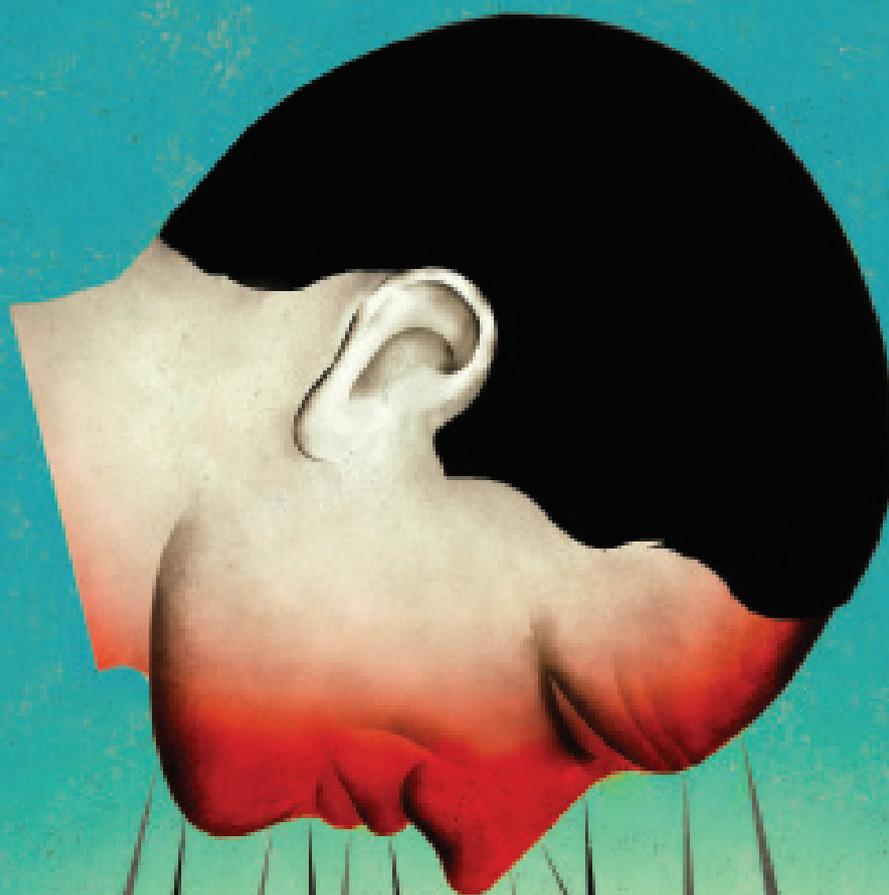


Facing Pain



Diagnosing and treating the intense facial pain known as trigeminal neuralgia.

BY DEBRA GORDON, M.S.

The pain came out of nowhere and struck like lightning, as if it were a bolt of electricity in her face. Then, just as suddenly, it was gone. But Mary Harmon of Broken Arrow, OK, knew it would return, triggered by putting on makeup, drinking a glass of water, even a slight breeze on her face. The fear of another attack of pain from trigeminal neuralgia (TN) kept Harmon at home, forcing her to give up her beloved gardening, the church choir, her life.

No wonder they used to call TN the “suicide disease.”

“It’s a horrible pain,” says Gary S. Gronseth, M.D., professor and vice chairman of the neurology department at the University of Kansas in Kansas City, KS, and Fellow of the American Academy of Neurology (AAN). “When you ask patients to rate their pain on a scale of zero to 10, they typically give a number greater than 10,” he says.

TN is a neurologic condition that affects less than 1 percent of the population in the United States but about 14 percent of those with nerve-related (neuropathic) pain. Some experts think the prevalence of TN is much higher because the condition is often misdiagnosed as a dental or other medical problem. It occurs more often in women, generally appearing in middle or late middle age. However, many men and some children also have TN.

Although effective medications are available for the condition, one study found that TN patients still have “moderately severe” pain despite 94 percent taking medications

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—GARY S. GRONSETH, M.D.

generally considered effective, likely due to undertreatment and lack of knowledge about TN among doctors. It is estimated that nearly half of primary care physicians misdiagnose the condition.

Harmon, now 75, is among the people whose TN pain continued despite taking drugs. Although her symptoms started in the early 1990s, it took until 2007 before she finally saw a neurologist and received a diagnosis.

WHAT CAUSES TN?

The trigeminal nerve is the major nerve serving the face. Its three branches carry sensations from the eyes, mouth, and jaw to the brain. The pain of TN, which often occurs on the right side of the face but can affect both sides, typically originates in the maxillary nerve, which runs along the cheekbone and serves the nose, upper lip, and upper teeth, or the mandibular branch, which controls sensation in the lower cheek, lower lip, and jaw.

The two main types of TN are classical and symptomatic. Classical TN is the most common, occurring suddenly with no obvious trigger. Symptomatic TN is related to some underlying condition such as a tumor, aneurysm, multiple sclerosis, meningitis, or Lyme disease, says Charles Argoff, M.D., member of the AAN as well as professor of neurology and director of the Comprehensive Pain Center at Albany Medical College in Albany, NY.

The worst part of the disease, besides the pain itself, is the fear of the attacks, according to Dr. Argoff. “They come out of nowhere, and a person can have multiple attacks within the same day. It makes people grimace and jerk their head,” he says. In fact, for a long time, TN was called *tic douloureux*, which means “painful tic” in French, because of the way patients would jerk their heads during an attack in response to the pain.

As for the cause of the classical form of the disease, Dr. Argoff says, “Exactly what causes the pain in classical TN is a matter of debate.” The most commonly held hypothesis is that the pain occurs when a vein or artery presses upon the trigeminal nerve where it enters the brain stem, which is the lower part of the brain that connects to the spinal cord.

According to this hypothesis, the contact creates inflammation that damages the nerve by stripping its myelin sheath, a layer of insulation that forms around nerves. Damage to the myelin sheath, which may occur gradually and initially go unnoticed, interferes with the ability of a nerve to conduct sensation normally, says Dr. Argoff. As a result, the nerve will fire at the slightest provocation, leading to jolts of pain out of proportion

with the trigger. Sometimes surrounding nerve cells are affected as well, increasing the sensory signals and the pain.

Yet, notes Dr. Argoff, “Some people with TN have normal looking arteries and nothing compressing the nerve, so it’s important to keep in mind that another explanation is also likely.”

DRUGS FOR THE PAIN

Rosalyn Tarnoff (not her real name), a 69-year-old woman from Erie, PA, had her first TN attack in her late 40s. She experienced the pain on both sides of her face. For about a year, she bounced between her primary care doctor and dentist; neither of them could find any reason for her pain. Finally, in agony, she insisted that her doctor refer her to a neurologist.

The neurologist sent Tarnoff for an MRI to rule out any underlying problems and then started her on the most commonly prescribed medication for TN: carbamazepine (brand name Tegretol). Although best known as an antiepileptic drug (AED), carbamazepine was actually approved more than 40 years ago to treat TN.

Still, Tarnoff was skeptical. “I didn’t like taking the medicine. I’m not a medicine person,” she says. But the pain was so severe, she really had no choice.

About 80 percent of patients with TN have a “dramatic response” to carbamazepine or its chemical cousin, oxcarbazepine (Trileptal), says Dr. Gronseth. Neurologist Kavita M. Grover, M.D., of Henry Ford Health System in Detroit, MI, assistant professor at Wayne State University in Detroit, MI, and member of the AAN, says it is

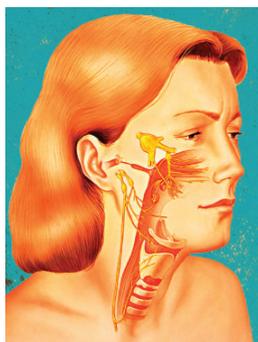
“quite gratifying to see that the correct medication at the right dosage” can lead to such an improvement.

Patients on carbamazepine require regular blood tests to monitor levels of the drug because it can cause hyponatremia, or low sodium levels (oxcarbazepine does not). More common, less serious side effects include drowsiness, unsteadiness, and vomiting.

However, about 20 percent of people with TN “wind up on a merry-go-round” of other AEDs and antidepressants, says Dr. Grover, for which there is very little evidence of their effectiveness.

That’s what happened to Tarnoff. Although the carbamazepine brought her pain down to a manageable level, it didn’t entirely erase it. In November 2010, she got sick with an unrelated illness, and the pain returned in full force despite the carbamazepine.

Her doctor started her on another AED, pregabalin (Lyrica), but the side effects were too much for her. “I couldn’t sleep, I had double vision, and I felt like I was having nightmares when I was awake,” Tarnoff recalls. Finally, she pulled out the patient



TRIGEMINAL NERVE
The trigeminal nerve, in yellow, has three major branches: ophthalmic, maxillary, and mandibular.

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information form she'd received with the drug and learned these were all possible side effects. She stopped taking the medication immediately. The next day, her TN pain returned.

After prescribing several other AEDs, her doctor finally suggested surgery.

Prescribing one medication after another can be a fruitless pursuit that just delays the inevitable, according to Dr. Gronseth. Meanwhile, “the patient is suffering,” he says. While it's reasonable to try one more medication after the carbamazepine, he says, “if the person is not better, it's time to talk about some type of procedure.” (Go to <http://bit.ly/ybeifg> to read the AAN's guideline on evaluating and treating trigeminal neuralgia, which was authored by Drs. Gronseth, Argoff, and colleagues.)

Trigeminal Neuralgia: The Basics

According to the U.S. National Library of Medicine, symptoms of trigeminal neuralgia (TN) include:

- ▶ Very painful, sharp, electric-like spasms that usually last a few seconds or minutes but can become constant
- ▶ Pain on one side of the face, often around the eye, cheek, and lower part of the face (although it can occur on both sides of the face)
- ▶ Pain triggered by touch or sounds
- ▶ Pain triggered by common, everyday activities, such as brushing teeth, chewing, drinking, eating, lightly touching the face, shaving the face

Patients may undergo an MRI or CT scan during the diagnosis process to make sure they don't have a tumor pressing on the nerve or other medical problems that can cause TN. However, the condition is typically diagnosed by ruling out other medical conditions.

Once diagnosed, patients may need to take medication indefinitely, says Dr. Grover. “This condition has almost a cyclical pattern,” she explains. “There are times when it becomes quieter, and then phases when it is very active.” So medication doses may need to be adjusted accordingly.

Finally, joining a support group, whether in person or online, can help people with TN feel more in control of the condition, experts say.

SURGERY

Although four types of procedures are available to treat TN, little evidence exists as to which is best. The four procedures include peripheral nerve blocks, percutaneous procedures, gamma knife, and microvascular decompression.

Peripheral nerve blocks involve the doctor attempting to block the nerve with anesthetics such as lidocaine.

Percutaneous procedures involve inserting a tiny needle through the base of the skull where nerves exit and damaging the trigeminal nerve with heat, an injection of glycerol (a form of alcohol), or balloon compression. Damaging the nerve can short-circuit its ability to transfer pain signals.

While more effective than nerve blocks, percutaneous procedures may need to be repeated. Overall, studies suggest that about half of patients who undergo the procedure report they are pain free at five years. More than half, however, experience facial numbness and difficulty chewing.

Gamma knife is the least invasive procedure. Doctors use a robotic device to deliver beams of high-dose radiation to destroy the nerve. It can take weeks or even months for the pain to dissipate, although sometimes patients have immediate pain relief. The main risk is facial numbness. About half of patients report they are still pain free at three years.

Finally, in microvascular decompression, a piece of the skull is removed behind the ear, exposing the trigeminal nerve. The surgeon then removes any blood vessels in contact with the nerve. It is the most invasive procedure, yet many experts believe it has the best results. However, it also entails the greatest risks, with between 0.2 and 0.5 percent of patients dying from the procedure. Complications include leaking spinal fluid, serious bruising, meningitis, double vision, and facial muscle weakness. About 10 percent of patients lose their hearing in one ear.

A PAIN-FREE FACE

Tarnoff was not a good candidate for decompression surgery because the neurosurgeon could not confirm that the nerve was compressed. She is now back on the carbamazepine and has been prescribed another AED, levetiracetam (Keppra). The pain is better, she says, but not completely gone.

Harmon, however, underwent gamma knife radiotherapy. She was awake during the 45-minute procedure listening to the Frank Sinatra music piped into the operating room. The pain disappeared immediately.

Although she's had a few TN episodes since then, “they were very scattered and very brief,” says Harmon. And she is back to gardening and singing in the church choir.

“A pain-free face is a lovely thing,” she says.

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