Pearls & Oysters: Post Pipeline Headache Phenomenon: Nummular Headache Presenting After Intracranial Aneurysm Stenting

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Pearls
• Nummular headache is a headache disorder characterized by focal head pain in a well-circumscribed oval or elliptical-shaped area.
• Post-pipeline embolization headache is a recently described headache phenomenon that should be considered on the differential diagnosis for patients presenting with new onset headaches after intracranial flow-diversion stenting.

Oysters
• While nummular headache is considered a primary headache disorder, a similar headache phenotype can occur secondary to lesions of the bone, scalp and underlying intracranial structures.
• New headache following flow-diversion stenting could be related to in-stent stenosis or branch occlusions of perforating arteries, so additional evaluation is recommended.
Abstract

We report on a 31-year-old right-handed female with past medical history of pre-syncopal episodes and migraine headaches who presented to the outpatient clinic with a nummular headache after intracerebral stenting, which was different than her previous migraines. This represents “post-pipeline embolization headache” phenomenon, which is a relatively new term to describe a new or different headache in individuals who recently underwent intracranial vascular stenting as a treatment for cerebral aneurysms.

Case Report

A 31-year-old right-handed female with past medical history significant for pre-syncopal episodes and migraine headaches presented to the outpatient clinic for a new-onset persistent headache occurring immediately after intracerebral aneurysm stenting.

She described a long history of infrequent migraine headaches that had escalated in frequency and severity, without provocation. Magnetic resonance imaging (MRI) of the brain with and without contrast and a subsequent MR angiogram head without contrast revealed an unruptured saccular aneurysm measuring 5mm (height) x 4mm (width) x 4mm (neck), arising from the paraclinoid region of the right internal carotid artery. Two to three months after her headaches had increased in frequency, she had a cerebral angiogram with placement of a flow-diverting stent in the right internal carotid artery across the neck of the aneurysm.
Immediately after the procedure, she developed a new type of headache that was entirely different from her previous migrainous headaches. Instead of intermittent pain, she described a persistent sharp pain localized to an oval shape, about 1 inch wide and 3 inches long (longer over the anterior-posterior dimension) in the right parietal region. There was no rash in the region and no skin/hair change. The pain was present all day every day with associated allodynia, frequent paresthesia, and occasional sharp stabs in the oval area of pain. About 20 days per month, she would have brief episodes lasting seconds, where she would feel the paresthesia spread from the oval area to the hemicranium or the whole head. She would sometimes feel subjective warmth in the skin of the scalp in that region. She had not noticed any associated tenderness over the eyebrows or skull base. In the clinic, her neurological exam was unremarkable. She underwent several imaging studies, including computed tomography (CT) of the head without contrast, CT angiogram of the head and neck with and without contrast, MR angiogram of the head without contrast, and MR venogram, which were all unremarkable for acute pathologies. She also underwent a cerebral angiogram, which showed stabilization of her implanted flow-diverter device.

The patient was seen in our clinic 3 years after the onset of this persistent focal headache. For abortive headache therapies, she had previously tried butalbital approximately 2 days per week, acetaminophen (about 4 days per week), indomethacin (75 mg three times daily x 3 months), cyclobenzaprine, and ibuprofen without relief. For preventative headache therapies, she previously tried amitriptyline (small dose), topiramate (did not tolerate), and gabapentin (did not tolerate), without significant relief. During the clinic visit, she was diagnosed with nummular headache and started on a retrial of amitriptyline with a plan of alternative preventatives, including onabotulinumtoxinA, if she continued to have daily pain. During a follow-up visit, she
noted that the amitriptyline 50 mg daily was ineffective, but her response was confounded by her recent daily overuse of acetaminophen tablets and twice weekly use of butalbital/acetaminophen/caffeine tablets. Recommendations for the patient included initiation of onabotulinumtoxinA, continuation of amitriptyline 50mg daily, and weaning of daily over-the-counter pain medications.

**Discussion**

The best term to describe the patient’s new headache presentation is “post-pipeline embolization headache” phenomenon. This relatively new term describes a new or different headache in individuals who recently underwent intracranial vascular stenting as a treatment for cerebral aneurysms. A recent study performed a telephone survey of 88 individuals who underwent flow-diverting/Pipeline™ stenting for the treatment of cerebral aneurysms. Of the patients surveyed, 55% reported a new post-procedural headache with varying characteristics and associated symptoms. Individuals described their pain as dull, sharp, or throbbing, typically located ipsilaterally to the aneurysm/stent. The headaches started an average of 20 days following the procedure, and 69% of patients were still experiencing the headache an average of 21.6 months later. Most patients described intermittent rather than constant head pain, with an average headache frequency of 2 to 3 times per week, an average duration of 9.7 hours and an average 3.6/10 pain intensity. Interestingly, predictors for developing “post-pipeline embolization headache” include young age and prior history of headaches, both of which were present in our patient.

Numerous headache phenotypes can occur after post-flow diverter intracranial stenting. By her description, our patient’s post-flow diverter headache most closely resembled the phenotype of a nummular headache (NH), characterized by focal head pain in a well-circumscribed oval or elliptical-shaped area. The current ICHD-3 diagnostic criteria for NH
require the pain to be: 1) continuous or intermittent; 2) felt exclusively in the area of the scalp described as sharply contoured, fixed in size and shape, round/elliptical, and 1 to 6 cm in diameter; and 3) not corresponding better to another ICHD-3 diagnosis.²

The pain of NH is most often persistent, similar to our case, with up to 75% of published cases describing pain present for more than 3 months.² The painful area may be anywhere on the scalp but is typically located in the parietal region.² Pain is generally described as pressure-like, sharp or stabbing, and is of mild-to-moderate intensity, with occasional superimposed severe exacerbations.³ In a review of 250 patients with NH, the average age of onset was 45 years, with a female predominance.⁴ Like our patient, patients with NH commonly have abnormal sensation in the affected area and may describe variable combinations of paresthesia, dysesthesia, and allodynia in the affected area.² Treatment of NH resembles treatment for other primary headache disorders, with 55% of patients requiring preventative therapy.⁵ Gabapentin, indomethacin, tricyclic antidepressants and onabotulinumtoxinA have been reported effective in select cases.³-⁵ Unfortunately, NH can become refractory to abortive and preventive analgesic therapies, as seen in our patient’s presentation.

While NH is considered a primary headache disorder, numerous possible “secondary” etiologies have been described (see Table 1). Headaches resembling NH have occurred following trauma and in association with bony lesions such as fibrous dysplasia, craniosynostosis, Paget’s disease, and Langerhans histiocytosis.³ Some secondary cases have described pathology corresponding to the affected area, such as varicella-zoster viral infection, calcified hematoma, and linear scleroderma; other cases have a less clear association with the location of pathology, such as contralateral NH following a pituitary adenoma resection.³ Pertinent to our case, there
have been several cases of NH associated with vascular pathologies, although each of these were associated with fusiform areas of superficial vessels near the area of pain.³

Our patient developed ipsilateral NH following placement of a flow-diverting stent in the right paraclinoid ICA. Pain from stimulation within the ICA is not without precedent. In classic studies performed by Ray and Wolff in 1940, the intracranial carotid artery was found to be sensitive to pain brought on by stretching, stroking, and faradic stimulation.⁶ Focal referred pain has also been demonstrated ipsilateral to balloon inflation within the distal ICA.⁷ While the intracranial portion of the ICA is generally thought to refer pain to the frontotemporal region due to innervation from the trigeminal nerve, there is significant anatomic variation in the innervation of intracranial structures, with overlapping innervation by both trigeminal and the upper cervical nerves.⁸

Endovascular treatment of aneurysms using flow-diverting devices such as Pipeline™ stents is becoming relatively common. Most complications are asymptomatic, however severe complications can also occur including in-stent stenosis or potential branch occlusions of perforating arteries with a risk of ischemia. Therefore, any patient presenting with new unexplained headache following an endovascular stent procedure for an intracranial aneurysm requires additional evaluation. If the evaluation shows no structural abnormalities and the new post-procedural headache remains unexplained, post-pipeline embolization headache should be considered in the differential.
References


**Table 1: Reported secondary etiologies of nummular headache**

<table>
<thead>
<tr>
<th>Scap</th>
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<tbody>
<tr>
<td>Subcutaneous cyst</td>
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<td>Linear scleroderma</td>
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<td>Varicella-zoster viral infection</td>
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<td>Calcified hematoma of scalp</td>
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<td>Insect bite in the affected region</td>
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<td><strong>Bone</strong></td>
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<tr>
<td>Bone hemangioma</td>
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<td>Fibrous dysplasia</td>
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<td>Craniosynostosis</td>
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<td>Langerhans histiocytosis</td>
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<td>Paget’s disease</td>
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<td>Osteoma</td>
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<td>Cholesterol cyst in the bone</td>
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<td><strong>Intracranial</strong></td>
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<tr>
<td>Arachnoid granulation</td>
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<tr>
<td>Pituitary tumor (improved after resection)</td>
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<tr>
<td>Cranial surgery, trans-sphenoidal resection of pituitary tumor</td>
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<tr>
<td>Arachnoid cyst</td>
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<tr>
<td>Subtentorial meningioma</td>
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<td>Intracerebral cavernoma</td>
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<tr>
<td><strong>Vascular</strong></td>
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<td>Superficial artery aneurysms</td>
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<tr>
<td>Post-vascular stenting for aneurysm (current case)</td>
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<tr>
<td><strong>Other</strong></td>
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<tr>
<td>Head trauma</td>
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