Retroperitoneal fibrosis

Description

Retroperitoneal fibrosis is a disorder in which inflammation and extensive scar tissue (fibrosis) occur in the back of the abdominal cavity, behind (retro-) the membrane that surrounds the organs of the digestive system (the peritoneum). This area is known as the retroperitoneal space. Retroperitoneal fibrosis can occur at any age but appears most frequently between the ages of 40 and 60.

The inflamed tissue characteristic of retroperitoneal fibrosis typically causes gradually increasing pain in the lower abdomen, back, or side. Other symptoms arise from blockage of blood flow to and from various parts of the lower body, due to the development of scar tissue around blood vessels. The fibrosis usually develops first around the aorta, which is the large blood vessel that distributes blood from the heart to the rest of the body. Additional blood vessels including the inferior vena cava, which returns blood from the lower part of the body to the heart, may also be involved. Obstruction of blood flow to and from the legs can result in pain, changes in color, and swelling in these limbs. Impairment of blood flow in the intestines may lead to death (necrosis) of intestinal tissue, severe pain, and excessive bleeding (hemorrhage). In men, reduced blood flow back toward the heart (venous flow) may cause swelling of the scrotum.

Because the kidneys are located in the retroperitoneal space, retroperitoneal fibrosis may result in blockage of the ureters, which are tubes that carry urine from each kidney to the bladder. Such blockages can lead to decreased or absent urine flow and kidney failure. When the kidneys fail, toxic substances build up in the blood and tissues, leading to nausea, vomiting, weight loss, itching, a low number of red blood cells (anemia), and changes in brain function.

Frequency

Retroperitoneal fibrosis occurs in 1 in 200,000 to 500,000 people per year. The disorder occurs approximately twice as often in men as it does in women, but the reason for this difference is unclear.

Causes

No genes associated with retroperitoneal fibrosis have been identified.

Retroperitoneal fibrosis occasionally occurs with autoimmune disorders, which result when the immune system malfunctions and attacks the body's own organs and tissues. Researchers suggest that the immune system may be involved in the development of retroperitoneal fibrosis. They propose that the immune system may be reacting abnormally to blood vessels damaged by fatty buildup (atherosclerosis) or to certain drugs, infections, or trauma. In many cases, the reason for the abnormal immune system reaction is unknown. Such cases are described as idiopathic.

Inheritance

Most cases of retroperitoneal fibrosis are sporadic, which means that they occur in people with no apparent history of the disorder in their family. In rare cases, the condition has been reported to occur in a few members of the same family, but the inheritance pattern is unknown.

Other Names for This Condition

- Ormond disease
- Ormond's disease

Additional Information & Resources

Genetic and Rare Diseases Information Center

- IgG4-related retroperitoneal fibrosis (https://rarediseases.info.nih.gov/diseases/9568/index)

Patient Support and Advocacy Resources

- National Organization for Rare Disorders (NORD) (https://rarediseases.org/)

Clinical Trials

- ClinicalTrials.gov (https://clinicaltrials.gov/search?cond=%22Retroperitoneal fibrosis%22)

Catalog of Genes and Diseases from OMIM

- FIBROSCLEROSIS, MULTIFOCAL (https://omim.org/entry/228800)

Scientific Articles on PubMed

- PubMed (https://pubmed.ncbi.nlm.nih.gov/?term=%28retroperitoneal+fibrosis%5BMAJR%5D%29+OR+%28%28Ormond+disease%5BTIAB%5D%29+OR+%28Ormond

's+disease%5BTIAB%5D%29%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+720+days%22%5Bdp%5D)

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Last updated July 1, 2013