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# Arterial surgery for sporadic hemiplegic migraine: preliminary results

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## Abstract

My aim was to report for the first time (to my knowledge) the successful treatment of 3 patients with sporadic hemiplegic migraine by surgical cauterization of terminal branches of the external carotid artery under conscious sedation. Since the operations (between 1 and 2 years) none of the patients have had further attacks of migraine or hemiplegia. This preliminary report suggests that for patients with sporadic hemiplegic migraine with a confirmed arterial component, surgical cauterization of selected terminal branches of the external carotid artery may be effective treatment.

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*Keywords:* Sporadic hemiplegic migraine; External carotid artery; Vascular surgery; Migraine surgery

## Introduction

Sporadic hemiplegic migraine is defined as migraine with an aura including motor weakness but with no first- or second-degree relative having an aura that included motor weakness.<sup>1</sup>

Attacks have the same clinical characteristics as those of familial hemiplegic migraine, namely motor weakness and at least one of the following: fully reversible visual symptoms including positive features (such as flickering lights, spots, or lines) or negative features (such as loss of vision), or both; fully reversible sensory symptoms including positive features (such as pins and needles) or negative features (such as numbness), or both; or fully reversible dysphasic speech disturbance.

They also have at least two of the following: at least one symptom of aura that develops gradually over 5 min or more, or different symptoms of aura that occur in succession over

5 min or more, or both; each symptom of aura lasts more than 5 min and less than 24 h; and headache that fulfills criteria B–D for migraine without aura,<sup>1</sup> begins during the aura, or follows the onset of aura within 60 min.

Because sporadic hemiplegic migraine is rare there are no clinical trials of treatment available so the treatment is based on empirical data, the personal experience of the treating neurologist, and it involves trial and error. Acetaminophen and non-steroidal anti-inflammatory drugs are often the first choice of treatment for an acute attack. An effective treatment for the severe and often prolonged symptoms of aura is more warranted, but currently no such treatment is available.<sup>2</sup> Prophylaxis can be considered when the number of attacks exceeds 2/month, or when severe attacks pose a greater burden that requires reduction of severity and frequency. Propranolol,<sup>3</sup> flunarazine,<sup>4</sup> naloxone,<sup>5</sup> and verapamil<sup>6</sup> have all been reported to have a good therapeutic effect, but the studies were all too small to achieve a confirmed significant effect.<sup>7</sup> Sodium valproate, lamotrigine, and acetazolamide have also been suggested, and while less evidence is available for prophylactic treatment with topiramate, candesartan,

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and pizotifen, these drugs can also be considered.<sup>2</sup> None of these treatments, however, have been shown to be particularly effective.

## Methods

To find out whether the pain originates in the extracranial vessels, it is necessary to examine the patient while the pain is present. The diagnosis can be confirmed if the pain diminishes when blood flow to the painful area is interrupted by digital compression of the relevant vessels. When the pressure is removed the pain returns, usually within a few seconds. The vessels most frequently associated with the pain are: the main trunk of the superficial temporal artery; the frontal and parietal branches of the superficial temporal artery; and the occipital, maxillary, angular, and posterior auricular arteries. In most patients, multiple vessels are involved in the pain and all must be cauterized. All 3 patients were examined during an attack.

In patients with unilateral pain, the scalp vessels are still cauterized bilaterally, because there are extensive side-to-side anastomoses of some of the terminal branches of the external carotid artery,<sup>8</sup> and cautery of the vessels only on the affected side is sometimes ineffective.<sup>9</sup> It is only necessary to cauterize the maxillary artery on the affected side in side-locked unilateral headache. The techniques for cauterizing these vessels have been described elsewhere.<sup>9,10</sup>

## Case reports

### Case 1

A 38-year-old woman had had left sided migraine since childhood. Initially her main trigger was the hot sun, but when she reached puberty, hormone fluctuations and stress began to be more important. The first attack that was accompanied by paralysis occurred in 2003 when she was 30 years old. She experienced a stabbing pain on the left side of her head and fainted. When she regained consciousness the left side of her face was paralyzed. She could not recall how long the facial paralysis lasted, but it did recover spontaneously. Neurological examination and magnetic resonance imaging (MRI) were within normal limits. Shortly afterwards she had another, severe, attack, which rendered her unconscious until the following day. When she did regain consciousness she was disorientated for a few hours. Neurological examination showed no neurological deficit, and she was discharged with a diagnosis of migraine with aura.

As the years progressed the aura became worse, and 2 years after the first attack she started becoming completely paralyzed on her left side during attacks. When the pain lifted, so too did the paralysis. During her pregnancy she experienced no migraines, but after the birth of her son the

migraines returned and became steadily worse. She had constant headaches, but she also had 2 or 3 migraines/week. As the pain got worse, she would develop a complete scotoma of the left eye, she was unable to speak, and the left hand and left foot became paralyzed. After a particularly severe episode she was prescribed Maxalt (rizatriptan), which she used daily to prevent the migraines.

The patient had been prescribed Maxalt (rizatriptan) elsewhere, despite the fact that the triptans are contraindicated in sporadic hemiplegic migraine.<sup>11</sup> She used it regularly because it was the only medication that relieved her pain. As the triptans are potent constrictors of the extracranial vessels,<sup>12</sup> we made a clinical examination of the terminal branches of the external carotid artery during an attack to find out whether the pain was indeed extracranial. Digital compression of the superficial temporal, frontal, occipital, and internal maxillary arteries all reduced the pain. As we know that the pain of migraine that originates in the extracranial terminal branches responds well to cautery of the involved vessels,<sup>9</sup> and as the patient was aware that digital compression of the extracranial arteries relieved her pain, she elected to have cautery. The involved vessels were cauterized on 26 September 2011, and she has had no further attacks 2 years later.

### Case 2

A 35-year-old woman had her first attack of migraine as a teenager. She had also had several isolated episodes of visual aura. The first attack of sporadic hemiplegic migraine occurred when she was in her early 20s. She described it as follows: “the first hemiplegic migraine attack happened when I was at work on the ‘phone. I had been feeling odd all morning but at the time did not recognize this as aura. I was finding it difficult to find the right words whilst on the ‘phone, and then the ‘phone fell out of my (left) hand. Shortly after that I lost the right side of my visual field”. She was taken to hospital, but during the journey the paralysis in the arm stopped and the headache began. The next attack was a few months later, and then a few weeks after that a continuous cycle established itself of a day of aura, a day or more of pain, a day of exhaustion, and then possibly a day without symptoms, and then back to the aura. Despite drug treatment by her consultant neurologists her condition worsened, and she also developed chronic facial pain.

She experienced multiple sensory disturbances. Her visual disturbances she described as follows – “things looking too bright/sharp, violet spots, pin point white flashes in the periphery of my vision, partial loss of visual field either completely or to a broken glass or patterned distortion, Alice in Wonderland syndrome”. She also experienced sensitivity to multiple sounds: “I get very disorientated if, for example, the radio is on in the kitchen and the TV is on in the living room. If you add people having a conversation to the mix I find it unbearable”; to smell: “nausea from strong smells – in particular coffee, cigarette smoke, synthetic floral smells like air

freshener”; and to sensation: “I have numbness and tingling on the right side of my face, occasional odd sensation swallowing, a feeling if I put my hands together that they are both touching another hand which is not mine”.

Her motor disturbances comprised speech: “I find it difficult to string a sentence together and remember words, I mumble and feel like I have cotton wool in my mouth”, and movement: “A violent tremor occurs in my jaw like shivering but much faster/harder, loss of movement and sensation in my left hand side of varying amount from clumsiness and poor fine motor control to complete paralysis of my limbs”. She also had occasional disturbances of mood – brief feelings of elation or depression.

On clinical examination during an attack, the superficial temporal main trunk and frontal branch, the occipital, and the angular arteries bilaterally, and the maxillary and posterior auricular arteries on the right, were identified as being involved. These vessels were cauterized on 10 October 2012, and she has not had another attack of migraine or hemiplegia since then.

### Case 3

This patient presented with a 3-month history of intense frontal headache, predominantly on the right side, accompanied by weakness affecting the right side, which rendered her unable to walk. Neurological examination was normal except for a subjective decrease in sensation to pinprick and light touch in her right upper and lower limbs. The sensory loss had a sharp area of demarcation that stopped exactly at the midline. A MRI was within normal limits, the only changes being mild mucosal thickening of the paranasal sinuses. She walked with a bizarre, stiff-legged gait, but with no circumlocution, scissoring, or shuffling. She had been admitted to hospital during this time for a month, and despite a lumbar puncture that was within normal limits and the absence of fever, nausea, vomiting, and neck stiffness, had been treated for bacterial meningitis. She had then been referred to a psychiatrist, but she did not keep the appointment.

On clinical examination during a headache (she was in a wheelchair at the time), digital pressure on the superficial temporal, occipital, frontal, posterior auricular, and angular arteries all gave temporary relief. The affected arteries were cauterized under sedation on 8 May 2013. Since then she has had no further headaches, and has recovered full use of her limbs.

### Discussion

Sporadic hemiplegic migraine is defined as migraine with an aura including motor weakness but with no first- or second-degree relatives having an aura, including motor weakness.<sup>1</sup> Several drugs have been recommended, but these were based on isolated case reports and series were too small to show a significant effect. The triptans and ergots are currently

contraindicated,<sup>11</sup> the triptans because of the hypothesis that the neurological symptoms in hemiplegic migraine are the result of vasoconstriction. By increasing vasoconstriction, the triptans could theoretically increase the risk of cerebral infarction.<sup>13</sup>

It has been suggested, though, that the proscription of triptans may be depriving patients of a safe and effective option for treatment.<sup>13</sup> In one study of 76 subjects with hemiplegic migraine, it was concluded that triptans seem to be safe and effective for most patients with hemiplegic migraine.<sup>14</sup> Despite the restriction on the use of triptans, one patient in the present study had been prescribed rizatriptan, which had proved to be effective in pain control without side effects. This was fortuitous, as it indicated that the extracranial vessels could be the origin of her pain, and this was confirmed on clinical examination.

### Conclusion

Cautery of extracranial arteries has previously been reported to be effective in migraine when the arterial component to the pain has been confirmed clinically.<sup>9,15–22</sup> This is to my knowledge the first report of the use of surgical cautery of the terminal branches of the external carotid artery in sporadic hemiplegic migraine. Although this is a small sample, the results so far seem to be most promising.

### Ethical statement

All patients in the study gave informed consent.

### Conflict of interest

There is no conflict of interest to declare.

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