Chapter 1

Do I Have a Brain Tumor?

Tumor

In all likelihood, you do not, but it is an important question to ask and one that medicine should take seriously and address adequately, because reassurance is key. In the question, brain tumor does not just stand for a tumor in the brain, which can be cancer but does not have to be.

Tumor in medicine is a general term for swelling, which can be due to a number of causes and, again, does not have to be cancer. In the brain, any swelling of significance causes headache, because a hard structure, the skull, surrounds it and does not give. Within the skull, swelling increases the pressure, and that is painful, although generally not terribly.

The swelling can be in part of the brain or between the brain and the skull, but it can also affect the brain as a whole. When the latter is the case, we call it benign intracranial hypertension or, better, idiopathic intracranial hypertension. We also refer to it as pseudotumor cerebri, because it presents as a brain tumor while it is not.

We see idiopathic intracranial hypertension, or IIH, most commonly in obese young women, and the cause is generally unknown. A blood clot in one of the big veins draining blood from the brain can be the reason for the swelling. Occasionally, the cause is the use of tetracycline, an antibiotic, or taking too much vitamin A, an old-fashioned treatment for acne. It typically causes persistent, low-grade headache, associated with impaired vision due to swelling of the optic discs.

The optic disc is the site of entry of the optic nerve in the back of the eye. We can inspect it by examining the eye ground, which is part of a physical examination called fundoscopy, important in the context of evaluating a patient with headache.

The optic nerve is an extension of the brain and swells when the pressure in the skull is increased. Thus, looking into the eyes and examining the eye grounds is an important way of obtaining reassurance that a brain tumor, or anything else that increases the pressure in the skull, is not the cause of the headaches.

Apart from general swelling of the brain or a brain tumor, increased pressure can be due to enlargement of the chambers of the brain, called hydrocephalus. Other causes of increased pressure in the skull are the presence of blood in or around the brain or a brain abscess.

Imaging

A much better way of addressing the brain tumor question than by examining the eye grounds through fundoscopy is to visualize the brain through techniques, such as computerized tomography (CT) or magnetic resonance imaging (MRI).

When we use computerized tomography to picture the brain, no contrast is required if the headache is acute and bleeding is what we want to exclude. Bleeding can result from injury to the head, from a stroke, also called brain attack, or from rupture of an aneurysm of a brain artery, which is a localized expansion that looks like a little balloon.

The rupture of such a little arterial balloon is a catastrophic event and is associated with headache that comes on like a blow to the head. In medicine, we call a headache that comes on this way—that is, like a blow to the head—a headache of very acute or peracute onset or a thunderclap headache. It is characteristic of a condition called subarachnoid hemorrhage, which is a bleeding between the two inner membranes surrounding the brain. It presents a medical emergency, because half of the people who suffer from it unfortunately die.

If we perform computerized tomography of the brain for reasons other than to exclude bleeding, as in the case of post-traumatic headache or headache of peracute onset, we should do it with intravenous iodine contrast. This is important in case a suspected blood vessel abnormality, such as an arteriovenous malformation, or AVM, causes the headaches.

An arteriovenous malformation is a congenital abnormality that is present at birth. When affecting the brain, it can manifest itself as migraine—in this case, migraine with aura. It is usually present in the back of the brain, where vision is located, and the resulting aura symptoms are visual in nature.

Aura symptoms in the context of migraine originate in the brain and occur before the onset of the headache or during the initial phase of headache development. They are either visual in nature, affecting vision on one side or the other; somatosensory, affecting sensation on one side of the body or the other; or involve speech.

In case of an arteriovenous malformation in the back of the brain, a so-called occipital arteriovenous malformation, the aura symptoms are visual in nature, and the resulting disorder is that of symptomatic migraine with visual aura. The following patient vignette is an example of an occipital arteriovenous malformation manifesting as migraine.

A thirty-seven-year-old man has been experiencing headaches since college. Five years ago, they occurred once per month; after that, two or three times per year. However, over the last ten days, he experienced them five times.

The headaches come on during the day, usually in the afternoon, build to their maximum intensity in five to ten minutes, and last six to twelve hours. They are located on the left, in the forehead and behind the eye. The headaches feel like an intense pressure, and in half of them, the pain is severe. They are associated with light sensitivity and, occasionally, also with nausea and vomiting.

A visual disturbance precedes the headaches by ten to twenty minutes, consisting of blue spots in the right visual field. The visual disturbance lasts for the duration of the headaches. Bright light, overexertion, red wine, monosodium glutamate (MSG), and drinking more than two cups of coffee bring on the headaches.

Brain magnetic resonance imaging revealed a large arteriovenous malformation in the left occipital lobe. With a special procedure, surgeons made it smaller, causing permanent loss of vision in the process. Unfortunately, the headaches continued unabated, along with the visual disturbance.

Superior to computerized tomography of the brain is magnetic resonance imaging. When performed to address the brain tumor question, it does not require intravenous contrast, in this case gadolinium. As opposed to brain computerized tomography, it does not involve radiation, which is an advantage as well. Finally, it has better resolution, as we call it, allowing more detailed examination of the brain, including its blood vessels, the nerves originating from the brain stem, and the membranes surrounding it.

The brain itself is insensitive to pain, and therefore, any condition affecting it must be big enough to stretch its blood vessels, which are pain sensitive, to cause headache. Generalized swelling of the brain will do this, as well as enlargement of the chambers of the brain, or hydrocephalus. A tumor in the brain, whether cancer, bleeding, or abscess, will do it too, also as a result of associated swelling due to edema. Inflammation, usually from a viral infection, so-called viral encephalitis, also causes headache through localized or generalized swelling of the brain.

Meninges

In contrast to the brain, the membranes surrounding it, the meninges, three in total, are sensitive to pain like the brain blood vessels. The two inner membranes can become inflamed because of a viral, bacterial, or fungal infection called meningitis. This is a concern particularly in young children, teenagers, and adolescents.

In this case, the headache is associated with fever and with a particular kind of stiffness of the neck. This particular neck stiffness makes it difficult to bend the neck forward while it moves well in every other direction. We encounter the same kind of neck stiffness with subarachnoid hemorrhage, which is a bleeding between the two inner membranes of the brain. However, in the latter, there is no fever, and the headache has a very acute or peracute onset, like a blow to the head.

Bleeding can also happen between the two outer membranes surrounding the brain and between the outer membrane, called dura mater, and the skull. When it occurs between the dura mater and the skull, a so-called epidural hematoma, it is always due to a head injury, generally one that is impactful enough to fracture the skull. This is not the case when the bleeding happens between the two outer membranes of the brain, called a subdural hematoma. In this case, a head injury still causes the bleeding, but it is generally mild and, hence, easily forgotten.

Subdural hematoma predominantly occurs in the elderly, in whom shrinkage of the brain stretches the veins between the brain and the skull. This makes these so-called bridging veins vulnerable to injury, and a mild head injury, such as knocking the head against an open cabinet door, can cause them to bleed. The result is a venous bleed, which by its very nature is not as catastrophic as the arterial bleed from an aneurysm in subarachnoid hemorrhage. It generally causes a persistent, low-grade headache, which may be accompanied by memory impairment.

If we do not consider the possibility of a subdural hematoma in an older patient with recent-onset headache, we will fail to diagnose it. It may then be fatal, although we can easily detect it with the aforementioned imaging techniques and treat it with a relatively simple surgical procedure.

The outer membrane of the brain, the dura mater, can also be the source of a tumor, which is generally a benign tumor called meningioma. It is slow growing, develops over years or decades, and can manifest itself as chronic headache, including migraine. If it presents as migraine, it would be migraine without aura, and the condition would be that of symptomatic migraine without aura. The following patient vignette is an example of a meningioma manifesting as chronic tension headache.

A thirty-three-year-old woman has been experiencing headaches for seven years, initially occurring infrequently and mild in intensity. Over time, they gradually increased in frequency and have been almost daily for the last year. The almost daily headaches usually come on around noontime and are worst in the late afternoon or early evening. They are generally located in the right forehead but sometimes across the forehead. The headaches are not associated with nausea, vomiting, light sensitivity, or noise sensitivity.

Computerized tomography of the brain with iodine contrast revealed a meningioma in the front of the head, located on the right. Removal of the tumor resulted in resolution of the headaches.

Children

In children, brain tumors are often located in the small brain, or cerebellum, which is the part of the brain that resides in the back of the head and plays an important role in equilibrium. These tumors grow over weeks or months, ultimately compressing the fourth brain chamber, through which the brain fluid emerges from the chamber system.

The medical term for brain fluid is cerebrospinal fluid, or CSF, which is made in the chambers of the brain called lateral ventricles. The cerebrospinal fluid passes through the brain stem to the fourth brain chamber, or fourth ventricle, on its way flowing through the third ventricle, which is in the brain stem as well but in the front while the fourth ventricle is in the back.

The fluid emerges from the ventricular system in the brain and brain stem via the roof of the fourth ventricle and is absorbed in the blood through the veins on top of the brain. When something compresses the fourth ventricle, for example a tumor, the brain fluid cannot emerge from the ventricular system, and the chambers, or ventricles, enlarge to cause what we call hydrocephalus.

Hydrocephalus renders severe headaches, often associated with so-called projectile vomiting. They typically happen at night, waking the child from sleep or causing them to wake up in the morning with severe headache. Although the occurrence of headaches at night is something that we encounter in adults with nocturnal migraine or cluster headache, it is an ominous presentation in a child, requiring expeditious action in terms of brain visualization, preferably consisting of magnetic resonance imaging.

Sometimes, viral meningitis is mistaken for migraine, but here we are dealing with a single episode while migraine comes in repeated episodes, also referred to as attacks, over a longer period. With bacterial meningitis, such a mistake is unlikely to happen, because the patient is very sick with high fever and may be confused or even delirious. In both circumstances, a physician should perform a spinal tap or lumbar puncture for proper diagnosis, but not before picturing the brain with computerized tomography or magnetic resonance imaging.

Elderly

In an older patient with a relatively recent onset of headaches, subdural hematoma is not the only concern; we also need to consider two other medical conditions, namely, temporal arteritis and ophthalmic zoster.

Temporal arteritis, better termed giant cell arteritis, is a disorder that causes inflammation of the body's arteries including those of the head, leading to persistent, generally low-grade headache, often associated with fatigue, low-grade fever, and muscle pain. The muscle pain may happen particularly in the jaw muscles while chewing, which we refer to as jaw claudication.

We diagnose the condition through blood tests of inflammation, measuring the erythrocyte sedimentation rate, or ESR, and the C-reactive protein, or CRP, level. We confirm the diagnosis by microscopically examining a section of the blood vessel that runs in the temple—the superficial temporal artery.

Although the headache may be on one side only, it is prudent to take sections of both temporal arteries. These sections need to be relatively long, because the inflammation may not affect the entire length of the arteries.

Blindness and stroke are potential consequences of giant cell arteritis. Treatment requires the long-term intake of a corticosteroid and, hence, the need for solid confirmation of the diagnosis. We follow progress in treatment by repeating the aforementioned blood tests of inflammation.

Ophthalmic zoster is a presentation of shingles, which is due to reactivation of the chicken pox virus residing in the body. The virus inhabits nerve knots, known as ganglia—in this case, the so-called sensory ganglia, of which the trigeminal ganglion is one.

The trigeminal ganglion is located in the skull, at the base of the brain, and has three branches, hence the name trigeminal (tri = three). These are the ophthalmic branch, the maxillary branch, and the mandibular branch, which carry sensory information from the forehead, cheek, and jaw respectively to the brain.

We have a trigeminal ganglion on either side, each with a trigeminal nerve, and with ophthalmic zoster, the ophthalmic branch of one of them is inflamed. This causes pain on one side or the other of the forehead because of reactivation of the chicken pox virus in the trigeminal ganglion. Little blisters develop in the painful area, singularly or in small groups, several days after the onset of the pain. Sometimes, the blisters are difficult to see because they are hiding among the eyelashes or in the hairline.

The treatment of ophthalmic zoster is with an antiviral medication, like acyclovir or valacyclovir, and with a medication for nerve pain, such as gabapentin or pregabalin. It is also important to follow up with an eye specialist, or ophthalmologist, because the condition may affect the cornea and, therefore, can cause blindness.