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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

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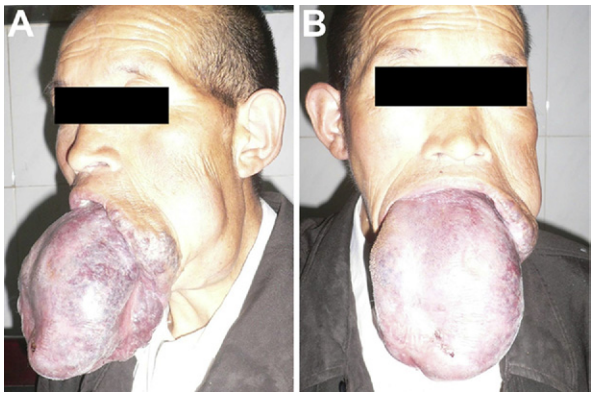

Fig. 1. The macroglossia with venous malformation.

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 $\times$ 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