Von Hippel-Lindau Disease

Von Hippel-Lindau disease (VHL) is a rare, genetic multi-system disorder. It is characterized by the abnormal growth of tumors in certain parts of the body (*angiomatosis*). The tumors of the central nervous system (CNS) are benign. They are comprised of a nest of blood vessels. They are called hemangioblastomas (or angiomas in the eye). Hemangioblastomas may develop in the brain, the retina of the eyes, and other areas of the nervous system. Other types of tumors develop in the adrenal glands, the kidneys, or the pancreas. Individuals with VHL are also at a higher risk than normal for certain types of cancer, especially kidney cancer.

SYMPTOMS

Symptoms vary among patients. They depend on the size and location of the tumors. Symptoms may include:

- Headaches.
- Problems with balance and walking.
- Dizziness.
- Weakness of the limbs.
- Vision problems.
- High blood pressure.

Fluid-filled sacs (*cysts*) and/or tumors (benign or cancerous) may develop around the hemangioblastomas and cause the symptoms listed above.

TREATMENT

Treatment varies according to the location and size of the tumor and its associated cyst. In general, the objective of treatment is to treat the growths when they are causing symptoms but while they are still small. This is so that they do not cause permanent problems by putting pressure on the brain or spine, blocking the flow of cerebrospinal fluid in the nervous system, or impairing vision. Treatment of most cases of VHL usually involves surgery to remove the tumors before they become harmful. Certain tumors can be treated with focused high-dose irradiation. Individuals with VHL need careful monitoring by a caregiver and/or medical team familiar with the disorder.

WHAT WILL HAPPEN

The prognosis for patients with VHL depends on the location and complications of the tumors. Untreated, VHL may result in blindness and/or permanent brain damage. With early detection and treatment the prognosis is significantly improved. Death is usually caused by complications of brain tumors or kidney cancer.

RESEARCH BEING DONE

The NINDS pursues a vigorous program of research aimed at preventing and treating disorders that cause tumors in the brain and spinal cord such as VHL.

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

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