CONDITION

Glucagonoma

Glucagonoma is a rare hormone-producing tumor, usually in the pancreas, that can cause a distinctive skin rash, weight loss, and high blood sugar.

WHAT IT IS
Glucagon-secreting neuroendocrine tumor, usually arising in the pancreas

BODY SYSTEM
Pancreas, hormones, skin, and metabolism

MAIN SYMPTOMS
Migrating rash, weight loss, high blood sugar, diarrhea

WHAT CAUSES IT
Usually sporadic; rarely linked to inherited MEN1

HOW IT IS DIAGNOSED
Fasting glucagon testing, tumor imaging, and tissue examination

MAIN TREATMENT
Surgery when possible; hormone and tumor-control treatments

CAN IT BE CURED?
Sometimes - surgery can cure localized tumors

IS IT AN EMERGENCY?
Sometimes - chest pain or sudden breathlessness

LONG-TERM OUTLOOK
Better when localized; advanced tumors often need long-term control

Overview

A glucagonoma is a rare type of neuroendocrine tumor. Neuroendocrine tumors begin in cells that make hormones. Most glucagonomas start in the pancreas, in cells that normally produce the hormone glucagon.

Glucagon helps keep blood sugar from becoming too low. A glucagonoma may release much more glucagon than the body needs. This can raise blood sugar and affect how the body uses fats and proteins.

The combination of a glucagon-producing tumor and its effects on the body is often called glucagonoma syndrome. Its best-known feature is a distinctive skin rash called necrolytic migratory erythema , or NME. The rash often changes position over time and may be sore or itchy.

Some pancreatic tumors contain glucagon but do not release enough to cause this syndrome. Doctors use symptoms, blood tests, scans, and examination of tumor tissue to work out whether a tumor is a glucagonoma.

Glucagonomas may grow slowly, but many have already spread by the time they are found. The liver is a common site of spread. Finding out whether the tumor is confined to the pancreas helps guide treatment and gives important information about the outlook.

Symptoms

Symptoms often develop gradually. They can appear months or years before the tumor is identified.

Necrolytic migratory erythema is the most characteristic symptom. It usually begins as red patches that may blister, crust, peel, or become darker as they heal. New patches may appear while older ones improve. Common sites include the groin, buttocks, lower abdomen, thighs, feet, and around the mouth.

Other possible symptoms include:

These symptoms are not unique to glucagonoma. Skin conditions, diabetes, nutritional deficiencies, liver or kidney problems, and other conditions can cause similar changes. A medical assessment is needed to find the cause.

- Unplanned weight loss, sometimes despite a reasonable appetite
- High blood sugar or newly diagnosed diabetes
- Increased thirst or urination
- Diarrhea or frequent bowel movements
- A sore, smooth tongue or cracks at the corners of the mouth
- Tiredness caused by anemia, meaning too few healthy red blood cells
- Muscle loss, weakness, or poor nutrition
- Low mood, irritability, or other emotional changes
- Blood clots, especially in a deep vein of the leg

Causes and risk factors

Most glucagonomas occur sporadically. This means they develop without a known inherited

cause. The exact reason that a pancreatic neuroendocrine cell becomes a tumor is usually not clear.

A small proportion are associated with multiple endocrine neoplasia type 1 , usually shortened to MEN1. MEN1 is an inherited condition caused by a change in the MEN1 gene. It increases the chance of tumors in several hormone-producing organs, including the pancreas, parathyroid glands, and pituitary gland.

A family history of MEN1 or several related hormone-producing tumors may lead to genetic counseling and testing. Having MEN1 does not mean that someone will definitely develop a glucagonoma.

Glucagonoma is not caused by eating sugar, having diabetes, or making everyday lifestyle choices. Diabetes may be an effect of the tumor rather than its cause.

Diagnosis

Diagnosis usually begins with a review of the rash, weight changes, blood sugar, digestive symptoms, and general health. The clinician may also ask about blood clots and any family history of endocrine tumors.

A fasting blood test measures glucagon. A clearly raised level can support the diagnosis when the symptoms and scans also fit. Glucagon can rise for other reasons, including severe illness and some liver, kidney, or pancreatic conditions, so the result is not interpreted by itself.

Other blood tests may check:

CT or MRI scans can show the pancreas, liver, and nearby structures. Endoscopic ultrasound may be used to examine the pancreas from inside the stomach or small bowel. This test uses a thin flexible tube with an ultrasound device at its tip.

Many glucagonomas have somatostatin receptors on their cells. Somatostatin receptor imaging uses a small amount of radioactive tracer to find tumors with these receptors. It can help show whether the tumor has spread and whether certain treatments may be suitable.

Tumor tissue is usually examined after a biopsy or operation. A pathologist looks at the cells and tests for neuroendocrine markers and glucagon. The tissue also provides a tumor grade, which describes how quickly the cells appear to be growing.

- Blood sugar and longer-term glucose control
- Blood count for anemia
- Liver and kidney function

- Protein, amino acid, vitamin, and mineral levels
- Other hormones if an inherited endocrine condition is possible

Treatment and management

Treatment is planned around the tumor's location, size, grade, spread, hormone production, and somatostatin receptor status. Your general health and treatment preferences also matter.

Surgery is the main treatment when the entire tumor can be removed safely. It may offer a cure when the glucagonoma is confined to the pancreas. The type of operation depends on where the tumor sits and whether nearby structures are involved.

If the tumor has spread, surgery may still be considered in selected situations. It may remove much of the tumor or treat areas in the liver, but it is not suitable for everyone.

Somatostatin analogues are medicines that copy the action of the natural hormone somatostatin. They can reduce glucagon release and may improve the rash, diarrhea, and other hormone-related symptoms. In some people, they also slow tumor growth.

Other treatments for advanced or growing tumors may include:

Supportive care is an important part of treatment. It may include treatment for high blood sugar, nutritional support, skin and wound care, pain relief, and management of anemia. Blood-thinning treatment may be needed if a clot develops or when the medical team judges the clotting risk to be high.

The skin rash often improves when excess glucagon is controlled. Nutritional support may also help because low levels of amino acids, zinc, and other nutrients can contribute to skin damage. Supplements should be guided by the results of an assessment rather than taken as a substitute for tumor treatment.

- Targeted medicines that interfere with tumor growth signals
- Peptide receptor radionuclide therapy, which carries radiation to tumors with somatostatin receptors
- Treatments directed at liver tumors, such as blocking their blood supply or using heat
- Chemotherapy for selected tumors
- Radiotherapy for particular areas causing symptoms

Complications

Glucagonoma can affect several parts of the body.

High blood sugar may develop into diabetes. If it becomes severe, dehydration and disturbances in body salts can occur.

Ongoing weight loss and altered protein use may cause muscle wasting and poor nutrition. Broken or blistered skin can be painful and may become infected.

Glucagonoma is also associated with an increased risk of blood clots. A clot in a deep leg vein is called deep vein thrombosis, or DVT. Part of a clot can travel to the lungs, causing a pulmonary embolism. This needs emergency treatment.

The tumor may grow into nearby tissue or spread to other organs, most often the liver. Spread can affect which treatments are possible, but it does not mean that symptom control or tumor-directed treatment must stop.

Prevention

Most glucagonomas cannot be prevented. There is no known diet, supplement, or lifestyle change that reliably stops a sporadic glucagonoma from developing.

People with MEN1 may be offered regular monitoring for endocrine tumors. Monitoring does not prevent a glucagonoma, but it may help find pancreatic neuroendocrine tumors earlier.

If MEN1 is suspected because of personal or family history, genetic counseling can explain testing and what a result could mean for relatives.

Living with it and what to expect

The course of glucagonoma differs from person to person. Important factors include whether the tumor can be completely removed, whether it has spread, its grade, and how it responds to treatment.

After successful surgery for a localized tumor, symptoms such as the rash and high blood sugar may improve. Follow-up is still needed because a tumor can sometimes return. Appointments may include scans, blood tests, and review of skin, weight, nutrition, and glucose levels.

Advanced glucagonoma is often managed as a long-term condition. Treatment may control hormone symptoms and slow growth for months or years, but the response cannot be predicted for one person. Treatment may change if scans show growth, symptoms return, or side effects become difficult.

Day-to-day care may involve protecting sore skin, planning meals around digestive symptoms, checking blood sugar if advised, and rebuilding strength after weight or muscle loss. A dietitian familiar with cancer or pancreatic neuroendocrine tumors may help with nutrition.

Tell the care team about changes between scheduled visits. A returning rash, weight loss, worsening diarrhea, or rising blood sugar may mean that symptoms need reassessment. These changes usually call for a routine or prompt appointment rather than emergency care, unless severe symptoms are present.

When to get medical help

Arrange a medical appointment if you have a persistent or recurring blistering rash, unexplained weight loss, ongoing diarrhea, or symptoms of high blood sugar. These symptoms have many possible causes, but they should be assessed.

If you have already been diagnosed, contact your care team promptly about a spreading or infected rash, continuing weight loss, uncontrolled diarrhea, worsening weakness, or significant changes in blood sugar.

Seek emergency help for:

Chest symptoms or sudden breathlessness can be signs of a pulmonary embolism. Do not wait for a routine appointment.

- Sudden shortness of breath, chest pain, fainting, or coughing up blood
- New swelling, warmth, or pain in one leg, especially with breathlessness
- Severe confusion, extreme drowsiness, or inability to keep fluids down
- Signs of severe dehydration, such as very little urine with marked weakness
- Fever with rapidly worsening redness, swelling, or pain around broken skin

Common questions

Is glucagonoma a type of pancreatic cancer?

It is a pancreatic neuroendocrine tumor and may be malignant, meaning it can invade nearby tissue or spread elsewhere. It is different from the more common pancreatic cancer that begins in the ducts of the pancreas. These tumor types behave differently and are treated differently.

Is the glucagonoma rash contagious?

No. Necrolytic migratory erythema is caused by changes inside the body, including excess glucagon and altered nutrition. It cannot spread to another person through touch.

Can glucagonoma be cured?

Sometimes. Complete surgical removal may cure a tumor that remains localized. If it has spread, treatment usually focuses on controlling hormone symptoms, slowing tumor growth, and protecting quality of life.

Does glucagonoma always cause diabetes?

No. Many people develop high blood sugar or diabetes, but not everyone does. Blood sugar may improve when glucagon production is controlled, although established diabetes does not always disappear.

Why does the rash move around?

The rash develops in cycles. One area may blister, peel, and heal while a new area appears nearby or elsewhere. This changing pattern is why it is described as "migratory." The precise biological reason is not fully understood.

Is glucagonoma inherited?

Most cases are not inherited. A minority occur as part of MEN1, which can run in families. Genetic counseling may be appropriate when MEN1-related tumors or features are present in you or close relatives.

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