Life-Threatening Arteriovenous Malformation of the Maxillomandibular Region and Treatment Outcomes

A. CHUROJANA¹, R. KHUMTONG², D. SONGSAENG¹, C. CHONGKOLWATANA³, S. SUTHIPONGCHAI¹

¹ Department of Radiology, ³ Department of Otolaryngology, Faculty of Medicine, Siriraj Hospital, Mahidol University; Bangkok, Thailand

² Department of Radiology, Faculty of Medicine, Songklanagarin Hospital, Prince of Songkla University; Tambon Korhong, Thailand

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Summary

We report on cases of life-threatening maxillomandibular arteriovenous malformations (AVM) whereby patients had successful endovascular treatment with good outcomes. Out of a total 93 facial AVMs treated endovascularly between 1991 and 2009, five patients (5.4%) had maxillomandibular AVMs. All presented with uncontrolled dental bleeding. Endovascular procedure was the primary treatment of choice in all cases, either transfemoral approach with arterial feeder embolization or transosseous puncture, depending on the accessible route in each patient. NBCA (glue) was the only embolic agent used. Tooth extraction and dental care were performed after bleeding was controlled.

All five patients (8-18 years) with a mean age of 12.4 years presented with massive dental bleeding following loosening of teeth, dental extraction and/or cheek trauma. The plain films and CT scans of four patients with AVMs of mandibles and one of maxilla, revealed expansile osteolytic lesions. The mean follow-up period was 6.6 years (ranging between one and 19 years). Three cases developed recurrent bleeding between two weeks to three months after first embolization, resulting from residual AVM and infection. Late complications occurred in two patients from chronic localized infection and osteonecrosis, which were successfully eradicated

with antibiotic therapy and bony curettage. Complications occurring in two patients which included soft tissue infection, osteomyelitis and osteonecrosis were successfully treated with antibiotics, curettage and bone resection. No patient had a recurrence of bleeding after the disease had cured.

Initial glue embolization is recommended as the effective treatment of dental AVMs for emergent bleeding control, with the aim to complete eradicate the intraosseous venous pouches either by means of transarterial superselection or direct transosseous puncture. Patient care by a multidisciplinary team approach is important for sustained treatment results.

Introduction

Approximately 50% of arteriovenous malformations (AVM) occur in the head and neck¹ region and about 50% of intraosseous AVM are also found in this region². AVM of the maxillomandibular region is a rare disease³⁻¹⁰ but can be potentially life-threatening due to massive dental hemorrhage^{3,4,6,7,9-11}. Almost all patients presenting with AVM are children or adolescents^{3,4,6,7,9-11}. Endovascular embolization often has to be the initial treatment for this emergency condition with the aim for cure. However, primary segmental excision or radi-

cal resection of the involved bone after preoperative embolization has been reported¹². The purpose of this study is to report the life-threatening maxillomandibular AVMs successfully treated by endovascular embolization in our hospital during an 18 year period, as well as the treatment outcome.

Materials and Methods

Out of a total 93 facial AVMs submitted to endovascular treatment in our Radiology Department, Siriraj Hospital, Bangkok, Thailand between 1991 and 2009, five patients (5.4%) had maxillomandibular AVMs. There were three males and two females, ranging in age from eight to 18 years with a mean age of 12.4 years at initial presentation. All presented with massive dental bleeding following loosening of the teeth, dental extraction and cheek trauma. One patient had external carotid ligation before having recurrent massive dental bleeding. Four lesions involved the mandible, with the most extensive one involving the entire mandible. One AVM was disclosed at the alveolar process of the maxilla. All lesions exhibited characteristics of expansile osteolytic pattern on their plain films and CT scans.

The endovascular procedure was performed under general anesthesia in all cases. Angiography of branches of bilateral external carotid arteries (ECAs) was routinely done to assess the arterial feeders, venous drainage and intraosseous vascular lake of each AVM. Bilateral internal carotid arteries (ICAs) and vertebral arteries (VAs) were also studied to evaluate any danger of anastomosis and associated intracranial AVMs on the initial study. All AVMs received ECA supplies (mainly from inferior alveolar branch with anastomosis from facial-lingual system) with intraosseous venous pouches of varying sizes. The embolization procedure was planned either by superselective transarterial route or transosseous puncture or a combination depending on the accessibility route of each patient. N-butyl cyanoacrylate (NBCA) or glue was the only embolic agent used for this purpose. For embolization via arterial feeding, a mixture of glue and lipiodol (at concentrations varying from 33-50%) was carefully injected under fluoroscopic monitoring with awareness of the dangers of anastomosis or reflux. For transosseous embolization, a no. 18 gauge needle was punctured directly into the

AVM nidus under fluoroscopic guidance. Injection test was always done to confirm appropriate location of the needle (with the intention to position in the large venous pouches or area of active bleeding). About 50-75% glue concentration was used in each session accompanied by manual compression of the venous outflow tract to prevent emboli into the circulation. Procedure was terminated after an adequate embolization was confirmed by controlled angiogram and then the bleeding site was checked after packing removal. Antibiotics were prescribed in every transosseous embolization.

Tooth extraction and dental care was accomplished, after the bleeding was well-controlled. Repeated embolization was carried out if there was recurrent hemorrhage or residual vascular space in the extensive lesion. Follow-up CT scan and/or angiogram were considered in some cases with extensive involvement, repeated dental bleeding or suspicion of any complication. When cured, clinical follow-up was scheduled at one, three and six months.

Results

All maxillomandibular AVMs were successfully treated by embolization initially for bleeding control. Three patients had transarterial embolization of arterial feeders, one had transosseous puncture and one had combined.

The mean follow-up period was 6.6 years (ranging from one to 19 years) (Table 1). Three cases had recurrent bleeding between two weeks and three months after first embolization. Complete disappearance of AVM could not be achieved in one of these patients due to extensive involvement of the entire mandible (Patient no. 1 in Table 1) (Figure 1). One patient had partial embolization with residual AVM involving the dental root (patient no. 4 in Table 1), and the other case had soft tissue infection after complete obliteration of the AVM nidus (patient no. 5 in Table 1) (Figure 2). Despite single embolization, two patients had complete cure without recurrence of dental bleeding during one year of follow-up, one had transarterial access (patient no. 2 in Table 1), whereas the other had direct puncture due to prior external carotid ligation (patient no. 3 in Table 1) (Figure 3).

Repeated embolization was done in two patients, one had two procedures and the other who had an extensive lesion with multiple epi-

Table 1 Patient summary and results.

No	Age (year)	Sex	Time of F/U	Clinical presentation	Location of AVM	Immediate post-embolization outcome	No. of endovascular treatments and reasons	Summary of outcome
1	10	M	19 yrs	Massive hemorrhage with shock from dental extraction. Bilateral pulsatile swelling of lower face with angiomatous discoloration, thrill and audible bruit.	Entire mandible	Good control of bleeding.	Multiple embolizations during 7 years due to several recurrent bleedings and chronic infection. Last session was for preoperative bony curettage due to osteonecrosis.	Good control of bleeding from multiple embolizations combined with surgical curratage with glue cast removal, partial mandibular resection and antibiotics. No longer symptomatic.
2	15	F	8 yrs	Massive hemorrhage from Rt. lower gum.	Rt. mandibular body	Good control of bleeding.	Single embolization without recurrence.	Cured
3	18	F	1 yr	Bleeding at Lt. lower molar tooth S/P external carotid ligation.	Lt. mandibular body	Good control of bleeding.	Single embolization without recurrence.	Cured
4	11	M	4 yrs	Rt. cheek trauma with massive bleeding at right upper premolar area.	Rt. side of maxilla	Good control of bleeding.	2 embolizations due to recurrent bleeding of residual AVM (re-embolization at 3 months of F/U).	Cured
5	8	M	1 yr	Massive hemorrhage after dental loosening at Lt. lower molar teeth.	Lt. mandibular body	Good control of bleeding.	Single embolization. Recurrent dental bleeding at 3 months of F/U due to soft tissue infection subsequently resolved by curettage and antibiotic treatment.	Cured

sodes of recurrent bleeding needed several procedures which were a combination of multiple direct punctures and transarterial access. Not only glue but also absolute alcohol which was the exceptional embolic agent were used in this patient (patient no. 1).

Late complications which resulted from delayed infection occurred in two patients (patients nos. 1 and 5). Patient no. 1 was found to have chronic osteomyelitis and osteonecrosis of the mandible at seventh year of follow-up, which was subsequently treated with surgical

bony curettage and resection. Patient no. 5 had localized soft tissue infection with recurrent dental bleeding at three months after initial embolization. Clinical cure was finally achieved after antibiotics therapy and surgical curettage.

Illustrative Case

A ten-year-old boy with a three-month history of bleeding from a loose milk tooth presented with severe bleeding until shock from

Figure 1 A 10-year-old boy presented with massive dental bleeding and hypovolemic shock. A,B) Angiogram of bilateral ECAs revealed extensive AVM involving the entire mandible with large venous pouches. Multiple transarterial and transosseous glue embolizations were done for initial treatment. C) Patient had neck exploration and bone curettage with removal of residual subcutaneous AVMs and glue cast (D). E) 15 years of follow-up: patient had subsequent chronic osteomyelitis and mandibular necrosis with frequent minimal bleeding. The angiogram shows residual AVM from many tiny arterial feeders with early draining vein (arrows). Direct puncture glue embolization was done into the AVM with glue casts deposited within the mandible (arrow-heads). Culture specimens from I&D and bone curette contained *Pseudomonas aeruginosa*. F) Patient's picture at 17 years follow-up.

Figure 2 An 8-year-old boy presented with acute episode of excessive periodontal bleeding at left lower molar teeth, after intermittent bleeding for 3 months from dental loosening resulting to blood transfusion. A) Expansile lytic bone lesion of the mandible was seen on CT scan (arrow). B) Angiogram of left ECA showed AVM receiving arterial supplies from left facial artery and inferior alveolar artery. Note venous pocket (dark circle) corresponding to an expansile lytic lesion on CT (arrow). C) Complete disappearance of venous pocket after embolization (white circle) was seen. Note reperfusion of the right submandibular gland (double arrows). D) During transarterial glue embolization, glue cast deposited in the venous pouch corresponding with lytic lesion on CT. Three months after embolization, the patient came with prolonged yet minimal bleeding and left cheek swelling. E) Angiogram of left ECA proved total obliteration of venous pocket. No residual lesion could be seen. F) CT scan showed abscess with soft tissue inflammation (arrows). Note glue cast within the mandible (arrowheads).

F

tions had to be repeated urgently due to recurrent bleeding. During the following four years after first discharge, he had more than ten instances of severe dental bleeding, multiple events of cardiac arrest, emergency transarterial and direct glue and alcohol embolizations, multiple tooth extractions and even tongue necrosis. In the next three year period, he still suffered frequent minimal dental bleeding, transarterial embolization was tried as much as possible, then subsequent neck explorations with removal of subcutaneous AVM nidi and

glue cast (Figures 1C and 1D). After seven years, he developed osteomyelitis and osteonecrosis of the mandible with soft tissue abscesses, and incurred hepatitis B and C infections resulting from multiple blood transfusions. Although he had multiple embolizations, multiple surgical curratages with occasionally pieces of glue cast removed and partial bony resection, he was symptom-free at 15 years after first embolization with minimal disfiguration of the mandible but preserved function without recurrent hemorrhage or infection.

Figure 3 An 18-year-old female presented with severe bleeding at left lower molar tooth. The patient had previously been treated for the same condition with left external carotid ligation from the other hospital. A) CT scan showed an expansile osteolytic lesion at the left mandibular body (arrow). B) Lateral angiograms on arterial and parenchymal phases of left ICA and left VA revealed mandibular AVM being supplied by collaterals from VA and ICA status post left ECA ligation. E) Complete obliteration of the AVM was obtained immediately after glue embolization via direct transosseous puncture. (C, D, double arrows). Subsequent dental extraction was done and patient had no recurrent bleeding during the 1 year follow-up period.

Discussion

Arteriovenous malformation (AVM) of the maxillomandibular bone is a rare disease of the oral and maxillofacial regions. The pathogenesis could be errors of vascular morphogenesis³ which although asymptomatic at birth, increase proportionally to the growth of the persons affected. Specific triggering factors such as endocrine or hormonal factors (puberty and pregnancy), trauma, iatrogenic insults (biopsy, incomplete surgery and proximal embolization)

and infection can all cause development of the lesions and aggravate symptoms. All patients in our study presented in the pediatric age group or had hit puberty. They came with either spontaneous massive dental bleeding, or bleeding which followed extraction and trauma.

Intraosseous maxillomandibular AVM is usually a life-threatening hemorrhage at presentation. It needs rapid and efficient treatment by an experienced team. Endovascular embolization has proven effective for immediate bleeding control¹²⁻¹⁷ and has to be performed

on an emergency basis as the initial treatment of choice. In addition, embolization can preserve functional anatomy and morphology, in contrast to malocclusion or disfiguration which may result from surgery.

In our experience, NBCA (N-butyl cyanoacrylate glue) was the only embolic agent to serve this purpose, not only because of its effectiveness for immediate vascular occlusion, but also because of its affordability. We used a mixture of glue and lipiodol at a concentration greater than 50%, with the aim of complete obliteration of the intraosseous venous lake or varices, whilst minimizing glue migration into the distal venous outlet or even pulmonary circulation. The approach could be transarterial, direct percutaneous puncture, or a combination. Transarterial selective catheterization was the basic consideration, as a standard method for embolization. Nevertheless, this method was limited:

1) In cases of too small and tortuous arterial feeders but large venous varices, it was difficult to obtain adequate glue deposition in the large venous lake injecting via a small feeder, and multiple microcatheters with several injections were necessary.

2) In cases where there was no arterial route access due to prior surgical ligation or proximal arterial occlusion.

In the aforementioned circumstances, direct puncture was instead our choice. Despite access being transosseous, the bony cortex of the involved mandible was thin enough for a no. 18 needle puncture, otherwise routinely used for femoral artery puncture, to penetrate. Once the venous varix was reached, a connecting tube was applied to minimize the radiation dose to the operator. We found injecting glue concentrate via the puncture needle, which had a larger diameter than a microcatheter, was more feasible and controllable, and allowed sufficient filling of the large venous lake. If complete venous occlusion could be achieved, total obliteration of the feeders could be expected, with a consequent complete cure as the immediate outcome. Additionally, using a direct puncture approach for glue embolization shortened the duration of the procedure, thus reducing expense.

Proximal artery occlusion by either surgical ligation or transarterial embolization should be avoided, as these procedures are not effective enough to stabilize this type of AVM and its bleeding sequela. Furthermore, recruitment of multiple collaterals after proximal artery occlusion is inevitable: those predominantly in this

particular area may ruin the chances of curative and palliative embolization for this type of AVM. In our experience, we only carried out proximal artery occlusion as a preoperative precaution and/or in cases of catheterization failure and disconnection of shunt/nidus.

Absolute alcohol is a known potential embolic agent that produces vascular thrombosis and might inhibit angiogenesis. However its effect on high-flow AVM is not promising and it cannot deliver immediate hemostatic control. Moreover, it is difficult to titrate an effective dose to offset its complications. We experienced the problem of a delayed skin and subcutaneous necrosis and subsequent chronic osteomyelitis in one of our patients (patient no. 1), who had extensive mandibular AVM and was receiving intrarterial ethanol injection for the residual feeders. Thus we would be warier in future of using this embolic agent.

Recurrent bleeding may occur after primary embolization in cases of incomplete embolization and/or infection, as we found in three cases of our series within two weeks to three months. Partial treatment and primary surgical resection was stated by Lasjaunias et al. to be a cause of uncontrolled bleeding and could also be a trigger for arteriovenous lesions growth³. Thus, once treatment of maxillomandibular AVM is considered, one must plan for cure. For large lesions, repeated embolization (or combined surgery) to occlude as many feeders as possible should be done. In cases where AVM involves erupting teeth, particularly close observation and follow-up is required due to the potential of massive bleeding occurring once the tooth is loosened (cf. patient no. 4).

Dental care, in particular, tooth extraction corresponding to the site of embolization is a key success factor in terms of preventing delayed infection which may become complicated by dental root abscesses and necrosis and eventual recurrent bleeding.

Another complication of embolization which may occur is delayed mucosa dehiscence, with exposure to the glue cast. It may be aggravated by local infection. This was found in patient no. 1, but was managed safely by surgical curettage after repeated transarterial embolization of the residual feeding arteries and finally, consequent osseous regeneration and remodeling.

There is a role for combined surgery in complete eradication of residual lesions post embolization, or cosmetic reconstruction in cases of massive lesions¹⁸⁻²⁰.

In our series then, the maxillomandibular AVMs were effectively treated by transcatheter or direct puncture, using NBCA as the permanent embolic agent. The size of the lesions did not affect the immediate outcome in terms of bleeding control. Tooth extraction with prophylactic infectious controls was necessary to achieve good long-term results, and to minimize the risk of recurrent hemorrhage and osteomyelitis.

We agree with Persky et al. that endovascular therapy alone can effectively "cure" most mandibular and maxillary vascular malformations with limited soft tissue involvement¹⁷. For successful treatment of maxillomandibular AVMs, complete eradication of intraosseous

venous lakes including all AVM nidi should be the aim for a permanent solution²¹.

Conclusion

In conclusion, even though initial glue embolization is not risk-free, it is still recommended as an effective treatment strategy for maxillo-mandibular AVMs which can cause fatal hemorrhage. Complete eradication of the intraosseous venous pockets is definitely the aim of treatment to get a permanent, curative result. Patient care by a multidisciplinary team using an emergent treatment approach is important for sustained treatment results.

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Dittapong Songsaeng, MD
Department Radiology
Siriraj Hospital, Mahidol University
2 Prannok Rd. Bangkok-noi
Bangkok 10700 Thailand
E-mail: dsongaeng@gmail.com