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Clinical Mimics of SIH

Morning, everybody. I am really sorry that I can't be there to join you in person, but after that excellent introduction to the diagnosis of SIH by Dr. Birlea, I am going to talk through a few conditions that are frequently misdiagnosed in people who are suspected of having SIH. And I chose these because of what my patients report to me.

So let's get started. My only relevant disclosure is that I received grant support from the Spinal CSF Leak Foundation when I was at UT Southwestern. Thank you. And, uh, also I'm on the Medical Advisory Board for same. Let's start with chronic migraine, which I think is probably the most common misdiagnosis I hear about.

So, chronic migraine basically is defined as having headaches more often than not, at least 15 days out of the month for at least 3 months, and the headache on at least half of those days qualifies as being a migraine. No other cause was identified, and there may or may not be excessive use of acute medications.

The headache is defined using the criteria from the International Classification of Headache Disorders. And regarding the characteristics of the headache, there should be at least two of the following. The pain is one sided, it's throbbing or pounding, moderate to severe in intensity, and the one that helps me the most is "worse with routine physical activity."

And then, regarding associated symptoms, you need one out of two, either sensitivity to light and noise, or nausea and or vomiting. How do we distinguish chronic migraine or vestibular migraine from a spinal fluid leak? People with chronic migraine just don't wake up one morning with chronic migraine. It evolves over a period of time from less frequent migraine attacks.

Whereas spinal fluid leaks are often the other way around. They can start very suddenly. Sometimes they're thunderclap in onset. The most common time to experience a migraine headache is actually early in the morning. Sometimes when people first wake up, the migraine is already present. Spinal fluid leaks are typically the other way around.

People generally feel best when they first wake up in the morning, and their headache starts once they're out of bed or as the day progresses. When we ask people about the effect of lying down, It's important for us to choose our words carefully. Asking somebody, does your headache get better when you lie down?

Yes, that's usually the case with a spinal fluid leak, but sometimes people with migraine will misinterpret that question because people with migraine have headaches that are worse with activity and they prefer to lie down being motionless. The headache doesn't actually get better because they lie down.

The headache gets better because they go to sleep. Okay, that's totally different than with a spinal fluid leak, where it's not necessary to go to sleep. Of course, spinal fluid leaks produce orthostatic headaches, which are not present with chronic migraine. Other neurologic symptoms are possible with either, but the brain MRI usually helps us sort this out.

Let's move on to post-concussion syndrome or traumatic brain injury. Concussion is very common, and the most common symptom of concussion is headache. The risk factors for having a headache after a concussion are female sex, a family and personal history of headache or migraine, a history of prior concussion, and a pre-existing mood disorder.

The headache of concussion is typically characterized as migraine-like, less commonly as tension type–like, and least commonly like cluster headache or a different headache in that family. The pain is usually on both sides of the head. It's moderate to severe in intensity, and it can be effectively treated with acute or preventive migraine medications.

The other symptoms that can occur after a concussion often make it difficult for us to sort out whether the patient has post concussive syndrome or a spinal CSF leak. And those range from cognitive problems to sleep trouble, neck pain, dizziness, vertigo eye movement difficulties, and psychological issues.

And studies have been done looking at various aspects of those. factors that predict the persistence of a post-concussive syndrome at three months. So how are we going to sort this out? A history of head or neck trauma is required for either concussion or traumatic brain injury, but many people with a CSF leak have symptoms that start after head or neck trauma.

If the symptoms start after surgery, the surgery should be on the brain for or on the head for a TBI. but surgery on the spine can predispose to a CSF leak. The headache is typically migraine-like for a concussion, but it can be, sound like anything, but it's orthostatic at the CSF leak. You can have neck pain with either condition and vestibular problems with either condition,

but trouble with hearing and tinnitus are typically more commonly found with CSF leak. Brain fog can occur with either one, as can sleep difficulties, but people with a TBI or concussion typically experience insomnia, where those with a CSF leak have waking up in the middle of the night with headache.

Mood disorders don't really help us. And if the brain MRI shows changes of a spinal fluid leak, then we have a home run. New Daily Persistent Headache is a primary headache disorder with a distinct and very clearly remembered onset that becomes continuous and doesn't go away within 24 hours after onset.

It has to be present for at least three months to make the diagnosis. And it's not attributable to any other international classification of headache disorders diagnosis. When taking the history, it sounds like migraine or it sounds like tension type headache. But keep in mind that SIH is actually a secondary cause

of new daily persistent headache. So how do we tell the difference? Mainly because of the presence of orthostatic headache in a CSF leak and the presence of other neurologic symptoms with a leak. POTS can be extremely difficult to distinguish from a CSF leak. This is a condition that has no really apparent underlying cause, but it affects 1 to 2 percent of the US population, typically young white women who may have other coexisting conditions. It's diagnosed by demonstrating at least a 30 beat per minute increase in heart rate in adults, or 40 beats per minute or more in children, or an absolute heart rate of at least 120 beats per minute within 10 minutes of standing.

And there are a lot of secondary causes of POTS, including medications. deconditioning, post-COVID and others. Headache is common in people with POTS, and the overall prevalence of headache is about 37%. Orthostatic headache is reported to range anywhere from 2 percent to almost 60%, which is a pretty wide range.

Keep in mind that most symptoms of POTS get worse when the patient is upright. Most people with POTS have a non-orthostatic headache, and it sounds like migraine. A case control study of nine patients with SIH and 48 with POTS showed that orthostatic headache and neck stiffness were much more common with SIH.

Worsening with menses and myofascial pain were more common with POTS, as was syncope. And note that neck stiffness was only in present in people with SIH. But many of the symptoms were common to both. It is really complicated to distinguish POTS from SIH clinically. Orthostatic vital signs done in the office are quite helpful.

A tilt table test might be warranted, but neither one of these tests is always conclusive. Not all patients with orthostatic headache develop a headache during their tilt table test. And experiencing a headache during upright tilt doesn't correlate with orthostatic headaches at other times during daily activities.

Moreover, SIH patients can have an increased systolic blood pressure with head up tilt and heart rate variability with deep breathing identical to POTS. POTS can develop as a result of SIH due to deconditioning. So SIH patients may also have

POTS, as well as its associated manifestations. So which came first, the chicken or the egg?

Did the physical conditioning come first? Maybe arising from a CSF leak and leading to POTS? Do people with joint hypermobility have a higher predisposition to develop both of these conditions? It's really hard. So how are we going to figure this out? The headache of POTS usually sounds like migraine.

The headache of a CSF leak can pretty much sound like anything, but the location in POTS is typically in the front of the head or involves the whole head. In CSF leaks, the headache is often in the back of the head, but again, nothing specific about it. Orthostatic headache occurs, who knows, anywhere between 2 and 60 percent with POTS, but it's much more common with a CSF leak.

Joint hypermobility doesn't help us much, either condition can be associated with it. Eighth nerve involvement can also occur in both, but it's usually manifested a little differently. People with POTS usually experience dizziness that they describe as lightheadedness or wooziness. People with a CSF leak will describe a sensation of imbalance or spinning and may also have problems with their hearing or tinnitus. Syncope or fainting is common with POTS. We don't really see it very much with CSF leak. Tremulousness similarly is common with POTS. It can occur with a CSF leak, but it's not very common. The abnormal tilt table test can occur with either condition.

And when we're really, really stuck, we just hope that the brain MRI is going to be abnormal. Unfortunately, that doesn't always happen. Chiari malformations, in my opinion, are most commonly mistaken for CSF leaks because the imaging was read incorrectly. For example, this came off the internet from a radiology teaching site that was labeled as a Chiari malformation, when in fact it is clearly the brain of a spinal CSF leak with brain sag, pituitary enlargement, narrowing of the prepontine cistern, and just a little bit of tonsillar descent, whereas this one is a real Chiari malformation with a crowded posterior fossa

and descent of the tonsils. There are a lot of different manifestations of a Chiari, but the typical ones include headache that's provoked by coughing, sneezing, or laughing. Often the patients will have abnormal eye movements, and they may have other problems such as imbalance or incoordination.

So how are we going to sort this one out? Occipital headache is common to both. Orthostatic headache does not happen in Chiari unless the patient also has POTS. Neck pain is common to both. Syncope occurs in Chiari, or fainting, but it doesn't occur with a spinal fluid leak. And similarly to POTS, the dizziness in a Chiari is typically lightheadedness, whereas a spinal fluid leak is described as vertigo or imbalance.

And the answer should really be on the MRI. And in good hands, this should not really occur but there are distinct differences on the MRI between the brain of someone with a Chiari malformation and a spinal fluid leak.