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MISCELLANEOUS

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Arteriovenous malformation of the floor of the mouth: a case report

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Abstract Arteriovenous malformations of the head and neck are rare lesions with unclear pathogenesis. They usually present during childhood, growing proportionately to the child. Although preoperative superselective embolization followed by surgical resection is the treatment of choice, complete removal is often not feasible, leading to high recurrence rates. The case of a patient with an arteriovenous malformation of the floor of the mouth diagnosed late in her adulthood and its management are presented.

Keywords Arteriovenous malformations · Embolization · Head and neck · Surgery

Introduction

Various classifications of congenital vascular lesions had been suggested before 1982, when Mulliken and Glowacki presented a classification based on clinical and biologic behavior of these entities [1]. They formulated two entirely distinct groups of congenital vascular anomalies: hemangiomas and vascular malformations. Hemangiomas, though not obvious at birth, are usually of clinical importance in the 1st month of life [2]. They are most commonly found in the head and neck, starting as flat erythematous macules. Hemangiomas grow to considerable size in the first 2 years of life, and after a stable phase they involute to various degrees [2].

Vascular malformations are further subclassified according to the vessels involved: capillary, venular (port wine stains), venous, lymphatic or mixed (arteriovenous or venous-lymphatic) [3, 4]. The majority of vascular lesions presenting in the oral cavity and maxillofacial area are malformations. Vascular malformations are present at birth, though they are not always apparent. They usually undergo a slow hypertrophy without involution [4]. Rapid growth has been reported to be associated with periods of hormonal modulation such as puberty or pregnancy. Bone involvement occurs in 35% of patients with vascular malformation [5]. Arteriovenous malformations (AVM) may change rapidly owing to high flow with shunting, making them potentially destructive [5]. Of all vascular malformations, AVM is the most difficult to manage, since the only chance for cure is complete resection [4]. We present the case of a patient with an AVM located in the floor of her mouth, diagnosed late in her adulthood.

Case report

A 52-year-old female patient was admitted to our department with the diagnosis of a congenital vascular malformation located on the left side of the floor of her mouth. The malformation was asymptomatic for years. The patient reported a port wine stain on her left auricle that she had had since she was an infant, which had been managed successfully with an argon laser a year before. On admission, she complained of a stabbing pain in the submandibular area on the left, as well as swelling in the floor of the mouth on the same side, causing dysphagia and dysarthria, which had been aggravated during the last 12 months. Clinical evaluation revealed a mass located in the floor of the mouth on the left with dilated tortuous vessels. On palpation a thrill was felt over the mass. There was no sign of infection, bleeding or involvement of the oral cavity mucosa. Magnetic

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resonance imaging showed a multi-lobulated mass located in the floor of the mouth on the left side, which invaded the left mylohyoid muscle and displaced the left genioglossus muscle medially (Fig. 1). The major part of the lesion consisted of tortuous dilated veins with thrombi formation. Color Doppler revealed a high-flow malformation. Arteriogram and coil embolization of the left submental artery was performed the day prior to surgery (Fig. 2). The feeding artery was opacified during angiography and superselectively catheterized. A metallic coil (MREYE, Cook, Bjaeverskov, Denmark) of 30-mm length and 40-mm diameter was advanced through a 5-French catheter (Fig. 3). An incision was made about 3 cm beneath the mandible on the left, over the submandibular gland, and extended towards the chin. The lesion was excised after removal of the left submandibular gland and resection of the left mylohyoid and genioglossus muscles. The lingual and hypoglossus nerves were preserved. No blood transfusion was needed during surgery or the postoperative period, which was generally uneventful. The patient was discharged on the 4th postoperative day. Two years after surgery, there is no clinical or radiological evidence of recurrence (Fig. 4).

Discussion

AVMs are abnormal congenital communications between arteries and veins bypassing the normal capillary bed [4]. They have recently been classified with arteriovenous fistulas as high-flow vascular malformations. AVMs are present at birth, although they may not become evident until adolescence or early adulthood, and they persist throughout life [1, 4]. There is no gender predilection. The patients present with deformity and the complaint of "heaviness" in the area of the malformation [6]. Pain occurs in 50% of patients. AVMs may be found in facial muscles with the most common site being the masseter and temporalis muscle; they can be confused with parotid masses, and headache is the patient's main symptom. Involvement of the tongue, especially when there is a significant arterial component, results in major functional and aesthetic problems as well as bleeding and ulcerations [4]. Bleeding, infection and sudden enlargement often follow a direct trauma to the malformation [6]. Some malformations result in overgrowth of the facial skeleton and hypertrophy of the adjacent soft tissue caused by the increased blood supply [4, 5]. On clinical examination, there is always a strong pulsation to the mass, and a palpable thrill may be felt

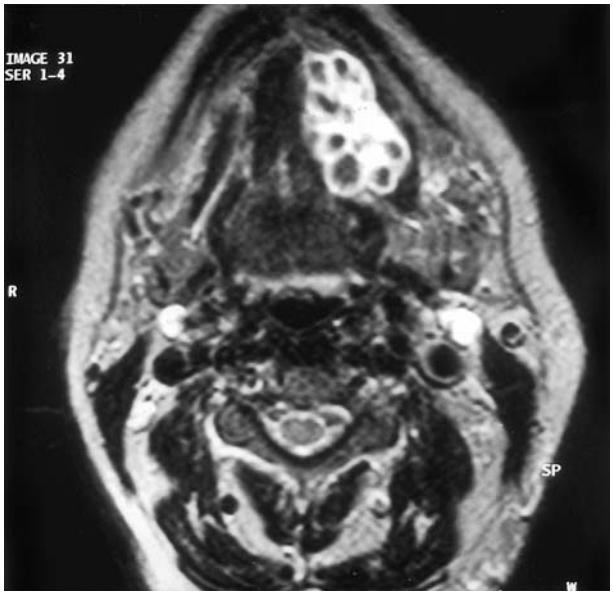

Fig. 1 Pre-embolization MRI, axial T2-weighted image, demonstrates a multilobulated mass in the left submandibular space, with well-defined margins. There is marked displacement of the adjacent mylohyoid muscle

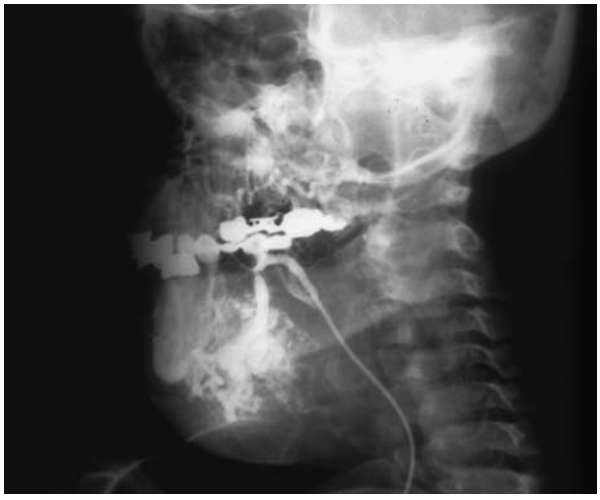

Fig. 2 Superselective catheterization of the left submental artery. The arteriovenous malformation in the submandibular space is confirmed

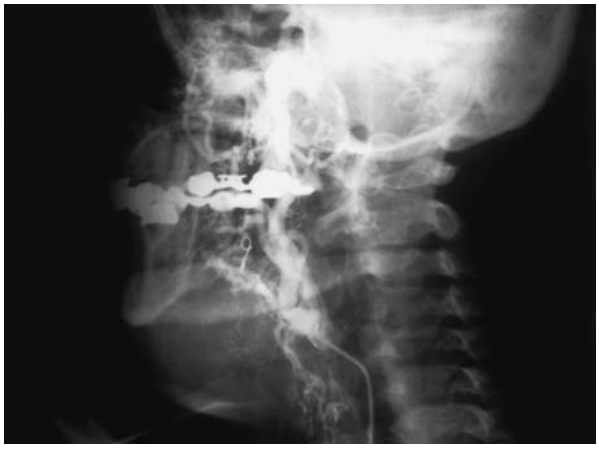

Fig. 3 A metallic coil was advanced through the 5-French catheter. Post-embolization angiography revealed a satisfactory result

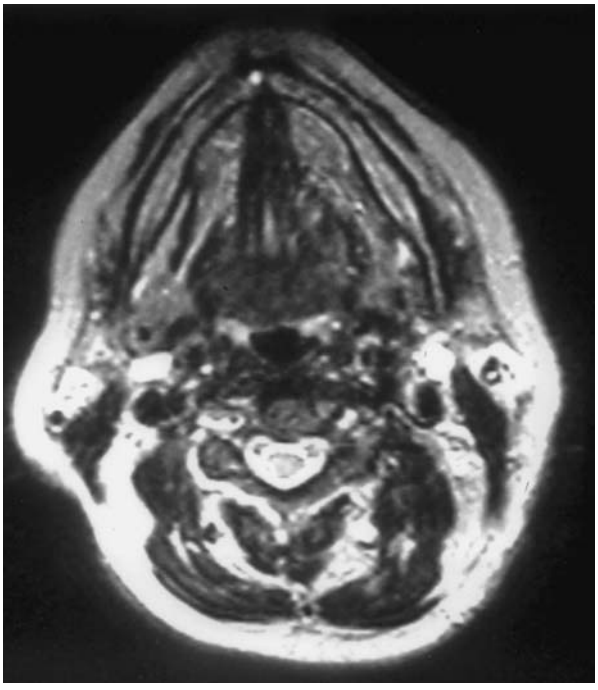

Fig. 4 Post-surgical MRI, axial T2-weighted image, reveals complete removal of the vascular lesion

over it. Dilated tortuous veins are usually noted, and sometimes a fibrotic change on the overlying skin is present because AVM shunts blood from skin. In advanced disease ulceration of the skin and bleeding may be noted [4, 6].

Arteriovenous communications consist of multiple microfistulous tracts, called the nidus [4]. Histologically, the nidus shows a heterogeneous mixture of vessel types, including tortuous arteries and dilated veins with thickened walls, dispersed within a complex background of numerous smaller arteries and veins. The AVM expands through hemodynamic changes that result from the shunting of the blood. For many years, AVMs may be low-flow lesions, but eventually they become high-flow and expansion becomes very rapid [6].

Differential diagnosis by histology alone is often impossible, and the establishment of the correct diagnosis needs clinical evaluation and imaging techniques. Computed tomography demonstrates bone structures involvement as well as the relationship of the lesion to the adjacent tissues. Magnetic resonance imaging is the most informative technique for the demonstration of both the extent of involvement and the rheological characteristics [3, 4, 7]. Gray-scale ultrasonography coupled with color Doppler has the advantage of providing a rapid, relatively inexpensive and noninvasive assessment of lesion morphology and vascular components [8, 9]. Some high-flow AVMs with rapid growth within the 1st year of life are very destructive and may be confused with hemangiomas, but they do not respond to steroids or vincristine as hemangiomas do.

It is of great importance to estimate whether a lesion in a child is a hemangioma or a malformation. Hemangiomas involute spontaneously. Steroids, vincristine or interferon-α-2a have been used to speed up involution or for patients with life-threatening lesions [3, 4]. There are limited indications for surgical resection such as obstruction of the visual axis, large hemangiomas with thrombocytopenia, obstruction of luminal structures or uncontrollable infection, bleeding and ulceration [3].

Vascular malformations are characterized by a lack of involution. They usually present a slow and steady growth caused by the progressive dilatation of preformed vascular channels [4, 6]. Once the diagnosis is made, therapy should not be delayed, because the AVM only becomes larger with time. Embolization alone as well as surgical ligation without resection, should be performed only for emergency situations or as palliative procedure since it lasts days to weeks because of the rapid development of new blood supply. When the therapy chosen is embolization alone, the uniformly sized microparticles are used to deposit an inert substance deep within the network of the lesion itself. Although good results have been reported with embolization therapy, most authors agree that the treatment of choice is embolization followed by surgical resection [10]. An experienced interventional radiologist should perform embolization therapy, since it carries severe risks. Different embolization materials have been recommended: autogenous muscle, fibrin, isobutyl-2-cyanoacrylate, polyvinyl alcohol particles and microcoils [3]. We used a metallic coil in our case. Their use for intravascular embolization was introduced in 1975 [11]. They are inserted through proximal vessels or directly into the vascular malformation. Platinum coils wrapped with Dacron strands are commonly used. They decrease blood flow and increase turbulence in an AVM leading to clot formation. An arteriogram and embolization of the malformation should be performed 1 to 3 days before surgery [4]. Preoperative embolization is carried out to reduce the risk of intraoperative bleeding, though some patients have considerable blood loss during surgery. Sometimes an AVM is so extensive that complete resection is not feasible without leaving a devastating defect. Sclerotherapy is used with pure alcohol for AVMs that are unresectable, but may cause complications such as pain, swelling and nerve injury [4].

Despite the fact that all vascular malformations are, by definition, present at birth and AVMs become clinically evident before the age of 30, our patient was asymptomatic until her early 50s. Her main complaint was swelling in the floor of her mouth on the left, causing progressively aggravated dysphagia and dysarthria starting a year before admission. No history of infection or bleeding was reported. There was no trauma history around the lesion, which could have made the diagnosis of AV-fistula possible. MRI showed concentric rings, which most likely represent thrombus formation in the early phases of phleboliths, which are seen in venous malformations. However, the malformation had

as feeder vessel the submental artery, which showed an unfamiliar course probably because of displacement from the lesion. The patient was successfully managed in our department with the combined therapy of elective embolization and surgical resection. She was followed-up every 6 months for the first 2 years postoperatively without any evidence of recurrence. The evaluation will continue annually for the next 3 years in order to complete a total follow-up of 5 years.

Although congenital vascular abnormalities of the head and neck are relatively rare, hemangiomas are the most common tumor of infancy; thus, it is mandatory for the specialist to establish the correct diagnosis in order to manage these lesions with the appropriate individualized treatment. Decisions must be made about the kind of treatment and the proper time of management. It is widely accepted that as soon as an AVM is diagnosed, it is best treated, when possible, with total resection after elective embolization.

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