

A Case Study: A Common Syndrome with an Unusual Presentation: Type III Achalasia following Herpes Zoster Infection Julie Forsberg, BSN, RN, CGRN

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Case Summarv

A 74 year old woman presented with an erythematous band like rash in a dermatomal distribution to her right upper back, extending to her right lateral chest. She was diagnosed with shingles, (Herpes Zoster), and discharged from the emergency department.

Twelve days later, the patient was admitted with odynophagia and dysphagia to liquids and solids, and could not even tolerate sips of water. An esophagogastroduodenoscopy was performed, and found to be normal, with no signs of esophagitis.

High resolution esophageal manometry (HREM) was performed, showing significant high pressurization and esophageal spasm throughout the body of the esophagus, Additionally, HREM revealed failed relaxation of the lower esophageal sphincter (LES). Based on these findings, the patient was diagnosed with Type III achalasia, possibly secondary to the shingles infection.

EGD with Botox injections to the LES was performed to provide temporary relief of symptoms.

Ten days post Botox treatment the patient was tolerating a general diet, without symptoms of odvnophagia or dvsphagia.

The interest of this case study is to consider the uncommon presentation of shingles coinciding with Type III Achalasia.

To understand the relationship of the two events, we have to consider the shingles virus itself, and the "Brain of the Gut".

The Enteric Nervous System (ENS) >Why the Brain of the Gut? The Enteric Nervous System (ENS) controls gut function, including motility. absorption, and secretion.

The Brain of the Gut:

>The Myenteric Plexus, a segment of the ENS, is a system of nerves (post ganglionic neurons) innervating the esophageal wall and LES. This plexus "manages" esophageal motility.

>The neurons in the plexus are distinguished between excitatory (Figure 1) and inhibitory (Figure 2) neurons.3



plexus result in coordinated waves of peristalsis pushing food forward

LES resting tone is also regulated by the balanced effect of excitatory and inhibitory neurons.

Herpes Zoster-Shinales



>This virus can reactivate late in life and cause a painful rash. Herpes Zoster (shingles), Anvone who has had varicella, or has gotten the varicella vaccine, can develop shingles.

autonomic nervous system ganglia.

>The shingles rash can occur anywhere on the body, but primarily appears on the trunk. The rash is described as itchy, painful and tingly.

>Varicella, the virus that causes chickenpox, is a virus that,

once resolved, remains inactive in the dorsal root ganglia and

Rash commonly seen in shingles

Post herpetic neuralgia is the most common complication. causing persistent pain where the rash was located.6



Normal HREM Swallow A. Normal peristaltic wave B. Relaxation of the LES

"If two improbable events occur together, the most probable explanation is they are linked "

Rafi Ali. MD

Putting it Together: Can the shingles virus cause achalasia?

>Achalasia is characterized by esophageal aperistalsis and failure of the LES to relax in response to a swallow. Symptoms include progressive dysphagia to solids followed by liquids.

> Causes of achalasia may be autoimmune, genetic predisposition, or viral. Herpes viruses exhibit a strong affinity for nerve fibers and can cause ganglion cell damage.²

> Viral infections such as shingles cause an immune-inflammatory reaction, activating T lymphocytes that attack ganglion cells in the myenteric plexus of the esophagus. The consequence of this reaction is destruction of inhibitory neurons (J. Pandolfino, personal communication, March 14, 2017),

>Nerve fiber damage results in loss of inhibitory neurotransmitters VIP and NO, causing loss of esophageal peristalsis and failure of LES relaxation.1

>Loss of esophageal peristalsis and failure of LES relaxation create the symptoms of dysphagia.

> Type III achalasia, characterized by pressurization in the body of the esophagus and esophageal spasm (Figures 3 and 4), is most likely the early stage of the disease process.⁵





B. Pressurization in the body of the esophagus

Figure 4 A. Failed relaxation of LES B. High amplitude esophageal

Conclusion

>In this case study, it is probable the shingles infection triggered an autoimmune-inflammatory response, resulting in significant decrease in ganglion cell concentration in the myenteric plexus, specifically the inhibitory neurons. The consequence of this process is loss of coordinated peristalsis in the body of the esophagus, and failure of LES relaxation.

>Local injection of Botox (Botulinum Toxin), was effective by counteracting the excessive LES tone that is seen in figures 3 and 4. The Botox blocked the release of acetylcholine (excitatory neuron), allowing relaxation of the LES. Symptom relief is due to lowered LES pressure.⁴

>The effect on the inhibitory neurons in the myenteric plexus due to the virus is likely irreversible.

>As of the time of this presentation, the patient continues to tolerate solids and liquids without dysphagia or odynophagia, but it is probable her symptoms will return when the effects of the Botox wears off.

A special thanks to Dr. Rafi Ali and Dr. John Pandolfino for their advice and support.

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