Surgical treatment of hemangiomaS and arteriovenous malformationS in upper extermity

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Hemangiomas belong to one of the most common type of tumors occurring in childhood. They are benign and often do not require any treatment, with the tendency to frequently disappearing spontaneously at the age of 5-7 years. The tumors usually develop in the head and neck area, whereas vascular malformations (AVMs) are rare congenital anomalies, which may occur anywhere. In upper limb, they may amount to 30-60% of cases. It is common for these changes not to require any surgical treatment. In case they do, the main indications for such treatment of upper limb AVMs may be as follows: signs of compression of the peripheral nerves, limited mobility of the fingers because of the tumor, which entails difficulties in the functioning of the whole arm, pain and aesthetic considerations.

Aim of the study was to present the results of surgical treatment of vascular tumors (hemangiomas and AVMs) in the upper limb that was applied in our department, as well as a broader discussion on epidemiology, diagnosis, and treatment of these changes.

Material and methods. In 2003-2011 period we treated 11 patients aged 24-39, suffering from vascular tumors (hemangiomas / vascular malformations) in the upper limb; the group included 3 men and 8 women. In this group we diagnosed 2 cases with arm hemangiomas, 2 cases of intramuscular hemangiomas of the forearm, 2 cases of hemangiomas in metacarpal area, 2 cases of AVMs of the finger and metacarpal area, and 4 cases of isolated vascular malformations (mainly arteriovenous malformations) of the fingers. All preparations were examined histologically.

Results. No complications or recurrence were found in 7 cases, recurrences of AVMs were observed in 4 patients with finger changes. In one patient with AVMs of the index finger, we observed a treble recurrence, which severely impaired functioning of his hand. We eventually suggested to him the finger amputation, to which the patient agreed.

Conclusions. Upper extremity is a rare location of hemangiomas and other vascular malformations requiring surgery. hemangiomas are more common in women, and the occurrence of AVMs in both males and females is similar. Surgical treatment of hemangiomas and AVMs of the upper extremities is doubtlessly an arduous task, which calls for great microsurgical skills.

Key words: hemangiomas, vascular malformations, arteriovenous fistula, surgical treatment.

Vascular changes are characteristic of childhood. They may already be present at birth (vascular malformations) or come a little later (hemangiomas). They may occur in any part of the body and in any organ. The presence of vascular changes of upper limbs makes it difficult for patients to accept them due to clinical and aesthetic reasons. Since there is a compression possibility of neighboring elements, e.g. vessels or nerves, this will be an indication for surgical treatment. In this paper we have put forward results of surgical treatment of the afore-mentioned changes and discussed all subject related problems.
MATERIAL AND METHODS

In the period January 2003 - November 2011 in our department we performed operations on a group of 11 patients stricken with vascular changes in the upper extremity (tab. 1), which included 3 men and 8 women. Hemangiomas were found in 6 patients, while in the other 5 we observed vascular malformations, mainly of distal or peripheral type. Our material shows that in case of hemangiomas, the ratio of female to male was 5:1, and in case of vascular malformation, the ratio amounted to 3:2 – which is to some extent a reflection of the results at which other papers have arrived when discussing treatment methods of these changes. A clear indication for surgical treatment of hemangiomas were signs of compression of the nerves, with lack of laser and/or radiological treatment of the neighboring area, or their failure.

As far as vascular malformations are concerned, the main indication for surgery was pain and aesthetic considerations. Limited treatment options for these changes along with less invasive methods have been associated with the distal or peripheral position. All preparations were sent for histopathological examination, which confirmed our clinical and macroscopic examination.

RESULTS

As long as vascular tumors are concerned, we did not observe any recurrence of hemangiomas. What’s more, the symptoms resulting from compression of the peripheral nerves have completely disappeared, while in vascular malformations cases, recurrences were found in four patients. In majority of cases, the size of recurrent vascular malformation was clinically insignificant. In one case, though, the recurrence was so significant both from clinical and aesthetic perspective that reoperation was considered as necessity. Another recurrence in this patient was observed two years later. The size and location of the tumor had a great influence on functioning of the whole hand, yet another operation of the same finger would pose a high risk of ischemia or necrosis. Following our consultations with the patient, he agreed to have the finger amputated. In addition, after surgical treatment of vascular malformations and finger metacarpal, we discovered a complication in the form of necrosis of the distal ring finger (fig. 1, 2).

DISCUSSION

Liston presented his findings regarding hemangiomas for the first time in 1843 (1). While majority of authors believe that they occur more frequently in females than in males, with the incidence of 3-4:1, all authors agree about one thing: that hemangiomas and vascular malformations are benign tumors. Hemangiomas are the most common tumors in children, proliferating during infancy and subject to spontaneous disappearance over the period of childhood. According to Mulliken and Glowacki (2) these tumors tend to occur within the first 4 weeks of life and are described by a characteristic life cycle. The authors claim that only 30% of hemangiomas are present at birth, while the vast majority (70-90% of cases) occur at 4 weeks of age. The first stage of development cycle is very fast and takes about 10-12 months. During this time hemangiomas change color from reddish purple to light red or light blue, just to turn dark red or purple at the second stage. In the third and final stage of development, tumors get contracted and start to fade away at the age of 5-7. Consequently, they decrease and often disappear. The thing that may remain is skin changes, though, including telangiectasia.

Hemangiomas are tumors histologically characterized by high metabolic activity, with an increasing replacement of endothelial cells, mast cells, fibroblasts and macrophages during proliferation manifested by changes in their size, shape, color and cohesion. The second

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stage of development begins in patients who are a couple years old. Here, due to a reduction in metabolic activity of the tumor, it gradually disappears (3, 4).

Hemangiomas can be confused with vascular malformations and even sarcomas because of the rapid growth phase. The best diagnostic method of hemangiomas is magnetic resonance imaging (fig. 3) or angiography, which also provide basis for differentiation between vascular malformations and sarcomas (5). An additional ancillary diagnostic examination may be carried out by either ultrasound with color Doppler function or CT scan with contrast, or both. On X-rays, regular hemangiomas can be visualized as tumors of soft tissue, to which calcifications are often to be presented.

In general, most hemangiomas do not require any treatment. Surgery is restricted to cases when clinical symptoms are associated
with the presence of hemangiomas. These symptoms include:

a) oppression by a tumor on the peripheral nerves and the associated paralytic symptoms (fig. 4),
b) restriction of mobility, e.g. occurrence of tumor angioma in the vicinity of fingers joints (fig. 5),
c) pain associated with hemangioma,
d) aesthetic reasons related to the size of hemangioma,
e) skin ulceration and secondary infection because of excessive compression of the skin by hemangioma (fig. 5) (6, 7).

The AVMs are congenital anomalies, which are rare and which can occur at any location.
Surgical treatment of hemangiomas and arteriovenous malformations in upper extremity

For the upper limb, they may be presented in 30-60% of cases. Macroscopically vascular malformations are abnormally widened conglomerates of arteries and veins, often accompanied by abnormal vascular fistulas. They can be arterial, venous, capillary, lymphatic, or mixed. The most often employed classification of vascular malformations is the one elaborated on by Mallan and Puglioni (9, 10). They classified the malformations as vascular malformations with dysplasia – a physiological formation with vascular structural changes, and to these with abnormal formation in a normal looking tissues. In contrast to hemangiomas, vascular malformations are present at birth and grow proportionally to their development, never disappear spontaneously and their incidence by gender is 1:1.

Vascular malformations are histologically similar to changes in non-neoplastic tumors, and develop as a result of abnormal vascular tissue of morphogenesis process. They are characterized by a normal cycle of cellular replacement through all stages of development, and are not subject to a spontaneous involution. The weave type of vascular malformation determines the vascular classification into subcategories (capillaries, venous, arterial and lymphatic or mixed (fig. 1, 6). Another parameter which specifies the nature of these changes is the velocity of blood flow within them. We classify these changes into a low flow (containing elements of veins, capillaries and lymph vessels) and high capacity (with elements of arteries). These changes havever may stay unnoticed until the puberty or even adulthood period. They occur most frequently as a single malformation, and in rare cases, a family history of vascular malformations has been identified (11,12). Treatment of vascular malformation is very difficult, and surgery should be the last resort because recurrences are not a rarity. If the indication for surgery is pain associated with these malformations, Koman (13) recommends that the treatment should be started with oral calcium channel blockers and/or α₂ agonists, and only when it proves ineffective, can you qualify a patient for surgery. Surgical treatment depends on the symptoms, location – close to major vascular trunks. Endovascular embolization is a procedure of choices, but unfortunately it is not always possible perform it. The most common methods of surgical treatment of vascular malformations include:

a) total excision of the tumor with ligation of afferent and efferent vessels.
b) ligation alone of the main blood afferent and efferent vessels.
c) excision of arteriovenous fistula with simultaneous vascular reconstruction.
d) four ligations technique, proximal and distal vessels are ligated.

CONCLUSIONS

1. Upper extremity is a rare location of hemangiomas and other vascular malformations requiring surgery.
2. Hemangiomas are more common in women than in men, the incidence of vascular malformations in both sexes is similar.
3. Hemangiomas and other extremely large vascular anomalies with intramuscular location often produce symptoms of compression of peripheral nerves, which is an indication for surgical treatment.
4. Surgical treatment of hemangiomas and vascular malformations of the upper extremities is difficult to apply and it requires expert knowledge of microsurgical techniques.
5. Surgical treatment of vascular malformations is associated with a relatively high recurrence rate.
6. Vascular malformations in upper limbs are often located distally, which may increase the risk of ischemic changes in the distal parts of fingers and which creates the necessity of partial amputation.

REFERENCES


