Three-dimensional computerized tomographic angiography diagnosis and surgical treatment of macroglossia with huge venous malformation: a case report

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Macroglossia caused by large venous malformation is extremely rare and difficult and risky for treatment. This report describes a case of macroglossia with severe venous malformation in a 50-year-old male patient. First, 3-dimensional computerized tomographic angiography (3D-CTA) was carried out before surgery for diagnosis. Then the patient underwent successful operation and follow-up for 2 years after surgery. Finally, the clinical outcomes of 3D-CTA examination and surgical procedure were evaluated. The experience of the diagnosis and surgery may be helpful to reduce the risk and increase the success rate of surgical treatment for macroglossia with huge venous malformation.

There are many causes for macroglossia. Some scientists categorize macroglossia into 2 types: congenital and acquired macroglossia.1-4 Congenital macroglossia may be caused by vascular malformations, Down syndrome, Beckwith-Wiedemann syndrome, Prader-Willi syndrome, or congenital hypothyroidism. Acquired macroglossia can be further divided into chronic and acute macroglossia. Chronic macroglossia is caused by tumor, amyloid degeneration, or myxedema. According to the etiology, acute macroglossia is classified by Renehan and Morton into 4 types: hemorrhagic, infection, infarction, and allergic edema.1 Congenital vascular abnormalities are some of most common diseases in oral and maxillofacial region. In 1982, Mulliken and Glowacki proposed a biologic classification based on the endothelial cell characteristics and the clinical behavior. Later, this classification was redefined by Mulliken and Young and adopted by the International Society for the Study of Vascular Anomalies in 1996.5,6 At present, it is the most widely used classification that includes vascular tumors and vascular malformation. Vascular malformation is subdivided into capillary, venous, lymphatic, arterial, and combined malformations. Vascular malformation is a typical cause of macroglos­sia.

In 2007, we admitted a patient of rare macroglossia caused by large venous malformation, which not only threatened the patient’s life, but was also difficult and risky to treat. In this report, we retrospectively summarize the diagnosis and surgical procedure of the macroglossia caused by large venous malformation. We hope that the experience we gained can be helpful for others to reduce the risk and increase the success rate in surgical treatment of macroglossia with huge venous malformation.

CASE REPORT
A 50-year-old male patient was admitted for hyperpyrexia and acute swelling of the tongue. The patient had a deformity of the tongue that protruded out of the mouth for >20 years. Shortly after birth, the patient was found to have a bluish
mass in the left cheek and in the tongue. At the ages of 8 and 17 years, the patient received surgeries in the tongue and the left submandibular region in another hospital. However, after surgery, the mass kept growing, especially in the tongue, which protruded out of the mouth and affected his face, eating, and speaking. In July 2007, the patient was admitted from the outpatient clinic with vascular malformation accompanying infection in the oral and maxillofacial region. After being admitted, the patient was subjected to a systemic medical examination and treated with antibiotics. After the systemic symptoms were better and the swelling tongue slightly went down (Fig. 1), the patient was prepared for the surgery.

**Methods of 3D-CTA examination**

Three-dimensional computerized tomographic angiography (3D-CTA) was performed with a 64-slice CT scanner (LightSpeed VCT; General Electric, Milwaukee, WI, USA). After obtaining a scout view to determine the 3D-CTA scan range, a 22-gauge plastic intravenous catheter was placed in the antecubital vein and connected to a power injector (Empower CTA; Ezem, USA). A bolus of 75 mL iopamidol (Iopamiro 370; Bracco, Milan, Italy) was injected into the antecubital vein at a flow rate of 4 mL/s. Dynamic scanning was started 5 seconds after the injection of iopamidol. The scan parameters were as follows: tube voltage 120 kV, tube current 450 mA, tube rotation time 0.4 seconds, detector collimation 64 × 0.625 mm, and pitch 0.561. Scan range was above orbit to the aortic arch. The reconstruction thickness of the 3D-CTA image was 0.625 mm and reconstruction interval 0.625 mm. These axial images were retrospectively reconstructed on a workstation (AW4.4l General Electric Medical System, USA).

**CT Scanning images**

CT scanning images showed that huge venous malformation occupied the tongue, oral floor, submandibular region, lips, left cheek, left parapharyngeal space, and lateral skull base. The mass filled the patient’s oral cavity, and even protruded from the mouth. The pharynx oralis of the upper respiratory track was almost obstructed (Fig. 2).

**3D-CTA images**

3D-CTA images showed that blood supply in the tumor was profuse and the tumor was huge, and there were many round, oval, or irregular “lipiodol pools” formed by expansion of blood sinus and accumulation of blood (Fig. 3). The macroglossia with venous malformation were supplied by bilateral lingual artery. The left external carotid artery had been ligated between the lingual artery and superior thyroid arteries, and the superior thyroid artery and ipsilateral lingual artery formed a collateral circulation. The upper and lower jaws were deformed, the upper and lower teeth were everted and kept open, and the dental arch and jaw were expanded.

Given the above data, the tumor was diagnosed as a huge venous malformation. The biggest risk of surgery for the patient with huge venous malformation was massive hemorrhage and respiratory tract obstruction.

**Surgical procedures**

The first surgery was performed under incision of trachea, tracheal intubation, and intravenous combined general anesthesia. We performed resection of venous malformation in the tongue, oral floor, lower lip, and left submandibular region, as well as a glossoplasty. The surgical procedures were as follows.

A horizontal incision in the bilateral submandibular region and a vertical incision at the midline of lower lip were designed. The left external carotid artery was ligated between the lingual artery and the superior thyroid artery, and the superior thyroid artery and ipsilateral lingual artery formed a collateral circulation (Fig. 3). Therefore, the left superior thyroid artery and the right lingual artery were ligated for reducing bleed during the operation. A vertical incision was made in the lower lip, and the mandible was cut and truncated from the middle. The tumors in the submandibular region, mouth floor, bilateral submandibular gland, and sublingual gland were resected. The remaining loosened and dislocated mandibular teeth were removed. The mucoperiosteum was dissociated from the alveolar crest towards the lip and buccal mucosa, and the tongue was resected and reshaped into a “V” shape.

A 1.5-cm section of bone was excised from the middle of mandible body, and the fractured bones were rigidly and internally fixed with a miniplate. Because the mucosa of the mouth floor was tightly adhered to the tumor, it could not be dissected and reserved. Therefore, the expanded ventral tongue mucosa and the mucoperiosteal flap on the mandible body were contrapuntally sutured together to cover the mouth floor. The tumor in the lower lip was resected, and the lower lip received “V” plasty. Then circle sutures were performed around the mandible body to eliminate the dead space. Finally, the incisions at the midline of the lower lip and submandibular region were contrapuntally closed.

One week after the first surgery, the patient was injected with 8 mg pingyangmycin (PYM) into the tumors of the left lips, angle of mouth, and cheek. The injection was given once a week for 2 weeks (Fig. 4, B).

In the second operation, a resection-plasty of the tumor in the left lips and cheek was performed under local anesthesia.

**Fig. 1.** The macroglossia with venous malformation.
(Fig. 4, C). A picture after 2 years’ follow-up after the operation is shown in Fig. 4, D.

Pathologic examination
Specimens were sent for pathologic examination with routine hematoxylin and eosin staining. The red blood cells within the expanded venous sinus from the tongue and mouth floor are shown in Fig. 5, A and B. Fibrosis formation of venous cavity in the lip and cheek after PYM intralesional injection of the venous malformation is shown in Fig. 5, C.

DISCUSSION
The diagnosis and differential diagnosis of deep huge vascular malformation are mainly based on the imaging examinations, such as color Doppler flow imaging, digital subtraction angiography (DSA), CTA, and magnetic resonance angiography.7-12 Many scientists believe that DSA is the gold standard for the diagnosis of vascular malformation, especially huge venous malformation. The application of 3D-CTA provides the diagnosis of huge venous malformation with another important measure of clinical examination. It is not only noninvasive, but also a 3D and direct method to show the venous malformation’s location, size, range, supplying vessels, and relationship with surrounding tissues.7

In the present case, through the 3D-CTA image, it was observed that the venous malformation occupied the tongue, mouth floor, submandibular region, lips, cheek, parapharyngeal space, and lateral skull base. The venous malformation filled the patient’s mouth and even protruded from the mouth, and the pharynx oralis of the upper respiratory tract was almost blocked. The venous malformation in the tongue and mouth floor was supplied by the bilateral lingual artery; the venous malformation in the left parapharyngeal area and ipsilateral skull base was supplied by the left ascending pharyngeal and internal maxillary arteries. The left external carotid artery had been ligated between the lingual artery and superior thyroid arteries; and the superior thyroid artery and ipsilateral lingual artery formed a collateral circulation. Because of long-term outward pressure from the venous malformation, the upper and lower jaw were deformed and kept open, the upper and lower teeth were everted, and the dental arch and jaw were expanded and deformed. In the tongue, mouth floor, cheek, parapharyngeal space, and lateral skull base, there were many round, oval, or irregular “lipiodol pools” formed by expansion of blood sinus and accumulation of blood. The formation of “lipiodol pools” in 3D-CTA may be because the veins in the venous malformation were extremely expanded, forming venous sinuses that accumulated blood. After adding the iopamidol injection contrast medium, iopamidol quickly developed in the venous sinus that directly connected to the supplying vessel, thus creating a circular, oval, or irregular shaped “lipiodol pool.”

Whether the resection of venous malformation in the tongue and tongue reconstruction can be successfully performed depended on the maintenance of airway and the control of massive hemorrhage during surgery. In this case, the upper respiratory tract of the patient was almost blocked; therefore, a tracheotomy was necessary, which facilitated anesthesia and airway management. According to 3D-CTA examination, venous malformation in the tongue and mouth floor were supplied by the bilateral lingual artery, the left external carotid artery had been ligated between the lingual artery and superior thyroid arteries, and the superior thyroid artery and ipsilateral lingual artery formed a collateral circulation. To ensure the surgery being successfully performed, the left superior thyroid artery and the right
lingual artery were ligated and adequate blood for transfusion prepared before surgery.

To control the intraoperative hemorrhage is of vital importance. Preoperative embolism by DSA in the supplying blood vessel can reduce the intraoperative bleeding. But it is reported that embolism of the supplying blood vessel may lead to cerebral embolism, pulmonary infarction, and other corresponding complications.

**Fig. 3.** Three-dimensional computerized tomographic angiography (3D-CTA) images of the macroglossia with venous malformation. Arrows in panels A and B indicate irregular “lipiodol pools”; arrow in panel D indicates the ligation of left external carotid artery. A, B, 3D-CTA images of the hard tissue and vessels of the head and neck. Because of the expansion of the venous malformation, the upper and lower jaws were deformed, the upper and lower teeth were everted and kept open, and both dental arch and jaw bones were enlarged and distorted. The blood supplies of the venous malformation in the tongue and oral floor were provided by the bilateral lingual artery, and the venous malformation in the left parapharyngeal space and lateral skull base were supported by the maxillary artery. There were several round, oval, or irregular “lipiodol pools” formed by venous sinus expansion and blood accumulation in the tongue, mouth floor, cheek, parapharyngeal space, and lateral skull base, indicating a profuse blood supply of the tumor. C, D, 3D-CTA images of venous malformation at tongue, lateral cranial base, parapharyngeal space, and cervical vessels. The venous malformation in the tongue was nourished by the bilateral lingual artery. The left external carotid artery has been ligated between the lingual artery and superior thyroid artery. The superior thyroid artery and ipsilateral lingual artery formed a collateral circulation. It was seen that round, oval, or irregular “lipiodol pools” varied in size in the tongue and mouth floor.
complications with an incidence of 5%-8%. In the present case, the patient refused to get a DSA examination and embolism owing to financial problems. Another method to reduce intraoperative bleeding is intraoperative ligation of the supplying blood vessel. However, it is not appropriate to only simply ligate the supplying blood vessel of venous malformation. Because blood supply is rich in the oral and maxillofacial region, ligation of the feeder blood vessel may cause the venous malformation to build a more extensive collateral circulation that makes the venous malformation harder to handle. The present patient was a good example of this situation. In the earlier surgery, the left external carotid artery was ligated, resulting in a further enlargement of the venous malformation rather than bringing it under control. No matter what method is used to control the hemorrhage, a sufficient volume of blood should be prepared before surgery. To stop hemorrhage and continue the surgery, the left superior thyroid artery and the right lingual artery were ligated, and the patient was transfused with a total volume of 3,000 mL blood during the operation. In summary, preoperative selection of surgical indications, a fully perioperative preparation, and specific and feasible surgical measures are the most important safeguard for a successful operation of huge venous malformation.

The first 2 measures were used to ensure the safety of the patient. Resection of the venous malformation in the tongue and the mouth floor can release the patient from the unexpected life-threatening danger. Glossoplasty was important in resuming the shape and function of the tongue, and finally work ability and a normal life. During the surgery, the lower lip and mandible were cut and truncated from the middle line. After removal of the remaining loosened and dislocated mandibular teeth, the mucoperiosteum was cut apart from the top of alveolar crest at the posterior molar to the middle incision. Then, the mandibular stump was clamped to dissociate the mucoperiosteum and expose the mouth floor and submandibular region. Finally, the venous malforma-

Fig. 4. The patient with macroglossia and venous malformation after surgical treatment: A, 1 week after the first surgery; B, after intralesional injection of pingyangmycin; C, after the second surgery; D, follow-up 2 years after the operation.

Fig. 5. Pathologic examination of the patient of macroglossia with venous malformation. (A, B) Red blood cells within the expanded venous sinus from the tongue and mouth. (C) Fibrosis formation of venous cavity in the lip and cheek after PYM intralesional injection of the venous malformation.
tion in the mouth floor and submandibular region was bluntly and sharply isolated and resected. Because the boundaries between the venous malformation and surrounding tissues were not clear, the bilateral submandibular glands and sublingual glands were simultaneously resected, but efforts were made to protect the lingual nerve and hypoglossal nerve as much as possible. Resection of venous malformation in the tongue and glossoplasty adopted the procedure of “V”-shaped resection and tongue reconstruction to retain enough length of the tongue. Thus, the patient could resume the shape and function of the tongue.

Coverage of the mouth floor is a very important step. In this patient, it was impossible to preserve the mucosa of the mouth floor, because it adhered to the venous malformation. To overcome this problem, the expanded tongue ventral mucosa and the mucoperiosteal flap at the alveolar crest and the lip and cheek were contrapuntally sutured together to cover the mouth floor and pressed, fixed, and shaped with iodoform gauze.

Because of the swelling of the tongue, the upper and lower jaws were abnormally developed, showing eversion and opening of the upper and lower teeth. According to the measurement and analysis of the upper and lower jaw in the 3D-CTA image, 1.5 cm of bone was cut off from the middle of the mandible, so that the expanded mandible could be largely back to normal, providing a basis for dental implantation in the future.

The explicit mechanism of PYM for the treatment of vascular malformation is still unclear. From the chemical aspect, it is similar to bleomycin with antitumor and antiangiogenic effects. Intravenous or intralesional injection of PYM could cause injury and detachment of endothelial cells, thickening of the vessel wall, and narrowing or occlusion of the lumen. Some researchers 18-20 have also demonstrated that intralesional injection of PYM is an effective method for the treatment of vascular malformation.

In conclusion, 3D-CTA shows great value in clinical diagnosis of huge venous malformation. It not only has the advantage of being noninvasive, but it also provides 3D and direct images to present venous malformation location, size, range, supplying vessels, and relationship with surrounding tissues. The specific 3D-CTA image for the huge venous malformation is that there are many “lipiodol pools” formed by venous sinus expansion and blood accumulation. 3D-CTA can provide clinical evidences for the surgical risk assessment of huge venous malformation.

The macroglossia caused by such a huge venous malformation is rather rare. The difficulty and risk of the surgery are also great. A complete preoperative imaging examination and evaluation should be conducted to design a personalized surgical plan to achieve the desired therapeutic effect. The biggest risk of the surgery for the huge venous malformation is intraoperative massive hemorrhage and respiratory tract obstruction. Preoperative embolism by DSA and intraoperative ligation of the supplying vessel are important strategies for reducing intraoperative hemorrhage. The choices of surgical indications, adequate perioperative preparation, and specific feasible surgical measures are most important for a successful operation of huge venous malformation.

REFERENCES


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