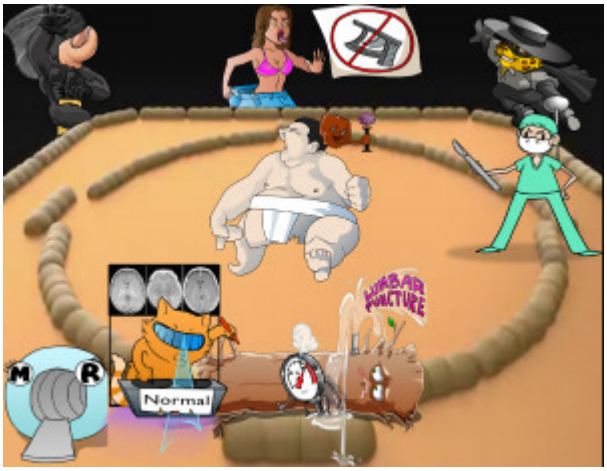
Pseudotumor Cerebri: Diagnosis and Management

Pseudotumor cerebri, or idiopathic intracranial hypertension, is characterized by elevated intracranial pressure with no known underlying cause. When there is sufficient clinical suspicion, definitive diagnostic tests are pursued such as a lumbar puncture, which would show elevated opening pressure. Labs and cultures of the collected cerebrospinal fluid should be normal. Neuroimaging with MRI or CT should also be obtained to exclude tumors or other underlying causes of increased intracranial pressure. Once diagnosis is confirmed, various treatment modalities are commonly pursued, with the most common being weight loss in obese patients. First-line pharmacotherapy is acetazolamide while topiramate may be helpful for headaches.



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Diagnosis

Lumbar Puncture with Increased Opening Pressure

Lumber-punctured with high-pressure gauge

A lumbar puncture is performed in order to confirm increased pressure. An elevated opening pressure greater than 200 mm H_2O and no sign of inflammation on cerebrospinal fluid analysis supports the diagnosis of pseudotumor cerebri.

Normal MRI or CT

CAT and MR-eyes with normal-sign

Neuroimaging with CT or MRI should be obtained in patients suspected of having pseudotumor cerebri. Imaging should demonstrate normal findings, as presence of a space-occupying lesion would indicate a specific underlying cause behind the intracranial hypertension, which would immediately rule out pseudotumor cerebri.

Management

Weight Loss

Woman with loose-fitting pants

For patients with pseudotumor cerebri who are obese, weight loss is recommended, as studies have shown reduction in symptoms following weight loss.

Acetazolamide

A Cheetah-Zorro

Carbonic anhydrase inhibitors such as acetazolamide are believed to reduce the rate of cerebrospinal fluid production, and their use has been shown to have associated improvement in outcomes in patients with pseudotumor cerebri.

Topiramate

Toe-primate

Topiramate is an anti-epileptic medication that inhibits carbonic anhydrase, and it's also commonly used in the treatment of migraine headaches, making it an attractive candidate for therapy in pseudotumor cerebri. It is thought to help mitigate symptoms by reducing production of cerebrospinal fluid, similar to acetazolamide.

Surgery

surgeon

For patients whose condition is refractory to pharmacologic management, there are surgical options that may be considered. One of these procedures is cerebrospinal fluid (CSF) shunting, in which a device is inserted to divert CSF into the peritoneum in order to relieve pressure. Another is optic nerve sheath fenestration, in which incisions are made in the optic nerve sheath, again to relieve pressure.

Avoid Triggers

Avoid-sign with Trigger

Part of the management of pseudotumor cerebri includes discontinuing any medications or supplements known to be associated with increased risk of developing the condition. There are multiple medications with these associations, however the most prominent to know are tetracyclines and vitamin A-containing compounds.