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Congenital Arteriovenous Fistulas Supplied by a Single Branch of the Maxillary Artery

Trygve O. Gabrielsen, John P. Deveikis, Joseph H. Introcaso, and Arnold G. Coran

Summary: We report four cases (two children, two adults) of congenital arteriovenous fistula supplied by a single large (pterygoid) branch of the second part of the maxillary artery deep to the parotid gland and mandible, with emphasis on the angiographic findings, therapeutic implications, and cause, with a review of the literature. Awareness of a predilection of congenital arteriovenous fistulas for this site, excellent-quality selective angiography, and careful attention to flow patterns help make the correct diagnosis. Endovascular balloon occlusion is the preferred treatment.

Index terms: Fistula, arteriovenous; Cerebral angiography; Catheters and catheterization, balloons; Arteries, carotid (external); Parotid gland; Pediatric neuroradiology

Four patients with congenital arteriovenous fistulas supplied by a single maxillary artery branch deep to the parotid gland and mandible will be reported, with emphasis on angiographic findings and congenital cause. Making the correct diagnosis of arteriovenous fistula has important therapeutic implications, which will be discussed. The pertinent literature will be reviewed.

Case Reports

Between 1976 and 1992, two adults and two children with congenital arteriovenous fistulas supplied by a single branch of the second part of the maxillary artery were evaluated and treated by one or more of the authors. To our knowledge, these were the only such nontraumatic fistulas which we have encountered in the facial region during 30 years of experience with craniofacial vascular lesions. The patients' clinical records and radiographic studies were retrospectively reviewed. All patients underwent extensive craniofacial angiography. Table 1 is a summary of their clinical and angiographic findings, therapy, and follow-up.

There was no prior facial trauma or surgery in any of these four patients. With the exception of case 4, the other three patients originally were misdiagnosed as having arteriovenous malformations, as opposed to fistulas supplied by a single branch of the second part of the maxillary artery. In cases 1 and 2, the initial angiographies were done elsewhere and were of suboptimal technical quality. Review of the films and repeat angiographies established the diagnoses of arteriovenous fistulas. All of the fistulas were located deep to the parotid gland and ascending ramus of the mandible. Figures 1 and 2 demonstrate the typical angiographic findings in these patients. The feeding arteries proximal to the fistulas were enormously enlarged, as were the draining veins (Figs 1 and 2A). In every case, the maxillary artery and its branches distal to the fistula were of normal caliber and were opacified in antegrade fashion, although flow in these vessels was much slower than through the fistula and feeding arteries in direct line with the fistula. There was absence of any sump effect via collateral flow before treatment of the fistulas in all four cases.

Case 1 was treated with transfemoral balloon embolization in 1990. Initially, the fistula could be closed by placing a number 16 Ingenor latex balloon (Nycomed-Ingenor, Paris) and multiple platinum microcoils at the fistula site, but the fistula reopened when the balloon and coils migrated into the varix on the venous side of the fistula. Therefore, simultaneously, a much larger number 12 Ingenor latex balloon was placed in the varix, and a number 9 Ingenor balloon was placed in the fistula itself. Both balloons were inflated with 2-hydroxyethylmethacrylate polymerizing agent and detached. Control angiography showed complete occlusion of the fistula, with greatly slowed flow in the very dilated external carotid artery.

In case 2, transfemoral endovascular closure of the fistula was performed in 1992 by simultaneously inflating two number 9 Ingenor latex balloons in the region of the fistula. The distal balloon was just on the venous side and the proximal balloon just on the arterial side of the fistula. Both balloons were inflated with dilute nonionic contrast material and detached. Control arteriography performed immediately afterward documented complete occlusion of the fistula (Fig 2B). The proximal balloon appeared to

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TABLE 1: Four patients with congenital arteriovenous fistulas supplied by a single branch of the maxillary artery

Case	Sex and Age at Initial Angiography	History and Presenting Physical Findings	Angiographic Findings	Treatment, Results, and Follow-up
1	M 45 years	Longstanding mass in left parotid region. Increasingly annoying pulsatile tinnitus.	Arteriovenous fistula supplied by single large branch of second part of left maxillary artery (Figs 1A and 1B).	Endovascular balloon occlusion. Immediate disappearance of tinnitus. No apparent recurrence during 6 months follow-up.
2	F 34 years	Pulsatile swelling of right side of face "all her life." Increasing tinnitus, interfering with sleep.	Right-sided arteriovenous fistula with features practically identical to case 1 (Fig 2A).	Endovascular balloon occlusion (Fig 2B). Immediate relief of tinnitus. No apparent recurrence during 9 months follow-up.
3	M 5 months	Pulsatile mass with thrill and grade V/VI bruit found recently at angle of mandible.	Arteriovenous malformation mistakenly diagnosed on initial angiography. Repeat angiography at age 18 months showed right-sided arteriovenous fistula similar to cases 1 and 2.	Surgical excision at age 22 months. Clinical follow-up until age 3 years, without recurrence.
4	M 5 years	Right-sided neck mass since shortly after birth.	Right-sided arteriovenous fistula with features practically identical to cases 1, 2, and 3.	Surgical excision. No fistula seen on angiography done 5 days after surgery, and no clinical recurrence on follow-up for 8 years.

occlude the maxillary artery at the site of the fistula, but this was felt to be of no clinical significance because there was ample collateral flow into terminal maxillary artery branches.

In case 3, the patient was 5 months old when we first saw him in 1976. He had a wide pulse pressure (97/37 mmHg). His neck mass was thought to be increasing in size on follow-up, and his heart size became questionably enlarged before repeat angiography at 18 months of age. At age 22 months, a third transfemoral angiography was done before surgery, with transient complete endovascular balloon occlusion which accurately located the fistula. However, no detachable balloon catheter was available. Leaving the catheter in place, as planned, we immediately took the patient to the operating room. All proximal branches of the right external carotid artery were successively ligated. The catheter was used as a guide to find the fistula medial to the mandible. The fistula was ligated and resected.

In case 4, the patient was a 5-year-old boy when evaluated and treated in 1979. He had a grade III/IV systolic injection murmur over the entire precordium but no cardiomegaly. Based on our prior experience with case 3, we successfully undertook similar surgical treatment with ligation and resection of the fistula.

Discussion

Although the clinical findings in these four cases consisted mostly of cosmetic changes or symptoms and signs related to fast turbulent flow, the shunt in the infant was sufficiently massive to affect his cardiac status.

There can be little doubt that all of the four arteriovenous fistulas reported here are congenital in origin. There was no history of facial surgery or trauma as a cause. Admittedly, the two adult patients might not have remembered remote trauma, but trauma of sufficient force to have produced an arteriovenous fistula almost certainly would have been remembered, especially by the parents of the 5-month-old and 5-year-old children. Severe blunt trauma to the face, with or without fractures, certainly may result in arteriovenous fistulas. However, they usually involve a major artery such as an internal carotid or vertebral artery. Pseudoaneurysm formation or other arterial injury involving a branch of the external carotid artery also may be produced by penetrating trauma, surgery, or blunt trauma of sufficient force to produce fractures. However, as already stated, any history of such trauma was lacking in all of the present cases.

It is striking that all of these four arteriovenous fistulas were located in exactly the same place, immediately deep to the parotid gland and ascending ramus of the mandible. In every one of the four cases, a single branch of the maxillary artery, probably a pterygoid branch, supplied the malformation. To our knowledge, we have never encountered any congenital arteriovenous fistulas in any other extracranial location in the head and neck region, despite our having seen numerous hemangiomas and arteriovenous malformations in practically every conceivable location.

Halbach et al (1) reported six patients, and Berenstein et al (2) reported one patient, all adults with similar fistulas of presumed congenital ori-

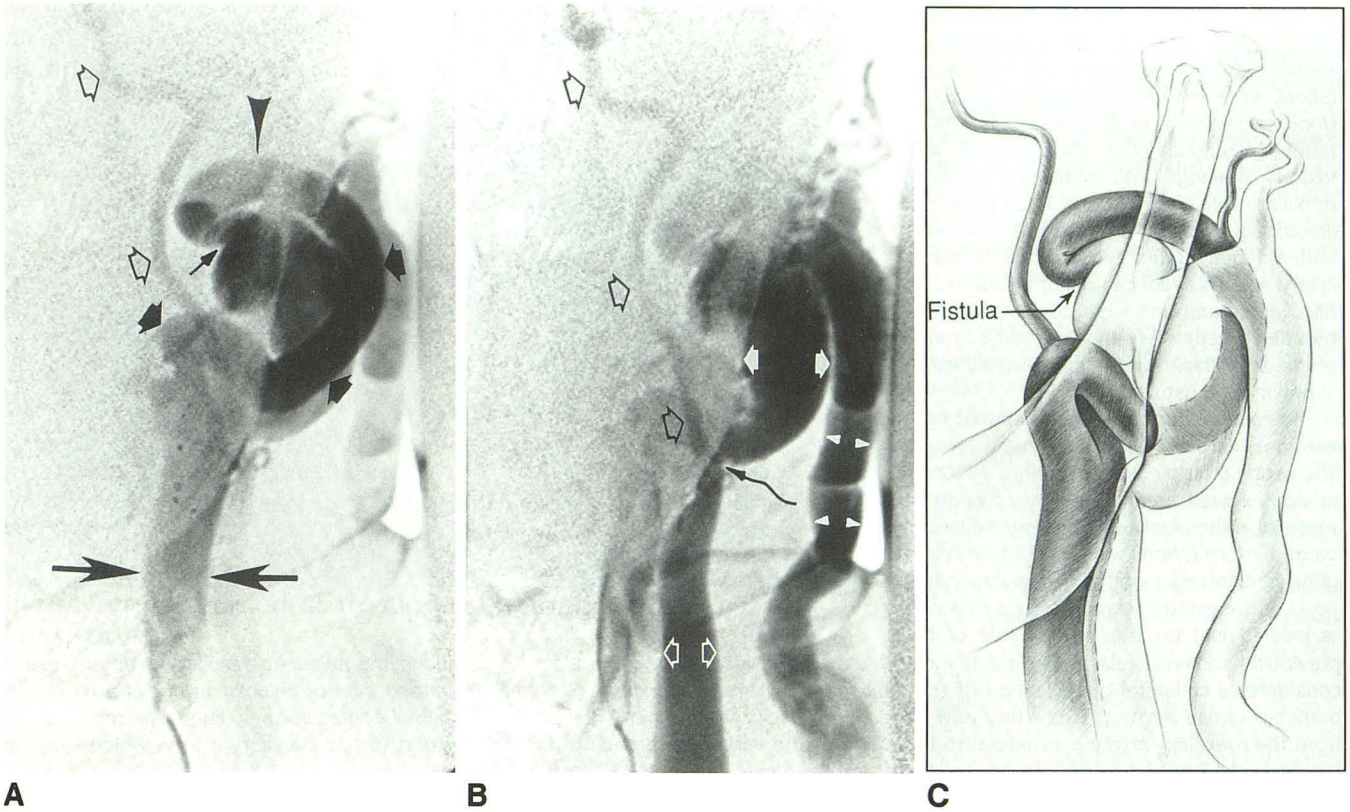

Fig. 1. Case 1. Preembolization frontal images of left common carotid digital subtraction angiography (*A* and *B*).

A, Common carotid (*large arrows*), external carotid (*wide solid black arrows*), and first part of maxillary (*long arrowhead*) arteries are enormously enlarged compared with normal size of the internal carotid artery (*open black arrows*). The maxillary artery distal to fistula (*thin, short arrow*) was opacified in antegrade fashion and was of normal size as shown in lateral views (not illustrated). Note very rapid flow in external carotid and proximal maxillary arteries compared with flow in the internal carotid artery, which is a reversal of the normal pattern, and very early opacification of grossly enlarged maxillary and other draining veins.

B, There appears to be a prominent web-like stenosis (*wavy arrow*) at the junction between the retromandibular vein (*wide solid white arrows*) and internal jugular vein (*open white arrows*), which may be partly responsible for the prominent degree of drainage through the external jugular vein (*small white arrowheads*).

C, A composite sketch of A and B, showing location of fistula and major arteries and veins relative to lateral border of mandible. Arteries are darkly shaded. Veins are lightly shaded.

gin, and all treated successfully by endovascular balloon occlusion. Similar fistulas in 2-year-old and 9-year-old children were treated in like manner by Kawakami et al (3) and Cluzel et al (4), respectively. Although the fistula was called spontaneous by Kawakami et al, it must be of congenital origin, since there was no history of trauma, and the lesion had been noticed 6 months after birth. An additional, similar lesion excised surgically in a 4-year-old girl was reported by Girevendulis et al (5). Including the present four cases, 14 such fistulas, nine in adults and five in children, have been recognized angiographically. All were treated successfully, 11 by endovascular balloon occlusion and three by surgical excision. Additional cases encountered by others may have been mistakenly diagnosed as arteriovenous malformations with more than one arterial feeder, as initially in three of our four cases.

Scialfa et al (6) described a similar fistula in a 9-year-old boy who was treated successfully by balloon occlusion, but they thought that the single arterial supply probably was from the ascending pharyngeal artery. The ascending pharyngeal artery also was cited as the supply to an arteriovenous fistula in an adult included in the paper by Berenstein et al (2). The material in that paper also included a 7-year-old boy with a fistula of presumed congenital origin involving the middle meningeal artery.

We are not able to offer any convincing embryologic basis for the apparent strong predilection for development of congenital arteriovenous fistulas in this particular region. Neither our patients nor those reported previously seem to have had associated soft tissue or bony anomalies in the craniofacial region, suggesting an isolated problem of vascular development.

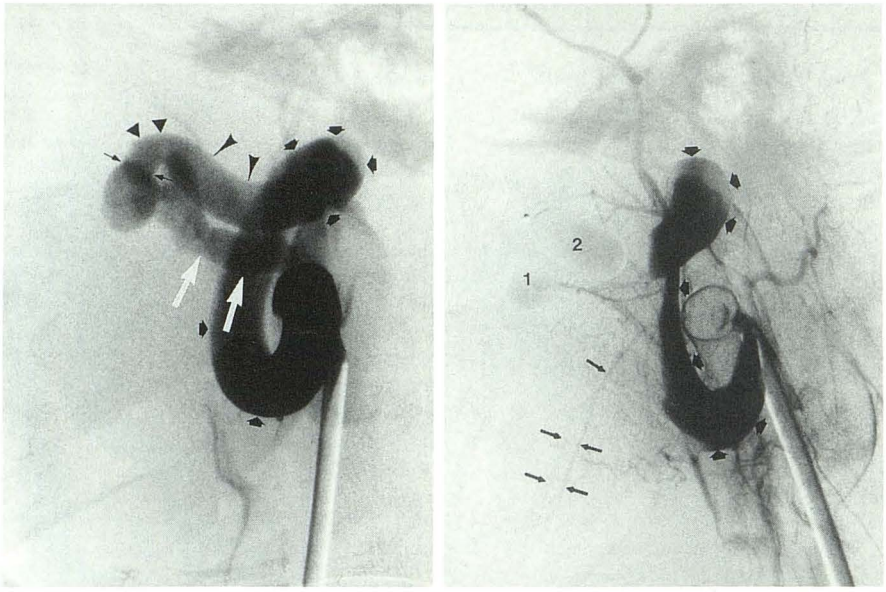

Fig. 2. Case 2. *A*, Preembolization lateral view of right external carotid angiography shows extremely enlarged external carotid (*short, wide arrows*) and proximal maxillary (*long arrowheads*) arteries supplying an arteriovenous fistula (*thin, short arrows*), with venous drainage into markedly enlarged maxillary vein (*wavy arrow*). Later films (not illustrated) showed huge retromandibular and external jugular veins. The maxillary artery and its branches distal to the fistula filled antegrade and were normal in caliber (not illustrated). Note the very large branch of the maxillary artery (*short arrowheads*) supplying the fistula.

B, Postembolization lateral view of right external carotid angiography no longer shows any arteriovenous shunting, and there is very slowed antegrade flow of contrast material in the persistently enlarged external carotid artery (*short, wide arrows*), with layering of contrast material in the dependent parts of the external carotid artery. Balloon1 is located just on the venous side of the previously shown fistula. Balloon 2 is located just on the arterial side and also has occluded the adjacent maxillary artery. There is considerable collateral opacification of terminal maxillary artery branches. Note well-visualized inferior alveolar artery and two of its branches (*small arrows*). This artery now appears to opacify via collateral flow, but its proximal course strongly suggests that its origin from the maxillary artery is proximal to the origin of the very large branch of the second part of the maxillary artery, which supplied the occluded fistula located between balloon 1 and balloon 2.

In cases of arteriovenous malformation with more than one arterial feeding vessel, enlarged collateral vessels from adjacent arteries with a sump effect supplying the arteriovenous malformation generally can be observed. No such collateral vessels or sump effect could be seen in any of the present four cases. As far as can be determined, several illustrations of previously published cases also have shown this feature (2–5), although van Halbach et al (1) reported seeing antegrade flow in the distal maxillary artery and its branches only after balloon occlusion of the fistulas. Interestingly, pterygoid arteries are said typically not to have collaterals with other arterial systems (7). This may be one possible explanation why no collateral channels from adjacent vessels contributed to these fistulas.

In every one of the four cases, it also was very striking that the maxillary artery and its branches distal to the location of the fistula were of normal caliber, whereas the common and external carotid and proximal maxillary arteries were enormously enlarged. In every case of congenital fistula reported here, the arteries in direct line to the fistula gradually must have become so enlarged, presumably both before and after birth, that they were capable of supplying antegrade flow also to all remaining external carotid artery branches. As far as can be determined, all illustrations of the above-mentioned previously published cases also have shown marked enlargement of the external carotid and proximal maxillary arteries and normal size of the distal maxillary artery and its branches (1–5). There may be improved, sometimes markedly improved, antegrade flow in the distal maxillary artery and its branches after balloon closure of the fistula (1–3).

In making a correct diagnosis, it is important to perform as selective angiography as possible, and of excellent technical quality. Making the correct diagnosis is essential for appropriate therapy. An acute awareness that these quite rare congenital arteriovenous fistulas apparently have a strong predilection for the pterygoid region should be a helpful diagnostic clue. It perhaps may be useful, in a practical sense, to regard these particular fistulas as a distinct entity, because all of these 14 fistulas of apparent congenital origin occurred in the same area, and congenital fistulas in other neighboring locations are extremely rare.

Although surgical treatment may be challenging, it certainly may be feasible, as in our two children 14 and 16 years ago. In considering

endovascular, curative occlusive therapy, it is absolutely essential to recognize the presence of an arteriovenous fistula, as opposed to an arteriovenous malformation. Arteriovenous malformations with more than one arterial feeding vessel usually can be treated with variable success with embolic agents such as cyanoacrylates, coils, and particulate embolic materials, either as definitive therapy or before surgical resection. However, such embolic materials generally are not suitable for closure of a fistula of large size. Nevertheless, corrective endovascular occlusive therapy of such a fistula also can be achieved via a transfemoral approach using detachable balloons. This can be achieved in a much less invasive manner than by conventional surgery.

Given the prerequisite endovascular occlusive therapeutic skill, we consider balloon occlusion the preferred method of treating such arteriovenous fistulas.

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Imaging Quiz: Request for Original Submissions

This issue marks the appearance of a new feature in the *AJNR*, the Imaging Quiz. The quiz can be found on page 658 and the diagnosis on page 774. We welcome other original submissions for this feature. This feature must fit on no more than two journal pages: a quiz page and a diagnosis page. Therefore, the approximate length of submissions should be two to three double-spaced pages of text, including references, and up to 10 figures. The submissions will undergo peer review. We look forward to receiving your contributions for this feature.