

What is retroperitoneal fibrosis/Ormond's disease?

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Retroperitoneal fibrosis, also sometimes referred to as Ormond's disease, is a disorder characterized by inflammation and the development of scar tissue behind the membrane that surrounds the digestive system, called the retroperitoneal space.

Epidemiology

Retroperitoneal fibrosis is an uncommon disease that affects 1 person per 200,000-500,000 population per year, depending on the specific location.

It can affect any individual, but most patients present between the ages of 40 and 60 years old, and it is more prevalent in males than females. In many cases, diagnosis is delayed due to the non-specific nature of the symptoms.

The prognosis for patients with the disorder varies according to the type. It is relatively good for idiopathic retroperitoneal fibrosis, with a small long-term effect on quality and quantity of life. However, it is poor for malignant retroperitoneal fibrosis, with an average survival of 3-6 months following diagnosis.

Cause

The exact cause of retroperitoneal fibrosis is not known. However, it has been associated with various immune related conditions, suggesting that an autoimmune response may be involved in the pathology of the condition.

Approximately 1 in 3 patients with the disorder has been subject to immune suppression previously, due to malignancy, medications, aortic aneurysm or an infection, further supporting this hypothesis.

Symptoms

There are various symptoms associated with retroperitoneal fibrosis, although they are not specific to the condition and may be linked to other health conditions with similar presentation.

Over time as the condition worsens, the fibrotic tissue can lead to dull general pain in the lower abdomen, back or side. Other symptoms may include:

- Fever
- Peripheral oedema
- Phlebitis
- Deep venous thrombosis
- Weight loss
- Nausea and vomiting
- Fatigue
- Renal function abnormalities

Eventually, the fibrosis can obstruct the blood flow to the lower parts of the body, leading to a disruption in the normal bodily function. This can cause pain and swelling, in addition to damage to certain organs, such as the kidneys.

Diagnosis

The diagnosis of retroperitoneal fibrosis is often very difficult as the symptoms are non-specific and may also be indicative of a number of other health conditions.

If it is suspected, there are a number of indicative factors and tests that can help to make the appropriate diagnosis. For example, many patients also suffer from hypertension or renal insufficiency. Blood tests can help to detect abnormally elevated levels of C-reactive protein, urea, creatinine and other parameters. Diagnostic imaging techniques can also be used, particularly to evaluate the severity of the condition.

In some cases, a biopsy for immunohistochemical examination is needed to assess the disorder, which can help to

guide treatment plan decisions.

Management

The recommended treatment for patients with retroperitoneal fibrosis is not clear, due to inadequate therapeutic trials and research in the area.

It is currently suggested that surgical and pharmaceutical techniques are integrated to provide optimal care. The aim of treat is to preserve renal function and other organs, exclude malignancy and to provide relief of symptoms.

Surgical techniques include ureterolysis, intraperitoneal transposition, percutaneous nephrostomy, ureteral stenting, laparoscopic surgery and endourologic treatment.

Pharmacotherapy may include administration with glucocorticoids (e.g. prednisolone), immunosuppressants (e.g. azathioprine) or estrogen receptor antagonists (e.g. tamoxifen).

In addition, there are various other management techniques that may be employed to address the specific symptoms that are most problematic for the patient.

References

- <http://ghr.nlm.nih.gov/condition/retroperitoneal-fibrosis>
- <http://www.healthline.com/health/retroperitoneal-fibrosis#Overview1>
- <https://www.nlm.nih.gov/medlineplus/ency/article/000463.htm>
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Further Reading

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