Spontaneous Thrombosis of a Middle Meningeal Arteriovenous Fistula With Subsequent Pseudoaneurysm Formation: Case Report and Review of Literature

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BACKGROUND AND IMPORTANCE: Middle meningeal artery (MMA) pseudoaneurysms and middle meningeal arteriovenous fistulas (MMAVFs) are rarely reported after head injury. We report an unusual case of delayed MMA pseudoaneurysm formation after spontaneous thrombosis of an MMAVF, and review existing literature on MMAVF treatment and results.

CLINICAL PRESENTATION: A 59-yr-old male presented with a 5-d history of worsening left-sided headaches, followed by nausea, lethargy, and difficulty with speech. Non-contrast computed tomography demonstrated a left temporal intraparenchymal hemorrhage (IPH) and an acute left-sided subdural hematoma (SDH). Cerebral angiography found abnormal shunting between the right MMA and the right sphenoparietal sinus, consistent with an MMAVF. During the course of admission, the patient’s neurological condition deteriorated requiring craniotomy for evacuation of SDH and IPH. Given the presumed incidental nature of the contralateral MMAVF, conservative management was recommended. Follow-up imaging 2 mo after surgery revealed spontaneous thrombosis of the right MMAV. Repeat imaging 5 mo later revealed an MMA pseudoaneurysm at the prior fistulous site, which was subsequently embolized with Onyx, occluding the pseudoaneurysm and the MMA both proximal and distal to the pseudoaneurysm.

CONCLUSION: Spontaneous thrombosis of an MMAVF is rare and only seen in 13.1% of cases. However, subsequent delayed formation of an MMA pseudoaneurysm has not been described. Our case therefore demonstrates that MMAVF thrombosis may not indicate complete healing of the underlying injury to the MMA, and suggests the need for continued follow-up of such lesions despite initial apparent resolution.

KEY WORDS: Middle meningeal artery, Arteriovenous fistula, Pseudoaneurysm, Spontaneous thrombosis, Case report

The middle meningeal artery (MMA) is the first and largest branch of the maxillary artery, it enters the skull through the foramen spinosum, and supplies bone and dura mater. Damage to the MMA can lead to epidural hematoma, traumatic aneurysm, or arteriovenous fistula. Most middle meningeal arteriovenous fistulas (MMAVFs) are associated with trauma and occur between the MMA and neighboring dural veins or venous sinuses. MMAVF may also present with mild or nonspecific symptoms or be incidentally discovered during imaging performed for other purposes. Endovascular embolization is commonly used to treat MMAVFs, with surgery remaining an alternative treatment.
In contrast, spontaneous closure of these fistulas is rarely reported.\textsuperscript{2,4-8} We describe an unusual case of an MMAVF that spontaneously thrombosed but was then associated with delayed development of an MMA pseudoaneurysm, and review existing literature on MMAVF treatment and results.

**CLINICAL PRESENTATION**

**History and Examination**

A 59-yr-old diabetic hypertensive male presented with a 5-d history of worsening left-sided headaches, followed by nausea, lethargy, and difficulty with speech. He could not recall any trauma. Initial examination revealed mild dysphasia and dysnomia but he could follow commands and move all extremities with good strength. Non-contrast head computed tomography (CT) demonstrated a left temporal intraparenchymal hemorrhage (IPH) as well as an acute left-sided subdural hematoma (SDH). Magnetic resonance imaging (MRI) demonstrated no underlying tumor and CT angiography (CTA) showed no vascular etiology of the left-sided hemorrhage but revealed abnormal vascular structures in the contralateral right middle cranial fossa (Figure 1). Cerebral angiography similarly demonstrated no vascular abnormalities to explain the left-sided hemorrhage, but found abnormal shunting between the right MMA and the right sphenoparietal sinus via a dilated interdural vein along the lateral wall and floor of the right middle cranial fossa, consistent with an MMAVF (Figure 2). There was no associated cortical venous reflux.

**Clinical Course**

Given minimal deficits on initial exam, craniotomy was deferred, and the patient was admitted to the neurointensive care unit for observation. Five days later, the patient’s neurological condition deteriorated and CT found increased left subdural collection and worsening of midline shift, requiring craniotomy for evacuation of SDH and IPH. His neurological examination improved and he was discharged home on postoperative day 4 without deficit, with follow-up postoperative noncontrast head CT demonstrating no residual hematoma. Given the presumed incidental nature of the contralateral MMAVF, conservative management with serial imaging was...
recommended. Follow-up CT/CTA 2 mo after surgery revealed no recurrent IPH/SDH and spontaneous thrombosis of the right MMAV (Figure 1). Repeat imaging 5 mo later confirmed MMAVF thrombosis but now revealed an MMA pseudoaneurysm at the prior fistulous site, which was subsequently embolized with Onyx, occluding the pseudoaneurysm and the MMA proximal and distal to the pseudoaneurysm (Figure 2). Six months after embolization, the patient remained neurologically intact, was back at work, and had no residual pseudoaneurysm, MMAVF, IPH, or SDH on MRI/magnetic resonance angiography. The patient provided consent for this publication.
| First author and year | N | Sex | Age (yr) | Trauma | Fracture | Side of fistula | Pseudo-aneurysm | Presentation | Treatment | Radiologic outcome | Clinical outcome |
|-----------------------|---|-----|---------|--------|----------|----------------|----------------|-------------|-----------|-------------------|-----------------|
| Fincher 1951⁹          | 1 | F   | 24      | Y      | Y        | I              | N              | H/A, progressive pulsatile tinnitus, bruit | Crani (MMA ligation, MMA clipping at 2nd operation) | UR        | CR               |
| Wilson 1964⁸           | 2 | M; M| 23; 78  | Y; Y   | Y; N     | I; N/A         | N; N           | H/A, dysphasia, facial paresis; H/A, dysphasia, obtundation | Crani; SR  | UR; NR            | Speech improvement; CR |
| Jackson 1964¹⁰         | 1 | F   | 56      | Y      | Y        | I              | N              | Obtundation, nuchal stiffness | SR         | NR                | CR               |
| Jakarinen 1965¹¹       | 1 | M   | 25      | Y      | N        | N/A            | N              | H/A, pulsatile tinnitus, bruit | ECA ligation | NR                | CR               |
| Nakamura 1966¹²        | 1 | M   | 64      | Y      | N        | N/A            | N              | H/A, left lateral gaze paralysis, double vision | Crani (coagulation, excision) | NR       | CR               |
| Handa 1970¹³           | 1 | M   | 52      | Y      | Y        | I              | N              | H/A, bilateral decrease of vision | Crani (MMA and MMV ligation) | UR       | UR               |
| Sicat 1975¹⁴           | 1 | M   | 49      | Y      | Y        | I              | Y              | Persistent severe H/A | Crani (excision) | NR                | CR               |
| Ishii 1976¹⁵           | 1 | M   | 65      | Y      | Y        | I              | N              | Speech impairment | SR         | NR                | CR               |
| Kitahara 1977¹⁶        | 3 | F; M; M| 27; 42; 38 | Y; Y | Y; Y | I; I; I | N; N; Y | Blurred vision; Proptosis; Nuchal stiffness | SR; Crani (coagulation); Crani (excision) | NR; UR | CR; CR; Dysnomia |
| Occhiogrosso 1980¹⁷     | 1 | F   | <1      | Y      | Y        | I              | Y              | Restlessness, proptosis, progressive obtundation | Crani (MMA coagulation) | UR       | Mild weakness   |
| Freckmann 1981¹⁸       | 8 | F (3), M (5) | 50*     | Y (8)  | Y (8)    | I (8)          | N (8)          | Cranial hematoma | Crani (5), UR (4) | UR              | Death (3), PVS (2), severe deficits (2), CR (1) |
| Smith 1981¹⁹           | 1 | M   | 37      | Y      | Y        | C              | N              | Confused, combative, seizure, progressive coma | N/A        | N/A              | Death            |
| First author and year | N  | Sex | Age  (yr) | Trauma | Fracture | Side of fistula | Pseudo-aneurysm | Presentation | Treatment | Radiologic outcome | Clinical outcome |
|-----------------------|----|-----|----------|--------|----------|----------------|-----------------|--------------|-----------|-------------------|-----------------|
| Bradac 1981<sup>18</sup> | 1  | M   | 33       | Y      | Y        | I              | N              | Chemosis, proptosis, bruit, tinnitus | Embo (Ivalon) | UR                 | CR              |
| Satoh 1983<sup>6</sup>  | 1  | F   | 75       | Y      | Y        | C              | N              | Nausea/vomiting, pain, SAH | SR         | NR                 | CR              |
| Inagawa 1984<sup>19</sup> | 1  | F   | 70       | Y<sup>7</sup> | Y | I  | N  | Routine angiogram after clipping aneurysm | Crani (excision) | UR                 | UR              |
| Bhoopat 1987<sup>20</sup> | 3  | F; M; M | 23; 29; 24 | Y; Y | N; Y | N/A; N/A; I | N; N; N | Proptosis, chemosis; Proptosis, chemosis, tinnitus; Proptosis, chemosis, bruit | Embo (catgut); Embo (catgut, polyethylene tube, Gelfoam); Embo (Gelfoam) | UR                 | Improvement; CR; Recurrence |
| Tsutsumi 1990<sup>5</sup> | 1  | F   | 35       | UR     | N/A     | N              | N              | Routine angiogram after craniotomy (for SAH) | SR         | NR                 | CR              |
| Komiyama 1994<sup>21</sup> | 1  | M   | 58       | Y      | Y        | I              | N              | Decreased level of consciousness | Embo (silk suture) | UR                 | CR              |
| Touho 1995<sup>22</sup>  | 1  | M   | 27       | Y      | N        | N/A            | N              | Progressive bruit | Embo (coil) | NR                 | CR              |
| Terada 1997<sup>23</sup> | 1  | F   | 73       | Y<sup>9</sup> | N | N/A  | N  | Mild H/A | Embo (PVA) | NR                 | UR              |
| Wang 2000<sup>24</sup>   | 1  | M   | 21       | Y      | UR      | N/A            | N              | Proptosis, diplopia | Embo (NBCA) | UR                 | Choroidal infarction |
| Tsumoto 2001<sup>25</sup> | 1  | M   | 57       | Y      | Y        | I              | N              | H/A, frontal bone defect | Embo (PVA, NBCA) | NR                 | CR              |
| Matsumoto 2001<sup>26</sup> | 1  | F   | 65       | Y      | Y        | I              | N              | H/A, paresis, vertex epidural hematoma | Crani       | UR                 | CR              |
| Kawaguchi 2002<sup>27</sup> | 1  | M   | 44       | Y      | Y        | I              | N              | Optic nerve injury | Embo (liquid agents) | NR                 | Focal deficits |
| Tsutsumi 2002<sup>28</sup> | 1  | M   | 23       | Y      | Y        | I              | Y              | Bruit, chemosis | Embo (coil) | NR                 | CR              |
| Chandrashekar 2007<sup>4</sup> | 1  | M   | 42       | N<sup>8</sup> | N | N/A  | N  | LOC, seizures, alcoholic withdrawal | SR         | NR                 | UR              |
| Liu 2006<sup>29</sup>    | 1  | M   | 22       | Y      | N        | N/A            | N              | Bruit, blurred vision, exophthalmos, diplopia, blepharoptosis | Embo (detachable balloon) | UR                 | CR              |
| First author and year | N  | Sex | Age (yr) | Trauma | Fracture | Side of fistula | Pseudoaneurysm | Presentation | Treatment | Radiologic outcome | Clinical outcome |
|-----------------------|----|-----|----------|---------|----------|----------------|----------------|--------------|-----------|--------------------|------------------|
| Unterhofer 2009⁰⁰ | 1 | M  | 53       | Y       | Y        | I             | N             | Pulsatile exophthalmos, chemosis | Embo (coil) | UR                | Severe disability |
| Sakata 2009¹¹ | 1 | F  | 48       | Y       | Y        | I             | N             | H/A, LOC, spontaneous SAH | Embo (coil) | NR                | NR               |
| Sacho 2014¹² | 1 | F  | 41       | Y**    | N        | N/A           | N             | Pulsatile tinnitus, change in H/A pattern | Embo (coil) | NR                | CR               |
| Ko 2014³  | 1 | M  | 24       | Y       | Y        | C             | Y††           | Progressive pulsatile tinnitus | Embo (coil) | NR                | CR               |
| Champeaux 2016¹³ | 1 | F  | 45       | Y²²    | N        | N/A           | N             | Pulsatile tinnitus, pain | Embo (coil) | NR                | Facial pain recurrence |
| Almefty 2016¹⁴ | 9 | F (5), M (4) | 60^   | Y (4); N (5) | Y (4); N (5) | UR (4); N/A (5) | Y (1); N (8) | SAH (3), IPH (4), SAH/IPH (1), SDH (1) | Embo (NBCA [6], Onyx [2], coil [1]) | UR                | Focal deficits (4) |
| Yu 2017¹⁵ | 1 | M  | 8        | Y       | N        | N/A           | N             | Proptosis, chemosis, bruit | Embo (coil, Onyx) | NR                | CR               |
| Park 2017¹⁶ | 1 | M  | 69       | Y       | N        | N/A           | Y             | Stupor, ICH, SAH, SDH | Embo (NBCA) | Cerebral vasospasm | Residual paresis, dysarthria |
| Martinez 2018¹⁷ | 1 | F  | UR       | Y       | Y        | I             | N             | Pulsatile tinnitus | Embo (coil) | UR                | UR               |
| Kamble 2018¹⁸ | 1 | M  | 29       | Y       | UR       | N/A           | N             | Progressive proptosis, chemosis, pain | Embo (glue) | UR                | CR               |
| Fei 2019¹⁹ | 1 | M  | 47       | Y       | N        | N/A           | N             | Weakness, speech difficulties, midbrain hemorrhage | Embo (coil) | UR                | CR               |
| Tokairin 2019²⁰ | 1 | M  | 24       | Y       | Y        | C             | N             | Tinnitus | Embo (coil, NBCA) | NR                | CR               |
| Dahl 2019²¹ | 1 | F  | <1       | Y       | N        | N/A           | N             | Bruit, proptosis | Embo (coil) | NR                | CR               |
| Present case  | 1 | M  | 59       | N       | N        | N/A           | Y             | H/A, lethargy, speech difficulties | SR (residual MMA) | NR                | CR               |

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¹ Only mean age of the patients was provided.
² Iatrogenic: Mayfield skull fixation.
³ Iatrogenic: ipsilateral pterional craniotomy.
⁴ Iatrogenic: during tumor embolization.
⁵ History of trauma not provided however CT imaging most consistent with traumatic frontotemporal contusions rather than primary ICH.
⁶ Iatrogenic: bilateral arthroscopic temporomandibular joint surgery.
⁷ Careful review of angiograms suggest dilated venous portion of fistula in stead of pseudoaneurysm, but we count this case as pseudoaneurysm based on the manuscript.
⁸ Iatrogenic: percutaneous trigeminal rhizotomy.
⁹ History of trauma not provided however CT imaging most consistent with traumatic frontotemporal contusions rather than primary ICH.

Legend:
I = ipsilateral; H/A = headache; Crani = craniotomy; MMA = middle meningeal artery; UR = unreported; CR = complete recovery; N/A = not applicable; SR = spontaneous resolution; NR = no residual/recurrence; ECA = external carotid artery; PVS = persistent vegetative state; C = contralateral; Embo = embolization; SAH = subarachnoid hemorrhage; PVA = polyvinyl alcohol particles; NBCA = N-butyl cyanoacrylate; LOC = loss of consciousness; IPH = intraparenchymal hemorrhage; SDH = subdural hemorrhage; ICH = intracranial hemorrhage.
TABLE 2. Summary of Demographics, Treatment and Clinical Outcomes

| Characteristic          | Value   |
|-------------------------|---------|
| No. of patients         | 61      |
| Sex                     | Female  37.7% |
| Age                     | Mean (SD), years 44.97 (18.8) |
| Traumatic               | Yes 88.5% |
| No 11.5%                |
| Fracture                | Yes 60.7% |
| No 34.4%                |
| Time period*            | 1 d 9.8% |
| 2-7 d 16.4%             |
| 8-30 d 23.0%            |
| 31-90 d 4.9%            |
| 91-365 d 8.2%           |
| > 365 d 4.9%            |
| Nontraumatic            | 11.5%   |
| Unreported              | 21.3%   |
| Treatment               | Embolization 54.1% |
| Craniotomy              | 24.6%   |
| Spontaneous resolution  | 13.1%   |
| Deceased before treatment | 4.9%  |
| External carotid artery ligation | 1.6% |
| Unreported              | 1.6%    |
| Clinical outcome        | Complete recovery 55.7% |
| Focal deficits          | 16.4%   |
| Severe deficits         | 8.2%    |
| Death                   | 6.6%    |
| Symptom recurrence      | 3.3%    |
| Unreported              | 9.8%    |

*Time period between trauma and diagnosis of MMAF.

Literature Review

Methods

A literature review of PubMed using key words “middle meningeal arteriovenous fistula” and limited to human and English studies yielded 223 publications. Only articles reporting patients with documented MMAVF and known demographics, treatment, and radiographic outcomes were included. Four additional eligible studies were found through screening reference lists.

Results

This yielded 40 publications with 60 cases of MMAVF published between 1951 and 2019 (Figure, Supplemental Digital Content). Data on all 60 patients, with the addition of our patient, is summarized in Table 1. A summary of clinical characteristics, treatment and outcomes of reported cases is presented in Table 2. Most patients had suffered trauma (88.5%) and had a skull fracture (60.7%). The MMAVF was ipsilateral to the side of the fracture in 87.9% and contralateral in 12.1% (where reported) of the cases. Embolization was the most common treatment (54.1%), whereas spontaneous resolution was uncommon and only seen in 13.1% patients. Only 13.1% of MMAVFs were associated with an MMA pseudoaneurysm, of which all but 1 were discovered (and treated) upfront. However, no cases of MMA pseudoaneurysm were reported after spontaneous MMAVF thrombosis. Clinical outcomes were mostly favorable with complete recovery in 55.7% of patients; however, MMAVF led to death in 6.6% patients.

DISCUSSION

Background

The MMA is the main source of blood supply to the meninges, arises from the maxillary artery in the infratemporal fossa, continues its course through the foramen spinosum into the floor of the middle cranial fossa, and supplies a large portion of the bone and bone marrow of the cranium. The anatomy and course of this artery leaves it vulnerable to traumatic injury which can occur with or without an associated skull fracture and may result in an epidural hematoma, pseudoaneurysm, and/or rarely, an MMAVF. First described by Fincher in 1951, Freckmann et al. in 1981 found only 8 cases of MMAVF after reviewing 446 angiograms in patients with head trauma.

Clinical symptoms of MMAVF relate to its venous drainage pattern and may include bruit, proptosis, chemosis, pulsatile tinnitus, headache, seizure, and loss of consciousness. As the MMV has numerous adjacent interconnected channels (Figure 3), MMAVF drainage may include diploic veins, the pterygoid plexus, the cavernous, sphenoparietal, superior and inferior petrosal, superior sagittal, and greater petrosal dural venous sinuses, the superior ophthalmic vein, or infrequently a cortical vein. Treatment decisions are usually related to acuity of symptoms and presence of cortical venous drainage.

Review of literature found most MMAVF to be traumatic in etiology, ipsilateral to the side of the head trauma, and more often treated with embolization (Table 2). However,
rare cases of MMA VF contralateral to the side of injury have been reported, as have iatrogenic and spontaneous cases. For example, Champeaux et al reported a case of ipsilateral MMAVF 3 wk after percutaneous retrogasserian glycerol rhizotomy for trigeminal neuralgia management, and Sacho et al in 2014 described a rare case of MMAVF after arthroscopic surgery of the temporomandibular joint.

Present Case

Our patient is unusual in that he did not have a history of trauma, his MMAVF was contralateral to the site of parenchymal hemorrhage, and repeat imaging found spontaneous thrombosis of his MMAVF, obviating need for initial treatment. He was then found to have an MMA pseudoaneurysm on delayed imaging, which was subsequently treated with embolization. While not seen on prior imaging, we suspect the MMA pseudoaneurysm arose at site of the initial arterial injury to the MMA which led to the MMAVF. Our patient is unique in that his MMA pseudoaneurysm was only found in delayed fashion after his MMAVF spontaneously thrombosed. Pseudoaneurysms of the MMA are rare and likely share the same traumatic etiologic with MMAVF, with prior reports of MMA pseudoaneurysms noting their occurrence where they cross fracture sites in the temporal bone. It is therefore possible that our patient suffered head trauma but was unable to recall the event. A possible but less likely alternative would be a right-sided dural arteriovenous fistula (DAVF) with secondary left-sided hemorrhage due to venous hypertension from the DAVF. However, this would assume thrombosis of the (non-visualized) DAVF pial venous drainage at time of initial presentation, and would not explain the delayed appearance of the MMA pseudoaneurysm.

CONCLUSION

MMAFVs are rare lesions found primarily in patients with history of trauma. While most are treated endovascularly, MMAFVs may spontaneously thrombose, as occurred in our patient. However, subsequent delayed finding of an asymptomatic MMA pseudoaneurysm has not been previously described, and our case suggests that spontaneous thrombosis of the MMAVF may not necessarily indicate complete healing of the underlying arterial injury to the MMA, leaving such patients at risk for recurrent MMAVF or other sequelae. Delayed clinical and imaging follow-up is therefore recommended in patients with spontaneously thrombosed MMAVF.

Disclosures

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.
