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Glucagonoma

Glucagonoma is a very rare tumor of islet cells of the pancreas, which leads to an excess of the hormone glucagon in the blood.

Causes

Glucagonoma is usually cancerous (malignant). The cancer tends to spread and get worse if not treated.

This cancer affects some of the islet cells of the pancreas. As a result, the affected islet cells produce too much of the hormone glucagon.

The cause is unknown. Genetic factors play a role in some cases. A family history of the syndrome multiple endocrine neoplasia type I (MEN I) is a risk factor.

Symptoms

Symptoms of glucagonoma may include any of the following:

- Glucose intolerance (body has problem breaking down sugars)
- High blood sugar (hyperglycemia)
- Diarrhea
- Excessive thirst (due to high blood sugar)
- Frequent urination (due to high blood sugar)
- Increased appetite
- Inflamed mouth and tongue
- Nighttime (nocturnal) urination
- Skin rash on face, abdomen, buttocks, or feet that comes and goes, and moves around (necrolytic migratory erythema)
- Weight loss

In most cases, the cancer has already spread to the liver when it is diagnosed.

Exams and Tests

Your health care provider will perform a physical exam and ask about your medical history and symptoms.

Tests that may be done include:

- CT scan of the abdomen
- Glucagon level in the blood
- Glucose level in the blood
- Genetic tests

Treatment

Surgery to remove the tumor before it spreads is usually recommended. Chemotherapy may be done when surgery is not possible.

Support Groups

You can ease the stress of illness by joining a cancer support group. Sharing with others who have common experiences and problems can help you not feel alone.

Outlook (Prognosis)

Approximately 60% of these tumors are cancerous. It is common for this cancer to spread to the liver. Only about 20% of people can be cured with surgery.

If the tumor is only in the pancreas and surgery to remove it is successful, people have a 5-year survival rate of 85%.

Possible Complications

The cancer can spread to the liver. High blood sugar level can cause problems with metabolism and tissue damage.

When to Contact a Medical Professional

Contact your provider if you notice symptoms of glucagonoma.

Alternative Names

Glucagonoma syndrome; 4D syndrome; Endocrine-secreting islet cell tumor - glucagonoma

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Updated by: Sandeep K. Dhaliwal, MD, board-certified in Diabetes, Endocrinology, and Metabolism, Springfield, VA. Also reviewed by David C. Dugdale, MD, Medical Director, Brenda Conaway, Editorial Director, and the A.D.A.M. Editorial team.

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