Endovascular Treatment for Craniofacial Arteriovenous Fistula/Malformation

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Objective: To introduce our experience of endovascular treatment for craniofacial arteriovenous fistula/malformation (AVF/M).

Methods: We retrospectively analyzed the medical records of 13 patients (7 females and 6 males) with craniofacial AVF/M who were treated between 2001 and 2017 in our institution. We classified into three categories including single AVF (sAVF), multiple AVF (mAVF), and arteriovenous malformation (AVM). Treatment plans included 1) curative embolization, 2) preoperative embolization, and 3) palliative embolization. These strategies were decided by the discussion with plastic surgeons in every individual case.

Results: Complete cure by embolization alone was obtained in all six patients with sAVF, in two among three patients with mAVF, and in none among four patients with AVM. Curative embolization was aimed at in eight patients, and complete cure obtained in all eight patients. Preoperative embolization was aimed at in three patients, and three patients resulted in total resection by surgery after successful partial embolization. Palliative embolization was aimed at in two patients, and these patients were kept in a stable condition after partial embolization. No permanent complications related to embolization were counted.

Conclusion: Endovascular treatment for craniofacial AVF/M is safe and effective treatment, especially in the case with sAVF.

Keywords: head and neck, scalp, lip, embolization, n-butyl-2-cyanoacrylate

Introduction

Craniofacial arteriovenous fistula/malformation (AVF/M) is a relatively rare disease and several departments including pediatrics, dermatology, otolaryngology, plastic surgery, radiology, and neurosurgery are involved in treatment. Fundamental treatment of this disease is surgical excision, but it is often difficult when the lesion is large, and endovascular treatment also plays an important role. Inversely, when the lesion is small, complete cure by endovascular treatment alone can be expected. Embolization of the head and neck region requires sufficient anatomical knowledge, selection of an appropriate embolic material, and techniques compared with those in other regions because of the anatomical complexity and risk of neurological complication through latent dangerous anastomosis between the facial, head, and neck arteries and cerebral/retinal arteries. Moreover, esthetic problems and preservation of important functions, such as breathing, vocalization, and swallowing, are important points of treatment. Endovascular treatment is classified based on its role into 1) curative embolization, 2) preoperative embolization, 3) palliative embolization, and various liquid and solid embolic materials are differently used corresponding to the condition. Embolization by direct puncture may be useful depending on the lesion. We report practice and outcomes of endovascular treatment performed in our institution.
### Materials and Methods

We retrospectively evaluated the endovascular procedures in 13 patients with craniofacial AVF/M treated at our department between January 2001 and December 2017. In addition, the treatment outcomes (safety and efficacy) were retrospectively investigated.

Seven and six patients were females and males, respectively, and the age ranged from 14 to 72 years old (mean: 41 years old). Based on angiographic findings, a fistula with direct flow from the feeder to the dilated drainer was diagnosed as arteriovenous fistula (AVF), and that considered mediated by a nidus was diagnosed as arteriovenous malformation (AVM). AVF was further classified into those with a single fistula as single AVF (sAVF) and several fistulous points as multiple AVF (mAVF). In addition, the numbers of feeders actually embolized and treatment sessions, embolic materials, and treatment results were investigated.

A strategy of treatment was sufficiently devised with the plastic surgeon in charge of surgery, and the objective of embolization: 1) curative embolization, 2) preoperative embolization, or 3) palliative embolization, target vessels to be embolized, regions to be occluded, and optimum embolic material were investigated in each strategy.

Embolization was performed through the femoral arterial approach under local anesthesia. A guiding catheter was stably placed in the external carotid artery, a microcatheter was then inserted and advanced close to the lesion, and embolization was performed. An over-the-wire microcatheter was used as the first choice, but when the feeder was strongly curved or tortuous, a flow-guided microcatheter was used. For scalp lesions with difficulty in catheter insertion, embolization was applied by direct puncture of the lesion.

A liquid embolic material was injected from the feeding artery to the draining vein passing through the fistula site in AVF, and AVM was to be filled with a liquid embolic material from the feeding artery to the nidus as the general embolization strategy. For preoperative embolization, after discussion with the operator of excision, a solid embolic material was used to occlude the feeder side, or 4-n-butyl-2-cyanoacrylate (NBCA) was injected so as to stay in the feeder as feeder occlusion.

### Results

All 13 patients are listed in Table 1. The angiographic diagnosis was sAVF in six patients, mAVF in three patients, and AVM in four patients. Regarding the feeder, since it was difficult to identify the entire vascular architecture of lesions with a very small or complex nidus on angiography, the number of feeding arteries examined by highly selective angiography was presented in Table 1. The number was 1 in sAVF in all six patients, 2–7 (mean: 4) in mAVF, and 3–7 (mean: 5) in AVM. The rate of complete cure achieved by endovascular treatment alone decreased as the number of feeders increased.

The embolic material (partially overlapping) was NBCA in 10 (direct puncture in 2), coils in 4, polyvinyl alcohol polymer (PVA) in 3, Embosphere in 1, silk thread in 1, and ethanol in 1 (direct puncture). Excluding three patients with direct puncture, the embolic material was injected through a microcatheter super-selectively inserted through a feeding artery. In all three patients with direct puncture, the varix of AVF was punctured and the liquid embolic material was directly injected into the varix to occlude the lesion. The lesion was AVM and AVF in three and five of eight patients treated with NBCA injection, respectively, and complete occlusion was achieved in all three and five patients by feeder occlusion and injection into the fistula, respectively. Particulate embolic materials, PVA and Embosphere were used in five patients. It was used for feeder occlusion in palliative and preoperative embolizations in two patients each and curative embolization in one in whom it was used to occlude the feeders of other feeding arteries before curative embolization with NBCA. Silk thread was concomitantly used with coils to occlude a very larger varix of sAVF and achieved complete occlusion.

The number of embolization sessions was 1, 2, 3, and 4 in 10, 1, 1, and 1 patient, respectively. Regarding the results of embolization, complete occlusion was achieved in eight of the eight patients treated aiming curative embolization, and the lesion was entirely excised by subsequent surgery in three patients treated with preoperative embolization. Two patients treated aiming at palliative embolization from the beginning are currently under course observation after partial embolization performed as planned and no re-enlargement has been noted so far.

No permanent complication of endovascular treatment occurred, but skin necrosis developed in the patient treated with ethanol and it was temporarily accompanied by crust formation, being esthetically problematic, but remitted by conservative treatment (Case 1).
<table>
<thead>
<tr>
<th>No.</th>
<th>Sex</th>
<th>Age</th>
<th>Location</th>
<th>Symptom</th>
<th>Diagnosis</th>
<th>No. of feeder</th>
<th>Strategy</th>
<th>No. of session</th>
<th>Embolic material</th>
<th>Result</th>
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<td>3</td>
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AVM: arteriovenous malformation; CO: complete occlusion; mAVF: multiple arteriovenous fistula; NBCA: n-butyl-2- cyanoacrylate; op: operation; PO: partial occlusion; PVA: polyvinyl alcohol polymer; sAVF: single arteriovenous fistula
Representative case 1 (No. 2)

30-year-old female

The patient was referred to our hospital requesting endovascular treatment of a pulsatile mass which had been gradually enlarging for several years on the left forehead. A varix fed by the left superficial temporal artery (STA) and angular artery was noted on CTA (Fig. 1A), and the patient was diagnosed with mA VF flowing into this varix. The varix was visible as a pulsatile mass through the skin.

When the region around the lesion was compressed with a metal ring on the skin and the two feeders were manually pressed, pulsation of the varix was markedly reduced (Fig. 1B). After confirming this, the varix was directly punctured with a 23G butterfly needle (Fig. 1C). When DSA was first performed without compressing for confirmation, contrast medium was rapidly washed out from the varix toward the angular vein. Then, it was confirmed that when a 5:5 mixture solution of contrast medium and local anesthetic, 1% Lidocaine, was injected while compressing the region, contrast medium was retained in the varix, both feeders, and drainer around the varix, and the total volume was less than 1 cc (Fig. 1D). Subsequently, 1 cc of ethanol was injected under compressing as described above and this condition with compressing was kept for 5 minutes, followed by removing ethanol by aspiration (only 0.1 cc could be aspirated) and the procedure was completed. No marked event occurred in the patient after treatment, and the patient was discharged on the day following treatment with disappearance of the pulsatile mass.

On examination at the outpatient clinic after 1 month, the pulsatile mass had completely disappeared but severe reddening was noted on the skin. It aggravated and was accompanied by crust formation after 3 months but remitted after 6 months (Fig. 2), and the appearance became mostly normal after 1 year (no photograph shown. It was reported by the patient by phone call).

Representative case 2 (No. 12)

14-year-old female

Swelling of the left upper lip had been pointed out since infancy, but it was left untreated because swelling was mild. Swelling of the left upper lip became noticeable several years earlier, and hemorrhage in the oral cavity repeatedly occurred resulting in anemia from 3 months earlier, and the patient was referred to our hospital for endovascular treatment.

At the time of arrival, minor bleeding repeated every day. DSA was performed the patient with general anesthesia,
Representative case 3 (No. 8)

15-year-old male

The patient was referred to our hospital for a chief complaint of right pulsatile tinnitus aggravating for 1 year. A pulsatile mass was noted in the posterior region of the right auricle. Bruit was heard in the same region and resolved by pressing the carotid artery, and the size of the mass was also markedly reduced. On DSA, sAVF fed by the right occipital artery (OA) was noted, the drainers were subcutaneous veins in the upward and downward directions and transverse sinus through the emissary vein in the anterior direction (Fig. 5). No reflux into the brain surface vein was observed, but endovascular treatment was selected because of the presence of intracranial reflux and strong complaint of tinnitus.

Under local anesthesia, 6 Fr ROADMASTER (Nipro, Osaka, Japan) was stably placed in the right OA, and Headway-17 (Terumo Corporation, Tokyo, Japan) was coaxially inserted into the pouch of the emissary vein passing through the fistula. Since the lesion was a high-flow fistula (Fig. 6A), first, the pouch, then subcutaneous vein, and drainage side were filled with detachable coils to some extent (Fig. 6B) followed by filling 50% NBCA into the coil mass to the proximity of the feeder sandwiching the fistula (Fig. 6C), and AVF completely disappeared (Fig. 6D). The symptoms were resolved immediately after treatment and the patient was discharged on the following day without any marked

and AVM was detected in the left upper lip (Fig. 3A–3C).
The main feeders were the internal maxillary artery (IMA) and facial artery (FA) forming a nidus in the left upper lip, and blood flowed from the superior labial vein into the angular vein. According to information provided by the previous physician, the bleeding point was present in the left upper region in the oral cavity. Together with the angioGraphic findings, it was considered located in the peripheral region of the FA so that embolization was subsequently performed. 6 Fr ROADMASTER (Nipro, Osaka, Japan) was stably placed in the left external carotid artery, Magic 1.4 Fr (Balt, Paris, France) was then advanced to the superior labial branch of the FA, and this feeding artery was occluded with 25% NBCA (Fig. 3D–3F). After treatment, no marked event occurred in the patient and the hemorrhagic episode was completely resolved.

After 6 months, complete cure of the residual AVM was requested. For this esthetic treatment, preoperative embolization + plastic surgery was planned. Under general anesthesia, six branches from the IMA and FA were occluded with 25% NBCA but feeder occlusion was adopted to prevent necrosis of the lip (Fig. 4A and 4B). The lesion was excised by plastic surgeons on the following day, excision was easy taking 32-minute operative time and 10 cc blood loss, and a good-looking lip was formed (Fig. 4C and 4D). The postoperative course was favorable and esthetic satisfaction of the patient was high.
event. On CTA performed after 1 year, AVF had disappeared and the esthetic appearance was favorable (Fig. 6E).

**Discussion**

Previously, there have been various classifications of AVF/M and the contents were confused. After International Society for the Study of Vascular Anomalies (ISSVA) classification was proposed, treatment methods have been organized based on this classification. Craniofacial AVF/M is a relatively rare disease, and several departments, such as pediatrics, dermatology, otolaryngology, plastic surgery, radiology, and neurosurgery, are involved in treatment. In the ISSVA classification, AVMs are classified into AVF and AVM with fast flow, but in the present study, AVF was further classified into single and multiple fistulas. Of course, the likelihood of healing by endovascular treatment alone was high in the order of sAVF, mAVF, and AVM. The treatment strategy is basically the same as that for AVMs of the four extremities. However, the incidence of complication of endovascular treatment in the facial, head, and neck region is higher than that in the four extremities because in addition to the anatomical complexity, there is a risk of neurological complication due to latent dangerous anastomosis between the facial, head, and neck arteries and cerebral/retinal arteries. Moreover, facial lesions should be more carefully treated from the esthetic viewpoint. Furthermore, lesions located in deep regions and very large lesions may threaten important functions, such as breathing, vocalization, and swallowing. So, preservation of these important
functions is important in treatment. Fortunately, most lesions were relatively simple and not large in the present series and the outcomes of endovascular treatment were favorable compared with those in previous studies.\(^5\)\(^7\) In addition, we closely cooperated with the plastic surgery department in charge of surgical resection and the strategies (course observation, endovascular treatment alone, and surgery following preoperative embolization) were sufficiently discussed before treatment. These may also be the reasons of improvement of the outcome.
Regarding the objective of embolization, after sufficient discussion of a treatment strategy with the plastic surgeons, the role of embolization was clarified as follows: 1) Curative embolization aiming at complete cure by endovascular treatment alone, 2) preoperative embolization, and 3) palliative embolization for cases difficult to be completely cured even by combination with surgical treatment. Palliative embolization is performed in patients repeating hemorrhage, targeting hemostasis and prevention of hemorrhage by occluding the bleeding point and size reduction of large lesions for which complete cure cannot be expected. The objective 1) requires sending the embolic material from the arterial to the venous side passing through the fistula/nidus. Inversely, for the objective 2), even feeder occlusion is very useful. Setting the embolization target at the feeder, which is difficult to control for the operator, is important and sufficient preoperative discussion with the operator of the excision is necessary as is for cerebral AVM. For the objective 3), several issues, including the long-term effect of embolization, development of collateral circulation, and recurrence due to vascularization after feeder occlusion, should be considered. Thus, it is desirable that the embolic material reaches the venous side, similarly to that for 1).4,7) For this purpose, direct puncture of the lesion may be useful.

For the embolic material used in endovascular treatment, generally, PVA particles,4,8) Embosphere particles,9) detachable balloon,10,11) Gelform, silk thread, and coil14,16) are used as solid materials, NBCA4–7,10–12) and ONYX13) are used as liquid materials, and ethanol5,14–16) Bleomycin, and Picibanil (OK-432)17) are used as liquid curing agents. We frequently use NBCA because it has high complete curing ability and we are familiar with handling it. However, when the lesion is present at a distant site, NBCA is difficult to use. In addition, NBCA mass becomes indurated when a large volume is injected into a lesion directly below the skin, and it may become esthetically problematic. It has been pointed out that when NBCA is used for preoperative embolization for cerebral AVM, surgery is interfered with by its hardness. Since the surgical field is shallow in the
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For the hemorrhagic case (Case 2; No. 12) requiring semi-urgent embolization, since the feeding artery at the bleeding point could be accurately identified based on information provided by a physician engaged in the initial treatment, complete hemostasis could be acquired after treatment. Subsequently, the feeder was internally occluded with NBCA to excise the residual AVM in the left upper lip for the esthetical purpose. After discussion with a plastic surgeon, feeder occlusion was performed centering the feeder from the deep region so as to prevent necrosis of the lip by NBCA reaching the lip, which contributed to safer excision.

In the case with high-flow single fistula (Case 3; No. 8), tortuosity of the feeder was not so strong and an over-the-wire type microcatheter could be advanced into the dilated drainer immediately after the single hole fistula. After framing a ‘foothold’ with coils on the venous side, NBCA was injected entangling with the coils and complete occlusion could be obtained. Although it was a high-flow fistula, flow was reduced by the coils and both arterial and venous sides could be sufficiently filled with NBCA sandwiching the fistula, which may have resulted in complete occlusion.

In all patients, the appropriate embolic material and embolization method were selected after sufficient discussion with plastic surgeons, and this may have led to the success, but diluted NBCA should have been used for Case 1 (No. 2).

Limitations of this study include the following: 1) a small number of cases, 2) inclusion of many cases of simple lesions represented by sAVF compared with those in previous reports (accordingly, the treatment outcome was favorable), 3) retrospective study, and 4) variation of the content of cases and treatment procedure being not uniform.

Conclusion

Endovascular treatment for craniofacial AVF/M can be safely performed with understanding of the vascular architecture and with knowledge of the functional anatomy, and complete cure by endovascular treatment alone can be expected depending on the lesions. On the other hand, some lesions are complex and intractable. We would like to emphasize that treatment should be performed using an appropriate embolic material and appropriate method after sufficiently understanding the pathology of each case before treatment in close cooperation with the plastic surgery department (physician in charge of surgical treatment).
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Disclosure Statement

The author declares there is no conflict of interest related to this work.

References