About Adrenal Cancer

Overview

If you have been diagnosed with adrenal cancer or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- What Is Adrenal Cancer?

Key Statistics

See the latest estimates for new cases of adrenal cancer and deaths in the US.

- What Are the Key Statistics About Adrenal Cancer?

What Is Adrenal Cancer?

About the adrenal glands

The adrenals are small glands that sit above each of the kidneys. The kidneys are located deep inside the upper part of the abdomen.

The adrenal gland has 2 parts. The outer part, called the cortex, is where most tumors develop. The function of the cortex is to make certain hormones for the body. These hormones all have a similar chemical structure and are called steroids. They include:

- Cortisol causes changes in metabolism that help the body to handle stress.
- Aldosterone helps the kidneys regulate the amount of salt in the blood and helps regulate blood pressure.
- Adrenal androgens are hormones which can be converted to more common forms of the sex hormones estrogen and testosterone in other
parts of the body. The amount of these hormones that result from conversion of adrenal androgens is small compared to what is made in other parts of the body. The testicles produce most of the androgens (male hormones) in men. The ovaries produce most of the estrogens (female hormones) in women.

The inner part of the adrenal gland, called the medulla, is really an extension of the nervous system. Nervous system hormones such as norepinephrine and epinephrine (also called adrenaline) are made in the medulla. Tumors and cancers that start in the adrenal medulla include pheochromocytomas (which are most often benign) and neuroblastomas.

This document is about tumors and cancers of the adrenal cortex. It does not discuss tumors of the adrenal medulla. Neuroblastomas are covered in a separate document.

**Adrenal cortex tumors**

There are 2 main types of adrenal cortex tumors: benign (non-cancerous) and malignant (cancers). Most of these tumors are benign and are called adenomas. Cancers of the adrenal cortex are rare. These 2 types of tumors can sometimes be hard to tell apart when the cells are looked at under the microscope. Although experienced pathologists (doctors that are trained to diagnose diseases by looking at tissue under the microscope) can tell the difference in most cases, sometimes the only way to know for sure that the tumor is a cancer is when it spreads. If it spreads to lymph nodes or other organs and tissues, it is a cancer. Adenomas do not spread outside the adrenal gland.

**Adrenal cortex adenomas**

Most tumors of the adrenal cortex are not cancer. They are benign tumors known as adenomas. These tumors are small, usually less than 2 inches (5 centimeters) across. They usually occur in only a single adrenal gland, but sometimes affect both.

Most people with adrenal adenomas have no symptoms and are unaware that they have an adrenal tumor. Some of these adenomas are discovered by accident (incidentally) when CT or MRI scans of the abdomen are done because of an unrelated health problem. About 5% of people who have a CT scan of the abdomen are found to have an adrenal tumor that was not suspected. Many of these are nonfunctional, meaning that they don't make adrenal hormones. Sometimes these tumors are known by the nickname incidentalomas because they aren't causing problems and were only found by
accident.

Some adenomas produce too much adrenal steroid hormones. Sometimes the excess hormone can cause symptoms. Many of the hormone-related symptoms of adenomas are the same as those from adrenal carcinomas (cancers). These symptoms are discussed in the section, “Signs and symptoms of adrenal cancer.” Adenomas are much more likely than carcinomas to produce high levels of aldosterone, which can cause high blood pressure.

Treatment: Adenomas can be cured by removing the adrenal gland that contains the adenoma. Some adrenal adenomas that cause hormone-related symptoms can be treated effectively with medicines to block the production or actions of these hormones. This may be the best treatment choice for patients with other serious medical problems who might not be able to withstand a major operation.

The treatment of incidentalomas depends on the chance that it may be a cancer and whether or not it is raising hormone levels. When an adrenal tumor is found accidentally, tests are often done to see if it is making hormones. If it is, surgery is often recommended. Otherwise, surgery may only be recommended if it is likely to be a cancer. Small tumors are less likely to be cancer, and are often watched but not treated. The CT (or MRI) scan can be repeated in 6 to 24 months to see if the tumor has grown. If it has, it may need to be removed. If it hasn't grown, hormone levels will be watched over the next few years. If the tumor remains small and doesn't make any hormones, it may not need to be treated at all.

The remainder of this document refers to adrenal cancers only, and not to adenomas.

Adrenal cortical cancer

The type of cancer that develops in the cortex of the adrenal gland is called adrenal cortical carcinoma. It is also known as adrenocortical cancer (or carcinoma) or just adrenal cancer. In this document, the term adrenal cancer is used to mean cancer that starts in the adrenal cortex.

Adrenal cancer most often is discovered when:

- It is found accidentally on an imaging test done looking for something else.
- It produces hormones that cause changes such as weight gain and fluid retention, early puberty in children, or excess facial or body hair growth in
It starts causing symptoms because it has gotten very large. Large tumors can press on other organs in the abdomen, causing pain or a feeling of fullness. Generally, adrenal cancers are much larger than adrenal adenomas. An adrenal tumor larger than 5 or 6 centimeters (about 2 to 2 1/2 inches) is assumed to be a cancer. In one study, the average size of an adrenal cancer was about 13 cm (or 5 inches).

Most cancers found in the adrenal gland did not start there and are not adrenal cancers. Instead, they start in other organs or tissues and then spread (metastasize) through the bloodstream to the adrenal glands. For example, lung cancers, melanomas, and breast cancers often spread to the adrenals. Even when other cancers spread to the adrenals; however, they are still named after the place they started and are treated like other cancers that start in the same place. They are not considered adrenal cancer. Their treatment is described in our documents on these cancers.

What Are the Key Statistics About Adrenal Cancer?

Adrenal carcinomas are very rare and the real number diagnosed in the United States is not known. It is probably around 200 per year. They are much less common than benign adrenal tumors (adenomas), which are found fairly often among middle aged and elderly people. Adrenal tumors (most of which are adenomas) are found in about one in every 10 people who have an imaging test (like a CT or MRI) of the adrenal gland.

The average age of patients with adrenal cancer is around 46, but adrenal cortical cancer can occur in people of any age; even in children.
What's New in Adrenal Cancer Research and Treatment?

Research focused on adrenal cancer is currently under way. Imaging tests for diagnosing this cancer, medical laboratory tests to more accurately distinguish adenomas from carcinomas, and new treatments are being studied. Progress in this research tends to be slow because adrenal cancer is so rare. Other studies of more general aspects of cancer that can be applied to adrenal cancers as well as other types of cancers are also being done.

Chemotherapy

Although adrenal cancer can be hard to study, experts are looking for new drugs that may help as well as looking at the value of accepted treatments.

One ongoing important study (called ADIUVO) is testing the value of mitotane in the treatment of patients with early-stage adrenal cancers that have been removed with surgery. The goal of the study is to see if mitotane lowers the chance of the cancer coming back and helps patients live longer.

Targeted therapy

Targeted therapy is a newer type of cancer treatment that uses drugs or other substances to attack the programming that makes cancer cells different from normal, healthy cells. Each type of targeted therapy works differently, but all alter the way a cancer cell grows, divides, repairs itself, or interacts with other cells. Targeted drugs have been effective for several more common types of cancer but their value for adrenal cancer is still not known.

Cixutumumab is a targeted drug that shows promise in treating adrenal cancer. This drug blocks the effect of a certain hormone called insulin-like growth factor 2 (IGF2) that is suspected of increasing growth of adrenal cancers. In one study, giving this drug with another targeted drug called temsirolimus stopped tumor growth for months in many patients.

Some other targeted drugs have been studied in adrenal cancer, but have not been very helpful.

Genetics
Scientists are learning how changes in certain oncogenes and tumor suppressor genes can cause normal adrenal cortex cells to become cancerous. Understanding these genetic changes will help doctors develop better methods to diagnose this disease as well as treatments that are more effective and have fewer side effects than those currently available. Medical centers involved in research may ask their patients for blood samples and about diseases in other family members to learn more about adrenal cancer. This happens usually as part of studies. These studies are different from treatment studies. The goal of these studies is to enhance research of this rare cancer, to learn more about how adrenal cancer forms, and in the future find new targets for adrenal cancer therapy.

For example, there have been several studies looking at what genetic syndromes can lead to adrenal cancer (these syndromes were discussed in Adrenal cancer risk factors). International groups are working to understand how adrenal cancer develops. Hopefully, these efforts will give insight into how adrenal cancers develop and provide better targets for therapy.