Клінічні міркування / *Doctor's thinking*

Том 9, № 3–4 • 2024

PainMedicine Journal

Медицина Болю

www. painmedicine.org.ua

Міждисциплінарний • Науково-практичний журнал

DOI: 10.31636/pmjua.v9i3–4.6

First stage reconstruction following surgical excision of a large arteriovenous malformation of the lower lip and tongue

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Abstract. *Arteriovenous malformations (AVMs) are rare congenital lesions caused by deformity of the blood vessels. Around half of the AVMs are found in the craniofacial area, presenting with a progressive high-flow benign vascular lesion. Larger lesions involving the facial region may cause extensive deformities, detrimental functional disturbances, and a substantial risk of unexpected hemorrhage and airway obstruction. Patients may develop feeding difficulty early in childhood in cases involving large lesions in the intraoral structures, such as the tongue and the lip. Here we report a challenging case of arteriovenous malformation of an adolescent male's tongue and lower lip. The diagnosis was confirmed by computed tomography (CT) angiography revealing malformation of the inferior labial artery and branch of the external carotid artery. Treatment involved surgical intervention, with initial excisional surgery, which significantly reduced the lower lip lesion's size, followed by immediate reconstruction. The outcome of this initial surgery revealed a significant size reduction of the lesion of the lower lip, allowing for better alimentation, preventing potential airway obstruction, and resulting in an acceptable aesthetic improvement. Subsequent treatment is planned to reduce further the remaining lesion of the lower lip in a second-stage excision, as well as excision of the tongue AVM. The wide variations and severity of arteriovenous malformation in the facial region necessitate a tailored treatment plan for each case. Strict angiographic follow-up strategies may be needed considering its high recurrence rate.*

Key words: *Arteriovenous malformation, tongue, lower lip, vascular malformation, vascular anomaly.*

Introduction

Arteriovenous malformations (AVMs) are complex clinical conditions that challenge their resolution. They are rare vascular anomalies caused by angiogenesis disorder, characterized by abnormal communication between the affected arteries and veins. In this case, there is the absence of a capillary network that usually connects the artery and vein [1]. This bypass of a capillary network disrupts the normal blood flow and may have a variable clinical presentation. The lesion which results from AVMs can cause both functional and cosmetic impairment, primarily due to its unpredictable evolution [2].

Arteriovenous malformations are high-flow anomalies and account for around 6 % of all vascular malformations [3]. The location in which AVMs develop can be intracranially or extracranially. Collective studies show that cerebral AVMs account for an incidence of 0.69–1.32 per 100 000 population [4]. It concerns the high risk of intracranial hemorrhage and other neurological implications such as chronic headache, seizure, and neurological impairment [4]. On the other hand, the most common extracranial AVMs occur in the head and neck region, which accounts for half of all AVMs. Most head and neck AVMs affect the midfacial region, such as the cheek, nose, upper lip, and ear [5, 6].

The development of AVM starts during embryogenesis, and the absence of a capillary network leads to the direct shunting of blood from the feeding artery to the draining venous vessel through abnormal channels known as nidus [6]. This shunting eventually causes a reduction of capillary oxygen delivery to the tissue, subsequently causing ischemia. It is crucial to distinguish AVM from vascular tumours such as haemangioma. Untreated, these fast-flow arteriovenous malformations can cause concerns of ulceration, hemorrhage, and even right heart failure as a result of the chronic artery to venous shunting.

Case Presentation

A 16-year-old male patient presented to the Plastic Surgery clinic with an enlarged lower lip and tongue mass. The swellings were first observed since birth and gradually increased in size. The mass was initially reddish and had developed into purplish discoloration. The patient claimed warmth in the lesion area, with previous ulcerations and episodes of unprovoked bleeding, but denied any pulsatile sensation. According to the patient, the size of the mass had grown to an extent which caused difficulty during eating and difficulty in closing the mouth, causing drooling of saliva. The patient denied any pain or associated fever. History of tumour or similar illness in the family was denied.

The general status examination of the patient was unremarkable. However, upon examination of the facial region, a mass of the lower lip with a size of 8 cm by 4 cm, reddish-purple discoloration, soft consistency, irregular surface, and a clear border was revealed. There was also swelling of the tongue with reddish-purple discoloration, soft consistency, irregular surface, and poorly defined border. On palpation, both lesions were warm to the touch. However, no tenderness was observed. No prominent pulsation was palpable. On auscultation, no bruit was heard on the lesion. The patient was underweight, with a body mass index of 15. These findings may have been related to the difficulty in feeding due to the enlarged mass.

The patient had previously consulted a general surgeon and underwent CT angiography, which revealed malformation, with the lower lip receiving vascularization from the labial artery and the tongue receiving vascularization from branches of the external carotid

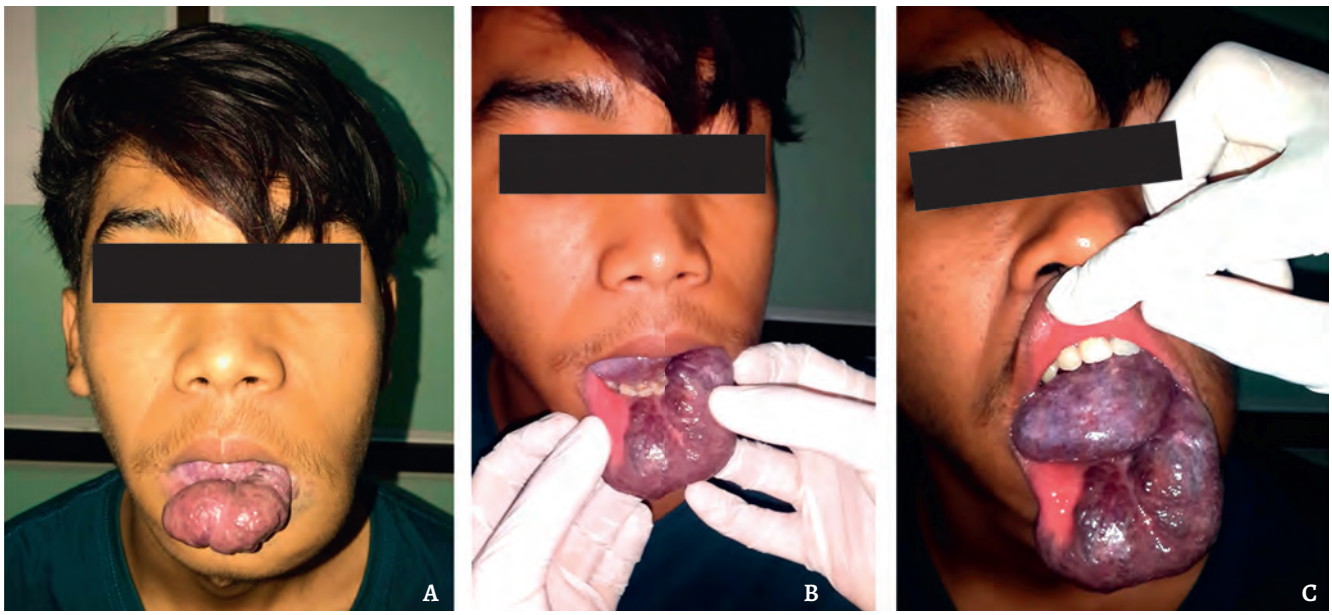

Figure 1. A 16-year-old male presenting with large arteriovenous malformations (AVMs) of the lower lip and tongue. (A) Extraoral view. (B–C) Intraoral view

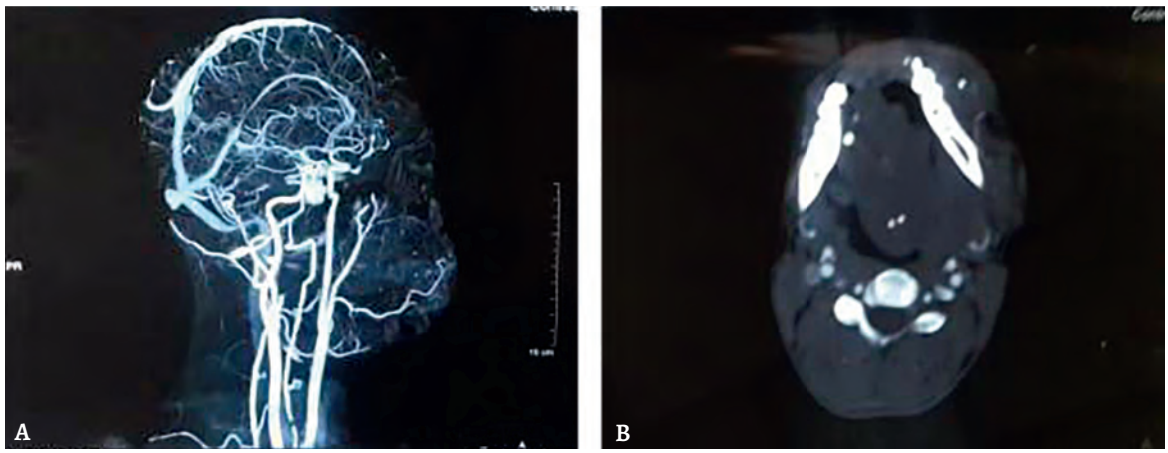

Figure 2. Radiographic examination of the head and neck region. (A) Computed tomography (CT) angiography. (B) Axial CT

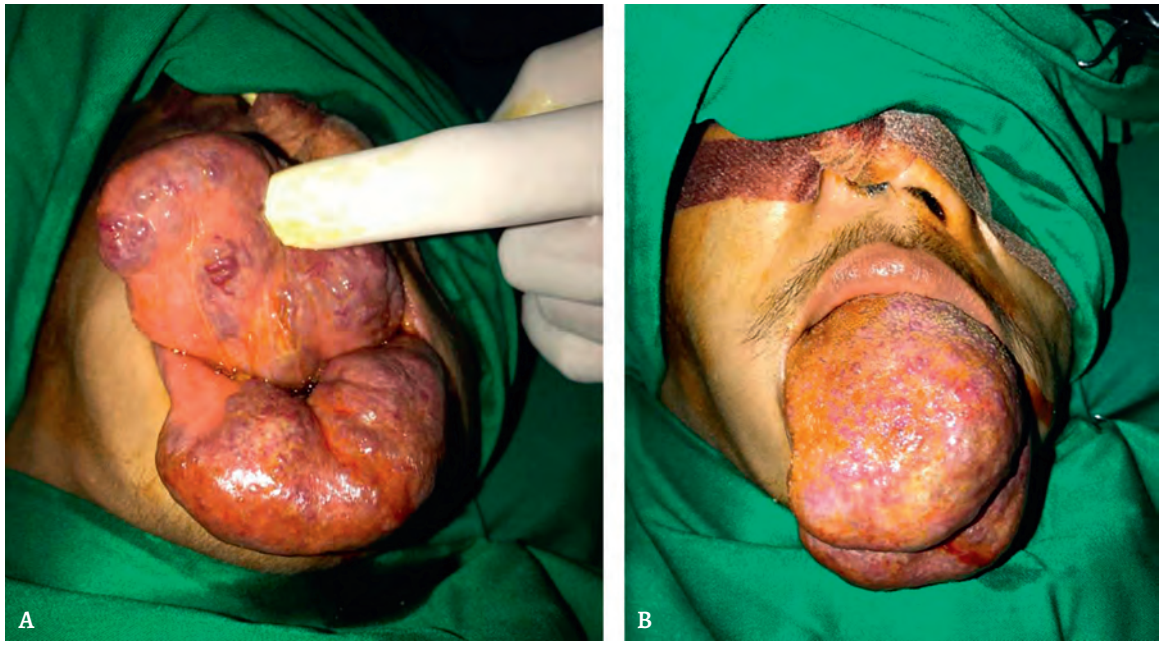

Figure 3. Intraoperative images showing the two separate lesions on the tongue causing macroglossia, and the lower lip

artery. An arterial aneurysm was observed at 1.6 cm × 1.58 cm towards the posteroinferior side. The patient had yet to receive any treatments prior to the referral.

The patient underwent surgical excision and reduction of the lesion on the lower lip, followed by immediate reconstruction. Hemorrhage was controlled by careful hemostasis technique throughout the procedure. Postoperatively, the lesion size was significantly reduced to 3.5 cm by 1.5 cm. No postoperative infection occurred related to the procedure. At eight months follow-up, no complication, such as facial muscle paralysis, numbness, or wound dehiscence, was recorded. The patient could comfortably close his mouth, and the saliva drooling subsided. Histopathology confirms the lesion as an AVM. However, due to incomplete excision during the first surgery, a second-stage excision is planned for further reduction or, if possible, complete resection.

Discussion

Head and neck AVMs can be challenging in their management, and understanding their characteristics helps in early diagnosis and prompt treatment. The oral cavity can involve the tongue, buccal mucosa, lip, palate, and gingiva. In this case, intraoral AVM was found in the lower lip and tongue. The lip accounts as a critical function-

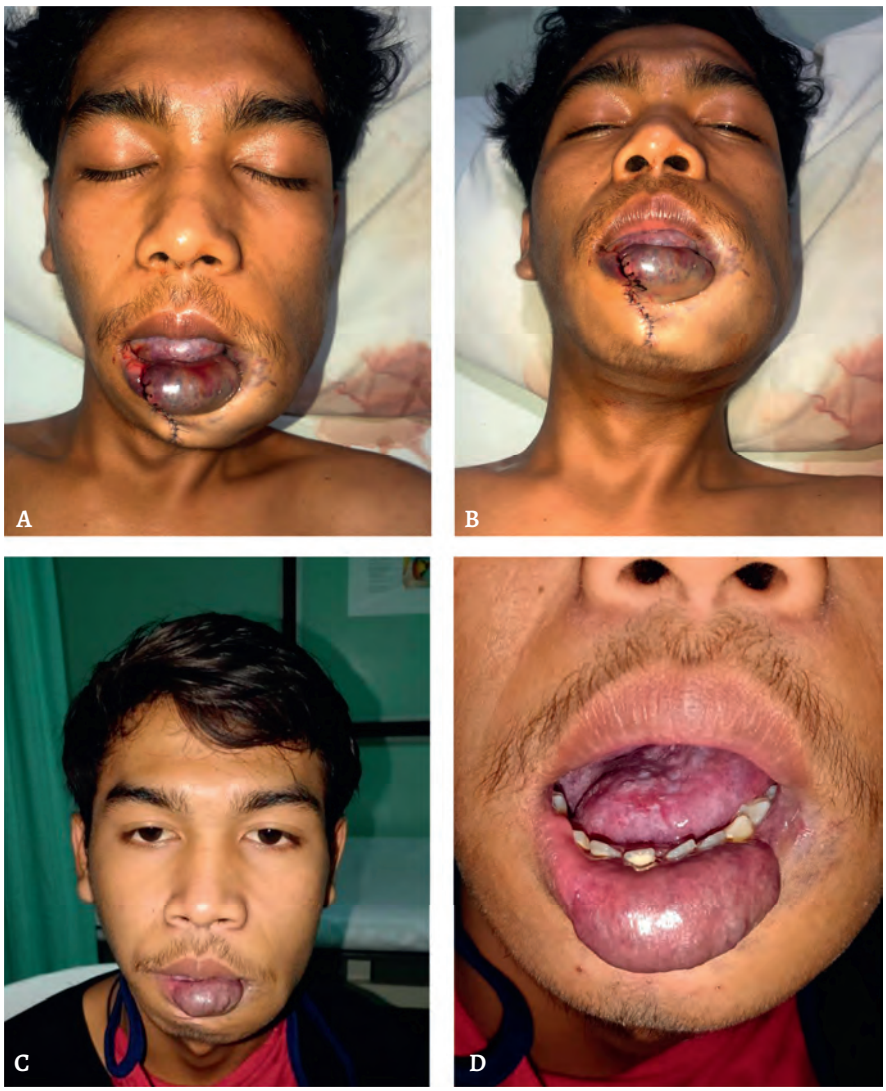

Figure 4. (A–B) Postoperative photographs after excision and reduction of lower lip AVM followed by immediate reconstruction. (C–D) Eight months follow up visit

al and aesthetic unit of the facial region, while tongue swelling may cause alimentation problems or even risk of aerodigestive obstruction. Therefore, early management is needed as lesion expansion may cause compression and eventually disrupts the surrounding tissues.

It is essential to differentiate AVMs and other vascular anomalies, such as hemangiomas commonly found in congenital vascular tumours that develop due to abnormal proliferation of endothelial cells that may involute spontaneously [5]. The detection and evaluation of AVMs characteristics are commonly done by utilizing Doppler ultrasound, conventional angiography, or computed tomography (CT) arteriography. Computed tomography (CT) can be employed to evaluate any involvement of bone components. Magnetic resonance imaging (MRI) has been used extensively to evaluate the adjacent soft tissue further if necessary [7]. The patient did not undergo further examination beyond the CT angiography from the previous hospital. Although MRI evaluation would be valuable additional information on the vasculature characteristic and soft tissue involvement, the high cost of this modality became a consideration factor. Therefore, we utilized CT with contrast as the primary imaging for presurgical planning.

The main goal of AVM surgical treatment is to eradicate the nidus and restore normal vasculature in the affected area. Several treatment options have been developed for managing vascular malformations, such as sclerotherapy, laser therapy, endovascular embolization, and surgical excision. With sclerotherapy, transcutaneous injection uses substances that induce endothelial destruction, resulting in thrombosis of the vascular

space. It is suitable for localized lesions that have undergone prior embolization treatment. Examples of sclerosing agents are sodium tetradecyl sulfate (STS), doxycycline, bleomycin, and absolute ethanol [5, 8]. However, a disadvantage related to sclerotherapy is the high flow characteristic of AVM which can wash out the sclerosing agent [5].

With embolization, an embolic agent is delivered using the endovascular catheter to an area proximal to the AVM to block the vascular flow. Ideally, it would fill the nidus and close the AVM. Improvement of symptoms can be expected even if the size of AVM is not significantly reduced. However, re-expansion commonly occurs due to remaining lesions. For example, embolic materials commonly used are n-butyl cyanoacrylate (n-BCA), ethanol, polyvinyl alcohol (PVA), gel foam powder, and coils. Some complications that may occur from embolization are the growth of new collateral vessels, tissue necrosis, thromboembolism, and nerve damage [5].

The photocoagulation capability of laser therapy has also recently been used as a modality to manage vascular lesions. Diode lasers are useful to de-vascularize AVM lesions, with a wavelength between 800 nm and 980 nm. A combination of laser therapy and embolization can also be of choice depending on the different characteristics of the lesions [9].

Surgical resection by itself has commonly opted for small, well-localized AVMs. Its recurrence rate is lower than that of embolization alone [8]. However, surgical excision or resection bears the risk of intraoperative hemorrhage and disruption of adjacent tissues. Commonly, the choice treatment consists of a combination of preoperative endovascular embolization followed by surgical excision [10]. Some lesions that cannot be accessed for embolization are commonly directly excised [8].

Additionally, a common complication following resection is a recurrence of the lesion. A report by Igari et al. showed less intraoperative blood loss in thigh AVM treated with preoperative embolotherapy prior to resection [11]. In addition to the benefit of reduced bleeding during surgery, embolization can also be considered to reduce symptoms, namely congestive heart failure, reducing the extent of surgery, and preventing recurrent bleeding episodes [12]. In our patient, consideration was taken to excise the lesion on the lower lip, followed by immediate reconstruction. Surgical excision is suitable in this case to correct the focal deformity. High-flow malformations such as AVMs tend to proliferate during puberty, with low chances of involution [13]. No embolization was performed prior to surgical excision in our patient. Instead, thorough presurgical planning was prepared, and intraoperative hemorrhage control was done with care. The amount of intraoperative bleeding was 200 ml, and no blood transfusion was needed during or after the procedure. Keshelava et al. previously performed an excision of a giant craniofacial AVM without embolization therapy due to multiple feeding arterial branches, showing satisfying results with no complications such as neurological deficit and no recurrence after four months [14]. Phillips et al. showed similar clinical results in patients with embolotherapy followed by surgery compared to surgery alone [15]. No ligation of the proximal feeding artery of the AVM was done, as it would stimulate the growth of collateral arteries and enlarge the lesion. Additionally, if therapeutic embolization is deemed necessary in the future, it would not be possible [8].

Several complications may occur after surgical excision of AVMs, such as muscle paralysis or numbness in the affected area, postoperative infection, hemorrhage that requires blood transfusion, and wound dehiscence [8]. In our patient, no severe complications and morbidity were recorded postoperatively. Although the size of the lesion on the lower lip has been significantly reduced, the patient is scheduled for a second-stage excision for further reduction to achieve a satisfying functional and aesthetic outcome. With large or diffuse lesions, the patient must understand the potential need for a second surgery and the risk of recurrence [8]. The macroglossia, due to the underlying AVM, disrupts the patient's mastication, speech, and swallowing ability. Richter et al. reported satisfying results after surgical excision of isolated tongue AVMs without embolization, while cases of advanced tongue AVMs needed a combination of embolization and surgery [16]. In our case, the two lesions have two different feeding arteries separate from each other. Thus, the decision to excise the lesion on the lower lip as a first-stage procedure without embolization was deemed suitable, considering the lip area allows for easily accessible feeding vessels and proper hemorrhage control done more confidently. Subsequent surgical resection of the tongue AVM may be feasible; however, the possibility of the need for embolization must be considered based on a thorough evaluation of the CT angiogram. The ultimate goal would be total resection. It was shown that patients with no recurrence within five months postoperatively are less likely to experience it long-term. However, within ten years, the risk of re-ex-

pansion is around 5.2% (17). It can be explained by the growth of collateral vessels and new vessels surrounding the primary lesion, as well as dilation of the vessels, due to proangiogenic activities induced by both surgical and endovascular procedures.

Conclusion

Considering the unpredictable clinical evolution, AVMs pose a significant challenge to physicians and surgeons in choosing the most suitable treatment option to optimize outcomes. Our experience is that surgical excision of the lesion may be completed without embolization in conditions where proper hemostasis can be performed with sufficient surgical experience. We recognize the need for continuing close follow-up for intraoral AVMs as such with other head and neck vascular malforma.

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Перший етап реконструкції після хірургічного видалення при великій артеріовенозній мальформації нижньої губи та язика

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Загальна лікарня Сангла Балі, Індонезія

Анотація. *Артеріовенозні мальформації (АВМ) — рідкісні вроджені ураження, спричинені деформацією кровоносних судин. Близько половини АВМ зустрічаються у черепно-лицевій ділянці, проявляючись як прогресуюче доброякісне ураження судин із високим кровотоком. Великі ураження ділянки обличчя можуть спричинити значні деформації, серйозні функціональні порушення та суттєвий ризик несподіваної кровотечі й обструкції дихальних шляхів. У пацієнтів можуть виникнути труднощі з годуванням у ранньому дитинстві у випадках великих уражень внутрішньоротових структур, таких як язик і губи. У статті ми описуємо складний випадок артеріовенозної мальформації язика та нижньої губи у підлітка чоловічої статі. Діагноз підтверджено за допомогою комп'ютерної томографії (КТ) з ангіографією, яка виявила мальформацію нижньої губної артерії та гілки зовнішньої сонної артерії. Лікування включало хірургічне втручання, первинне висічення ураження, що значно зменшило розмір ураження нижньої губи з подальшою негайною реконструкцією. Результати первинної операції показали значне зменшення розміру ураження нижньої губи, що дозволило полегшити годування, запобігти потенційній обструкції дихальних шляхів і забезпечило прийнятне естетичне покращення. Подальше лікування планується для додаткового зменшення залишкового ураження нижньої губи під час другого етапу висічення, а також видалення АВМ язика. Широкі варіації та тяжкість артеріовенозної мальформації в ділянці обличчя вимагають індивідуального плану лікування для кожного випадку. Зважаючи на високу частоту рецидивів, можуть знадобитися суворі стратегії подальшого ангіографічного спостереження.*

Ключові слова: *артеріовенозна мальформація, язик, нижня губа, судинна мальформація, судинна аномалія.*