The treatment of extensive arteriovenous malformations in the head

Piotr Wójcicki, Karolina Wójcicka

Plastic Surgery Department, Medical University in Wrocław, Specialist Medical Center in Polanica Zdrój

Kierownik: prof. dr hab. P. Wójcicki

Vascular tumours and malformations are revealed at birth and do not subside. The aim of the study was to present the principles and outcomes of treatment of patients with arteriovenous malformations treated at the Clinic of Plastic Surgery in Polanica Zdrój in the years 2009-2010.

Only one patient, who had not been treated previously, had the lesion on the cheek removed subtotally and the defect was closed by means of local repair. In the remaining patients, with primary lesions located in the auricle, scalp, and cheeks, the indications for operation included recurrent infections, ulcerations, and first of all, massive, life-threatening haemorrhages.

All the patients, treated for many years in other centres, had undergone numerous resection procedures, vessel ligations, embolizations and obliterations. The patients were followed up after the surgery every 6 months.

The therapy aim was achieved in all the patients. Vascular tumours were removed totally or subtotally, the lost structures were reconstructed and permanent healing of the wound was achieved. None of the patients developed recurrence of the disease, infection, or bleeding.

Key words: arteriovenous malformations, vascular malformations, vascular tumor.
Depending on the vascular component, the following vascular malformations can be distinguished: capillary, venous, lymphatic, arterial, or mixed, i.e. arteriovenous malformations. Taking into account the blood flow, the following types are distinguished: low-flow (capillary, venous, or lymphatic) malformations and high-flow (arterial or arteriovenous) malformations (11).

Vascular malformations affect men and women equally, however haemangiomas are 6 times more common in girls (12).

There is no one universal therapeutic modality for patients with vascular malformations. Every patient should be carefully diagnosed and treated by a multi-specialist team. In case of arteriovascular tumours, Schobinger staging may be helpful in evaluation of lesions and determination of indications for therapy (13). Stage 1 includes only arteriovenous shunt, stage 2 – a mass associated with a bruit and a thrill, stage 3 – ulceration, bleeding and pain, stage 4 – secondary circulatory failure.

**CASE REPORTS**

There were 5 adult patients with arteriovenous malformations located in the head treated at the Clinic of Plastic Surgery in Polanica Zdrój in the years 2009-2010. In four of them the lesions were recognized at birth, in one – at preschool age, manifested as gingival bleeding. All the patients had been treated primarily in other centres (tab. 1).

In patient K.K. (25 year old) the lesion on the interior surface of the cheek was noticed in the neonate period. The tumour developed slowly, involving more and more area in the oral cavity as the boy was growing up. The patient had not been treated before. He was operated on in 2010. The lesion was resected from the oral vestibule and the cheek.

The patient F.A. (23 of age) with malformation localized to the right auricle, had a series of laser therapy and two tumour embolizations performed in the years 2001-2008. The patient was qualified for surgical treatment in view of intensive growth of the tumour, recurrent infections and chronic ulcerations in the upper pole. In January 2009 the patient had embolization, followed by a surgery a week later. The vascular tumour was removed in whole, with simultaneous flap reconstruction and modeling of the auricle (fig. 1 A, B, C).

In the patient P.A. (22) the skin lesion localized to the skull cap was first noticed as a slight discoloration in infancy. When she was growing up, a slow but constant growth of the tumour was observed, which became more intensive in adolescence. Despite several consultations in other centres, no indications for treatment had been determined. In 2007 the patients underwent a cycle of monthly sclerotherapies, after which a further progression of the disease took place. The tumour increased its size, causing elevation of the skin on the periphery. The tumour growth was associated with chronic infection, recurrent bleeding, and enlarging necrosis in the central portion. Several days prior to the planned surgery embolization was performed. The patient was operated on in December 2009 – the tumour was excised in whole (fig. 2 A, B, C).

The patient K.J. (32) was admitted to the Clinic for the risk associated with an extensive Table 1. Patients with a-v malformations of the head

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Localization</th>
<th>Type</th>
<th>Symptoms</th>
<th>Preoperative treatment</th>
<th>Surgery</th>
<th>Complication</th>
</tr>
</thead>
<tbody>
<tr>
<td>K.K.</td>
<td>25</td>
<td>scalp</td>
<td>a-v</td>
<td>bleeding</td>
<td>no</td>
<td>1 x resection</td>
<td>no</td>
</tr>
<tr>
<td>F.A.</td>
<td>23</td>
<td>ear</td>
<td>a-v</td>
<td>ulceration, bleeding, infection</td>
<td>laser therapy, 2x embolisation</td>
<td>1 x resection, local flap</td>
<td>no</td>
</tr>
<tr>
<td>P.A.</td>
<td>22</td>
<td>cheek</td>
<td>a-v</td>
<td>ulceration, bleeding, infection</td>
<td>sclerotherapy</td>
<td>1 x resection, 1 x expander</td>
<td>no</td>
</tr>
<tr>
<td>K.J.</td>
<td>32</td>
<td>cheek</td>
<td>a-v</td>
<td>2x massive bleeding</td>
<td>surgical interventions / embolisation</td>
<td>1 x resection, 2x corrections</td>
<td>no</td>
</tr>
<tr>
<td>G.M.</td>
<td>27</td>
<td>cheek mandible</td>
<td>a-v</td>
<td>many massive bleeding</td>
<td>surgical interventions / embolisation, laser therapy, cryotherapy</td>
<td>5 x resection, forearm flap</td>
<td>maceration</td>
</tr>
</tbody>
</table>
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Fig. 1A. Vascular malformation tumour localized to the right auricle after a series of laser therapy and tumour embolizations.

Fig. 1B. After tumour resection in whole, with simultaneous flap reconstruction (Z plasty) and modeling of the auricle.

Fig. 1C. Final outcome.

Fig. 2A. Arterio-venous tumour of the scalp after a cycle of monthly sclerotizations. 4 days before surgery.

Fig. 2B. Expanded skin of the scalp, before skin graft replacement.

Fig. 2C. Final result.
tumour involving the right cheek and the lower right eyelid. The vascular malformation localized to the cheek and present since birth started to enlarge dramatically at adolescence. For this reason the patient underwent ligation of the external carotid artery branches. The patient had had three deliveries. During every pregnancy the tumour increased its size, assumed intensive staining, and its surface revealed a palpable pulsation. Prior to the last delivery the patient had massive haemorrhage. Her life was saved thanks to quick resuscitation action. A successive massive haemorrhage occurred a year later on a trial of a diagnostic puncture of the tumour. Since 2008 the patient had been attended at the Institute of Psychiatry and Neurology. Subsequent embolization was performed in hypothermia with the use of a harmonic knife. Haemostatic sutures were placed in order to reduce peripheral bleeding. The tumour was excised in whole, and only an area of discoloured skin was left in the upper part of the cheek and on the eyelid. Two successive corrective procedures were performed to reduce the size of the area.

The patient G.M. (27) had vascular malformation localized to the mucosa below the mandibular body on the right side, which was diagnosed at early preschool age. Several years later recurrent and constantly exacerbating gingival bleeding set in. Following a subsequent incident, the patient underwent ligation of the facial artery in a paediatric surgery department. In view of lack of improvement, the child was transferred to the Department of Maxillo-Facial Surgery, where her external maxillary artery was ligated. Prior to admission of the patient to the Clinic, she underwent numerous laser therapy and cryotherapy procedures as well as two embolization procedures.

Since 1997 the patient had four partial resections of the tumour, embolization and series of injections with concentrated levels of ethanol performed at the Polanica Hospital. Periods of remission were followed by the tumour growth on the periphery, often associated with massive bleedings especially from the gingivae above the hypertrophic mandibula. Angiotomography was performed to visualize a network of surrounding vessels.

In February 2009 the patient underwent subtotal resection of the tumour. Lesions involving almost whole cheek, oral vestibule, lateral parts of the upper and lower lips were removed in hypothermia. The mandible was split and its pathologically hypertrophied, intensively bleeding external part involving the body and the ramus on the right side was removed. The wound was sutured situationally. During the surgery and in the first 24 hours after the operation the patient was given 5 liters of blood. After 3 weeks the residual wound was covered with skin grafts, producing complete healing.

After a year a cheek contracture was resolved and residual lesions in the region of mandibular angle were removed. The defect on the cheek was reconstructed by means of island flap from the forearm, with the skin directed towards the lumen of the oral cavity. The wound surface of the flap on the external side was covered with skin graft. After 6 months the patient underwent modeling of the upper lip (fig. 3 A-G).
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Fig. 3C. Pathological hypertrophy of the mandible

Fig. 3D. Island forearm flap

Fig. 3E. State after island flap division

Fig. 3F. Island flap from forearm, with the skin directed towards the lumen of the oral cavity. Outside surface covered by the skin graft

Fig. 3G. Final result

RESULTS

Histopathological examinations revealed the presence of arteriovenous malformations in all the cases. The patients were followed up every 6 months. The aim of treatment was achieved in all the patients. Vascular tumours were removed totally or subtotally, the lost structures were reconstructed and the wounds healed permanently. None of the patients developed recurrence of the disease, infection nor bleeding.

DISCUSSION

The treatment of vascular malformations is difficult and poses a significant challenge for the surgeon.

Indications for urgent surgical treatment include life-threatening situations, such as cardiac failure, obturation of the respiratory tract, severe coagulopathy as well as impairment of basic vital functions, i.e. breathing, nutrition, excretion; negative consequences of the tumour growth and its compression on the optic nerve, eyeball, structures of the middle
ear, occlusion defect, or disturbances of the motor functions.

Among vascular malformations, lesions of a mixed type, arteriovenous lesions are most difficult to treat. In such cases extensive and thorough diagnosis is essential in order to determine the exact type of the malformation, localization of the tumour, as well as evaluate potential risks. For this reason, then use of angiography, CT or magnetic resonance is especially advocated.

In majority of cases arteriovenous malformations do not require surgical treatment in infancy and early childhood, they only have to be checked up and observed regularly.

A surgical procedure, regardless of the patient’s age, is necessary when there is a risk of severe complications, such as haemorrhage, ischemia (gangrene, ulceration, arterial insufficiency), prolonged venous insufficiency with hypertension, imminent respiratory failure, the risk of sight and hearing loss, difficulties with food intake, cardiac failure (14). The indications concern stage 3 and 4 arteriovenous malformations according to Schobinger.

Relative indications determining the decision to institute surgical treatment are either of medical or of psychosocial character. In children the existing functional and developmental disturbances play an important role, in adults – poor quality of life associated with the risk of complications – bleedings, bone fractures, presence of deformities, which cause functional disturbances and cosmetic deformities, as well as difficult to mitigate pains.

In many cases the therapy for vascular malformations starts from sclerotherapy or embolization (15). Obturating material – microspheres or cyanoacrylic glue, less commonly – balloons, coils, self-adhesive coils or spongostan – are inserted into the vessel lumen. It seems that such management should consider the option of resection in case of lack of improvement. Otherwise, an increasing tumour may cause substantial complications.

On the other hand, large arteriovenous tumours should be embolized before the surgery, most commonly 2-5 days prior to the planned surgical resection, in order to minimize bleeding.

Ligature of a large cervical artery should be treated as an emergency procedure performed in order to stop massive bleeding. Intense growth of collateral circulation which usually follows the procedure hampers the surgical treatment in the future, thus the next step it should be the decision to perform a resection surgery.

It seems that postponing the arteriovenous malformation surgery in patients who reveal the symptoms of growth is not justified (16). A delay in making the decision may only lead to an increased probability of developing infection, necrosis or bleeding within the tumour and definitely hampers the procedure itself. Large mass of the tumour, supplied by extensive network of abnormal blood vessels, as well as pathologically hypertrophied bones, which form spaces filled with intensively inflowing blood, may be the reason of difficult to manage haemorrhage. Moreover, closure of extensive defects in the head poses more difficulties than closure of lesser defects after resection of primary tumours (17, 18).

Repeated embolizations, or vessel ligatures may permanently damage the recipient vessels used in microsurgical procedures, what significantly limits the range of reconstructive procedures.

In four patients the treatment was instituted in stage 3 and 4 of the disease. In all these patients, in view of persisting inflammation, there was a real risk of intracranial complications, such as meningeal infections and cerebral embolisms. In a 22-year-old woman, a disintegrating necrotic tumour on the scalp eliminated her from normal life for two years. The life of two remaining patients was at severe risk due to recurrent, severe haemorrhages.

CONCLUSIONS

1. Arteriovenous malformations located in the head may be a source of infection, prolonged pain, frequent bleedings, and in most severe cases, may be life-threatening due to recurrent massive haemorrhages.
2. Chronic, large arteriovenous tumours cause facial deformations.
3. Radical resection of the malformation is the most effective modality of treatment.
4. The surgical procedure should be preceded by embolization of the tumour.
REFERENCES


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Adress correspondence: 57-320 Polanica Zdrój, ul. Jana Pawła II 2