Management of extracranial congenital arteriovenous malformations of the head and neck

Report of five cases

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Five cases of congenital extracranial arteriovenous malformations are reported. The need for careful preoperative definition of all feeding vessels by selective arteriography is stressed. The radiographic phenomenon of “washout” may preclude the recognition of some arterial feeders at the time of the initial contrast studies. The surgical management of these patients using repeated selective angiography, embolization, and ligation of the major arterial supply, followed by total excision of the malformation, is described.

KEY WORDS: extracranial, arteriovenous malformation, selective angiography, radiographic washout, surgical management

EXTRACRANIAL arteriovenous (A-V) communications may be divided into two categories. The first, traumatic in origin, should be referred to as arteriovenous fistula; it is by far the most common group. The second category, congenital arteriovenous malformations, result from abnormal remnants of embryologic vascular communications. It is this less common anomaly that we discuss in this report.

In a recent review of the literature, Lawson and Newton documented only 18 cases of congenital A-V malformations involving the extracranial carotid-vertebral circulation. Many of these were incompletely studied angiographically, and the full complexity of the arterial supply remains unknown. They added three cases of their own, all of which were examined with selective four-vessel angiography.

Of 23 cases reported, only 11 were treated surgically. Not all of these lesions were totally removed; some recurred, and surgical deaths have been reported. The remaining 12 lesions were not operated on because symptoms were minimal or the lesions were considered too extensive for total removal.

The purpose of this report is to present experience with five cases of congenital extracranial A-V malformations and to describe the evolution of our approach to the surgical management of these patients.

Case 1
This 53-year-old woman was admitted...
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with a chief complaint of a rumbling noise in the right ear, throbbing headache and feeling of pressure behind the right mastoid. The symptoms had started abruptly 5 months earlier and had been progressive. There was no history of trauma. The general physical and neurological examinations were normal except for a machinery-like bruit heard over the right mastoid which was decreased but not abolished by carotid compression.

A right common carotid arteriogram demonstrated an A-V malformation in the right suboccipital region, fed from the right occipital branch of the external carotid artery (Fig. 1). Attempts to fill the lesion from the left and right vertebral and left carotid arteries with and without compression of the right carotid were unsuccessful. The patient refused excision of the fistula because of the possible cosmetic deformity imposed by removal of the suboccipital muscles. She did accept an alternative procedure which included muscle embolization and ligation of the major arterial feeder. She has been free of symptoms or objective bruit for 2 years.

Case 2

This 24-year-old man was admitted with the complaint of a faint buzzing in the right ear. He had been well until 2 weeks earlier when he suffered a mild cerebral concussion and hyperextension injury of the cervical spine. The general physical and neurological examinations were normal except for a bruit heard just behind the right mastoid tip. Plain x-ray films of the cervical spine showed enlargement of the foramen transversarium of C-1, suggesting an enlarged vertebral artery. The first series of angiograms in August, 1967, demonstrated an extensive A-V malformation in the right suboccipital

![Fig. 1. Case 1. Oblique subtraction views, arterial phase, showing the malformation (large black arrows) being fed by the occipital branch of the external carotid (small black arrow) and the early draining jugular vein (large open arrow).](image)
region being fed by the right vertebral, external carotid, and thyrocervical trunk (Fig. 2). There was no filling demonstrated from the left carotid or vertebral systems except for a left-to-right vertebral steal (Fig. 3). Because of the lack of symptoms he was discharged.

Approximately 1 year later the patient was readmitted with increasing bruit, headache, nystagmus, unsteady gait, and fatigability. Repeat angiography confirmed the malformation and the primary feeders, but also clearly demonstrated an increase in the left-to-right vertebral steal. Excision of the malformation was not possible because of the enormous venous channels and a false aneurysm seen on serial angiograms (Fig. 4). Using our experience of muscle embolization and ligation of feeder vessels from the previous case, we undertook an initial procedure to resolve the vertebral steal by clipping the right vertebral artery extradurally at C-1, embolizing it with muscle packs from below, and ligating the vessel at C-6. We intended to obtain further angiography to identify additional arterial feeders before attempting a total excision. The patient, however, free of the bruit, tinnitus, and unsteadiness, refused follow-up angiography and was discharged.

Six months later, because of return of the bruit and increasing headache, he was readmitted. Repeat angiography demonstrated filling of the anomaly from the contralateral left vertebral and the right occipital artery by enlarged posterior muscle collaterals and the ipsilateral thyrocervical

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**Fig. 2.** Case 2. Anteroposterior (left) and lateral subtraction (center and right) views in the early arterial phase, demonstrating the large right vertebral artery filling a massive suboccipital A-V malformation with early filling veins (right). The small arrow (left) indicates the thyrocervical trunk.

**Fig. 3.** Case 2. Submental vertex subtraction view demonstrating the left-to-right shunt to the malformation (large black arrow).
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Fig. 4. Case 2. Artist's recreation of serial angiograms. The major contribution to the malformation is from the vertebral artery depicted as the largest of the four feeding vessels. Repeat angiography, following embolization and trapping of the vertebral artery, clearly defined significant feeders entering the lesion from branches of the external carotid anterosuperiorly, the ipsilateral thyrocervical trunk anteroinferiorly, and muscle branches of both occipital arteries posteroinferiorly. There is a large false aneurysm of the vertebral artery surrounded by multiple venous channels draining via the jugular vein.

Physical examination was normal except for an audible bruit heard best over the left mastoid and along the left sternocleidomastoid muscle. Four-vessel angiography demonstrated an A-V malformation in the left suboccipital area with multiple fistulas in the occipital bone and dura. It was fed by multiple branches of the external carotid, the internal carotid via the tentorial artery of Bernasconi and Casinari (Fig. 5), and extracranial muscle branches of the left vertebral artery. There were no feeders from the right-sided circulation.

The major arterial feeders from the left external carotid artery were embolized with small pieces of muscle marked with metallic clips and ligated. Repeat angiography 6 days postoperatively demonstrated the persistence of early venous shunting despite the evidence of emboli within the A-V malformation. He was returned to the operating room, and the entire lesion was excised including portions of the dura and skull. The position of major feeding vessels could be anticipated during the dissection. As a consequence of this, blood loss was minimal. He has remained free of symptoms for 2 years.

Case 3

A 50-year-old machine operator was admitted for diagnosis of a "swooshing" sound in the left ear. Approximately 1 year earlier he had noted the sudden onset of this noise accompanied by decreased hearing in the left ear and post-auricular headache. Physical examination was normal except for an audible bruit heard best over the left mastoid and along the left sternocleidomastoid muscle. Four-vessel angiography demonstrated an A-V malformation in the left suboccipital area with multiple fistulas in the occipital bone and dura. It was fed by multiple branches of the external carotid, the internal carotid via the tentorial artery of Bernasconi and Casinari (Fig. 5), and extracranial muscle branches of the left vertebral artery. There were no feeders from the right-sided circulation.

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Case 4

A 36-year-old physician was admitted for evaluation of a 6-month history of at first intermittent, then persistent, machinery murmur in the left ear. The sound increased with exercise, and the only other symptoms were progressive suboccipital headache and marked fatigue. His physical and neurological examinations were normal except for the loud continuous bruit over the left mastoid which was obliterated by strong carotid compression.

Angiography demonstrated a lesion similar to that in Cases 1 and 3 with feeders from branches of the external and internal carotid arteries via the tentorial artery. There were several smaller feeding vessels arising from the vertebral artery. Approximately 3 hours following angiography he developed nausea, vomiting, and diaphoresis, followed by bradycardia and hypotension lasting 8 hours. The next morning his bruit was no longer audible, and to this date, 1 year later, has not reappeared.

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Case 5

This 38-year-old woman could recall a roaring sound in her right ear dating back to early childhood. She was asymptomatic and unaware of its significance until age 37 when a bruit was discovered by her gynecologist on a routine examination. She was seen by a general surgeon who ligated the right external carotid without preceding angiography. The bruit disappeared for approximately 3 months only to return in association with a soft distensible mass in the right neck and persistent headache. Since her surgery she had noted a progressive increase in fatigability. Her physical examination was normal except for a large collection of soft distensible veins along the right sternocleidomastoid muscle and a diffuse bruit heard best at the angle of the jaw.

Selective four-vessel angiography demonstrated a large extracranial A-V malformation being fed by the right internal carotid artery via the ophthalmic (Fig. 6 upper left) and the right vertebral by numerous posterior cervical muscle branches (Fig. 6 upper right). The left vertebral study demonstrated a steal phenomenon from left to right. Left common carotid and selective external carotid injections (Fig. 6 lower left and right) demonstrated multiple left to right collaterals via branches of the external carotid including the superior thyroid, ascending pharyngeal, and internal maxillary arteries.

Because of the marked A-V shunting demonstrated on angiography and her subjective complaints of increasing fatigue, cardiac output studies were performed which were elevated at 8.21 l/min. The cardiac outline on x-ray and fluoroscopy was normal. The extent of the lesion and the

Fig. 5. Case 3. Left: Selective external carotid angiogram demonstrating a large feeding artery (open arrow) and early filling of the sigmoid sinus and jugular vein (large closed arrows). Right: Selective internal carotid angiogram showing the dural feeder via the artery of Bernasconi and Casinari (two small arrows) and early draining of the sigmoid sinus (large arrow).
FIG. 6. Case 5. Upper Left: Lateral subtraction view of the right common carotid injection (note ligated external carotid) rapidly filling the internal jugular vein (large black arrow) through collaterals of the ophthalmic artery (two small black arrows) retrograde via the internal maxillary artery (large open arrow). Upper Right: Lateral subtraction view of selective right vertebral injection demonstrating numerous feeding muscular branches (large open arrow) and early filling of the internal jugular vein (large black arrow). Lower Left: Anteroposterior subtraction view of the left common carotid injection demonstrating early filling of the dilated right internal jugular vein (large black arrow) being fed by multiple collaterals including two large internal maxillary arteries (small black arrows). Lower Right: Submental vertex subtraction of the selective left external carotid injection (tip of catheter, small open arrow) with multiple right-to-left collaterals including the superior thyroid, ascending pharyngeal, and internal maxillary arteries. Large black arrow indicates early filling of the internal jugular vein on the right.

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lack of incapacitating symptoms led to a decision for conservative management.

Discussion

The management of congenital extracranial A-V malformations poses a distinct challenge. The goal of total excision demands a thorough knowledge of the arterial feeders, fistula location and venous drainage patterns. Careful selective angiography is mandatory in the definition of these lesions. The failure of clear recognition of minor arterial feeders at initial angiography, as in Case 2, may relate to the phenomenon of "washout" due to dye dilution within large pools of non-opacified blood in the malformation. It is this group of unrecognized vessels, not the major feeders, that often leads to recurrence of the lesion or excessive blood loss at the time of their excision.

Although spontaneous cures have occasionally been reported, as in our Case 4, we would agree with others that these lesions should be dealt with early and that total excision is the procedure of choice. Case 5 demonstrates the futility of simple vessel ligation; it will not obliterate the lesion and may make subsequent excision more difficult, as less accessible feeders to the malformation may increase in size and number. The combination of fistula embolization and vessel ligation may afford a cure in small lesions with single feeders, as in Case 1, but its real usefulness is as an initial procedure, as in Cases 2 and 3.

As a consequence of this limited experience we suggest that these patients be managed sequentially. First, we stress the need for selective angiography of the external and internal carotids, vertebrals and thyrocervical trunks to define the major arterial feeders. Second, the initial surgical procedure should be designed to correct any steal phenomenon, embolize and ligate the major feeding arteries. Third, selective angiography should then be repeated as soon as feasible to examine the full extent of the remaining malformation and its minor arterial supply free of "washout." Fourth, the lesion should then be excised including soft tissue, skull, and dura, completely surrounding and ligating the arterial feeders as they enter the anomaly.

This sequence, though disadvantageous in that repeat angiography and two surgical procedures are required, has the distinct advantages of increasing the chance of total excision, minimizing blood loss at the time of total excision (less than one unit of blood in our cases), and markedly reducing the postoperative soft tissue defects.

References


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