Glucagonoma

Glucagonomas are a type of pancreatic neuroendocrine tumor that originate from the alpha cells of the pancreas. The symptoms of glucagonoma can be remembered by the 6 'D's: declining weight (weight loss), dermatitis (necrolytic migratory erythema), depression, diarrhea, deep vein thrombosis, and diabetes mellitus. Diagnosis begins with measuring glucagon levels which will be elevated. An abdominal CT can help localize the tumor. Management options include octreotide and surgical resection.



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Characteristics

Pancreatic Neuroendocine Tumor
Pancreas Neuron-indy-car Tumor-guy
A glucagonoma is a functional pancreatic neuroendocrine tumor (PanNET). Other PanNETs include insulinomas, gastrinomas, VIPomas, and somatostatinomas. Glucagonomas represent 5% of PanNETs. It can present as a benign adenoma, but 50% of patients can present with metastatic disease.

Originates from A Cells
Afro-guy and Cell
Glucagonoma originates from pancreatic alpha cells. Physiologically, glucagon promotes gluconeogenesis, glycogenolysis, ketogenesis, and lipolysis. It works primarily when the body has low blood glucose. Glucagon triggers the liver to begin amino acid oxidation and gluconeogenesis.

Clinical Features

Weight Loss
Skinny-with-baggy-pants
Weight loss is one of the most common clinical features seen in glucagonoma. Patients present with weight loss of around 5-15 kg in most cases.

Necrolytic Migratory Erythema
Necrosis-crow-light-bulb Migrating Earth-red
Necrolytic migratory erythema is a hallmark of glucagonoma. It is a rash which is pruritic and painful. It mostly occurs on the face, extremities, and groin. It presents with central clearing surrounded by small erythematous papules that fuse to build large and indurated plaques. It is typically bronze or brown colored.

Depression
Depressed-emo
Depression can be present in up to 50% of patients with glucagonoma. The pathophysiologic mechanism is not well understood. It remains to be elucidated whether it occurs due to high glucagon or a result of chronic wasting disease. It is also thought to be associated with chronic dermatosis. Other neuropsychiatric conditions may occur, including agitation, dementia, psychosis, proximal muscle weakness, ataxia, paranoid delusions, and hyperreflexia.

Diarrhea
Toilet
Diarrhea can be present in 30% of patients with glucagonoma. It is associated with multiple causes, such as increased gut motility, malabsorption, and bacterial overgrowth.

Deep Vein Thrombosis
Deep V-neck Trombone
Deep Vein Thrombosis (DVT) can be present in around 40% of cases. The mechanism of this is not precisely understood, but it is thought to occur due to the increased production of factor X by pancreatic alpha cells.

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Diabetes Mellitus

Dyed-bead-pancreas

The excess of glucagon leads to glycogenolysis and gluconeogenesis in the liver, resulting in increased glucose in the blood (hyperglycemia). This can contribute to diabetes mellitus or glucose intolerance. It is often present in 80-90% of glucagonoma cases. Symptoms of glucagonoma can be described with 6Ds: Dermatitis (necrolytic migratory erythema), Diabetes (hyperglycemia), DVT, Depression, Diarrhea, and Declining weight.

Diagnosis

Increased Glucagon

Up-arrow Glue-King-Kong

Glucagonoma causes increased production of glucagon. Most glucagonoma cases can present with levels of glucagon 2-3 times higher than the normal range.

Abdominal CT

Cat-scanner of Abdomen

Abdominal CT can help visualize the precise site of glucagonoma. It is used as an initial diagnostic study due to its non-invasiveness. This diagnostic study is superior to ultrasonography due to its ability to visualize the pancreas' body and tail much better. Glucagonoma is often found (90% cases) in the body and tail of the pancreas.

Management

Octreotide

Octo-tree-ride

Somatostatin analogs, such as octreotide, are glucagon secretion inhibitors. These effectively reduce secretion of glucagon and control metastatic development. These also help in improving skin rashes and reversing the catabolic state from excess glucagon.

Surgical Resection

Surgeon

Surgical resection is the only curative treatment. It is recommended and effective in patients with localized cases. Some cases are inoperable due to advanced staging and/or metastases.

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