Shy-Drager Syndrome

Multiple system atrophy (MSA) with postural hypotension is also called Shy-Drager syndrome. It is a progressive disorder of the central and sympathetic nervous systems. The disorder causes a large drop in blood pressure when the patient stands up. This leads to dizziness or momentary blackouts. There are three types of MSA:

- Olivopontocerebellar atrophy (OPCA). This mostly affects balance, coordination, and speech.
- A parkinsonian form (striatonigral degeneration). This can resemble Parkinson's disease because of slow movement and stiff muscles.
- A mixed cerebellar and parkinsonian form.

In all three forms of MSA, the patient can have dizziness or momentary blackouts. Symptoms that happen early in the course of the disease include:

- Dizziness or momentary blackouts.
- Constipation. This may be hard to manage.
- Impotence in men.
- Urinary incontinence.

This syndrome may be hard to diagnose in the early stages. For most patients, blood pressure is low when the patients stand up. It is high when the patients lie down. Other symptoms that may develop include:

- Impaired speech.
- Difficulties with breathing and swallowing.
- Inability to sweat.

TREATMENT

Orthostatic hypotension in this syndrome is treatable. There is no known cure for the central nervous system degeneration. The general treatment course is aimed at controlling symptoms. These include:

- Anti parkinsonian medicine. L-dopa is an example.
- To relieve dizziness, increased salt and fluid in the diet may help.
- Medicine to elevate blood pressure is often needed. Salt-retaining steroids are an example. This medicine can cause side effects. It should be carefully monitored by a caregiver.
- Alpha-adrenergic medications, non-steroidal anti-inflammatory drugs, and sympathomimetic amines are sometimes used.
- Sleeping in a head-up position at night. This reduces morning dizziness.
- An artificial feeding tube or breathing tube may be surgically inserted. It is to help manage swallowing and breathing problems.

WHAT WILL HAPPEN

This syndrome usually ends in the patient's death. This happens 7 to 10 years after diagnosis. Common causes of death include:

- Aspiration. This is inhaling fluid into the lungs.
- Stridor. This causes high-pitched breathing sounds due to airway obstruction.
- Cardiac arrest.

RESEARCH BEING DONE

The NINDS carries out and funds research about disorders of the autonomic nervous system. This includes Shy-Drager syndrome. Research is aimed at discovering ways to diagnose and treat disorders of the autonomic nervous system. Ultimately the goal is to cure or prevent them.

Research is continually being done to learn more about this problem.

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