbs are treated with elastic compressions stockings and with anticoagulants medication to reduce thrombosis.

MATERIAL AND METHODS

A retrospective study of 25 patients treated during the period of 1991–2010 was performed. All male and female children with venous malformations, who have been treated at the Department of Pediatric and Adolescent Surgery in LKH Graz, were including into this study. During the acquisition of the data these parameters were analyzed: the discharge diagnosis, the clinical presentation, the results of conducted imaging, as well as the outcome after the operative or non- operative therapy. Data were collected from the open MEDOCs (Medical Documentation and Communications System) and from the KIS system. Data were typed into a sheet and descriptive statistical analysis were performed: gender (female or male); symptoms and signs: swelling, pain, cosmetic disturbance, impaired function, no symptoms, skin efflorescence; location: head and neck, trunk, upper extremities, lower extremities and generalized; imaging modalities: US, X-ray, MRI, CT; treatments modalities: lasertherapy, sclerotherapy, partial resection, complete resection and non-operative therapy or combinations between them; recurrence after the treatment.

RESULTS

The gender distribution of (13) female to (12) male in present study is is proportional, is quite equal.

Anatomic distribution

The locations of the venous malformations were in the head and neck in 4 patients (16%), trunk in 4 patients (16%), upper extremities in 6 patients (24%) and lower extremities in 11 patients (44%).
The most common localization of venous malformations was in the lower extremities, followed in upper extremities.

**Symptoms and signs**

The main symptoms were swelling and pain, followed by other symptoms such as cosmetic disturbance, impaired function and skin changes were less frequent.

As we can see from the Fig.2 the two most common early signs and complications of venous malformations were pain and swelling. The rarest treatment indications were cosmetic disturbance, impaired function and skin efflorescence.

**Imaging modalities**

Fig. 3 shows the variety of imaging techniques used in the frame of the evaluation process to diagnose venous malformations. In patients with VMs it was necessary to perform various diagnostic methods. However, the best diagnostic option is magnetic resonance imaging – angiography (MRI-MRA) followed by ultrasonography (US). In complex cases it was important to perform combinations with other imaging modalities such as CT and X-rays.

**Treatment modalities for VMs**

The most common treatment type was surgical resection represented with 68%. This therapy has taken first option place and was conducted on all venous malformations. Other treatment modalities were partial resection, non-operative therapy, laser therapy alone or in combinations with sclerotherapy and partial or total resection.
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figure 4. Treatment modalities for venous malformations

In 25 patients with venous malformations, different treatment methods were performed. The main therapy was surgical resection, followed by sclerotherapy, non-operative, laser therapy alone or with partial or total resection.

Recurrence after the treatment

According to the follow-up data 56% of cases were successfully treated. After they were treated, 44% of patients got recurrence, and another treatment was necessary.

Figure 5. Eleven patients got recurrence after the treatment

DISCUSSION

Vascular malformations present a complex group of inborn errors of vasculogenesis and/or angiogenesis which in grave cases can be suspected prenatally by ultrasound and/or prenatal MRI. Under the term “vascular anomalies” all vascular diseases, i.e. various vascular tumors and all various vascular malformations have been grouped. While vascular tumors may have different timing of appearance from the prenatal, over the postnatal period, up to old age, vascular malformations are usually present at birth, and may seldom occur later in life such as during posttraumatic repair of tissues after injuries, or after surgical intervention for treatment purposes like dialysis. First of all it is important to differentiate vascular tumors, from vascular malformations. The congenital vascular malformations are divided into slow-flow malformations (capillary, lymphatic, venous, or combined), and high-flow malformations (pure arterial, arteriovenous fistulas, arteriovenous malformations). Besides these there is further a group of complex combined syndromes with all their varieties, which not only have a component of a vascular malformation, but also other pathologic findings like: epilepsy, lipomas, overgrowth, etc. [1,2,6,10].

Concerning their extension and growth pattern venous malformations may be localized or infiltrative. They can furthermore occur anywhere in the body, but their most common occurrence is especially in the upper and lower extremities. Anatomic distributions of these lesions in the present study were in the neck in 16%, trunk in 16%, and all together in the upper and lower extremities in 68% of all patients. The most common symptoms in patients with venous malformations were swelling and pain. Other symptoms were less frequent in the patients in our study group. In the present study the clinical diagnosis has been further evaluated by various imaging modalities. The main imaging techniques performed were magnetic resonance imaging with angiography (MRI-MRA) in 51% of patients and ultrasonography (US) in 33% of patients. Other imaging modalities such as plain x-ray films or CT imaging were used in patients when more detailed imaging of the lesions was necessary and osseous involvement was suspected. Radiography helps to show organomegaly, osseous destruction and phleboliths, and CT may provide further information about the exact vascularization, and shows better calcifications and the skeletal involvement [2,11,12,13]. The preferred therapies in the present study were complete resection, partial resection, sclerotherapy, laser therapy, non-operative therapy, or combinations between them. VMs are difficult to treat and usually it is necessary to apply
more treatment modalities. The preferred treatment modality was complete surgical resection in 68% of patients, followed by sclerotherapy, partial resection, laser therapy and non-operative alone or in combination between them. Due to the dangers associated with sclerotherapy when the VMs involves vital structures like nerves and any others functionally organs, and when the symptoms cannot be treat with non-operative/conservative therapy then as an important therapeutic choice is surgical resection.

**CONCLUSION**

The present study confirms that the complexity of the diagnosis and treatment options of venous malformations need a close interdisciplinary cooperation of experts from different disciplines, to provide the best possible care for these patients. Through the collaboration of experts from different fields such as pediatric surgeons, plastic surgeons, vascular surgeons, oral and maxillofacial surgeons, orthopedic surgeons, pediatricians, dermatologists, radiologist, ear-nose-throat surgeons, ophthalmologists and others, an exact diagnosis and best treatment options can be found.

**REFERENCES**