Cancer Association of South Africa (CANSA)

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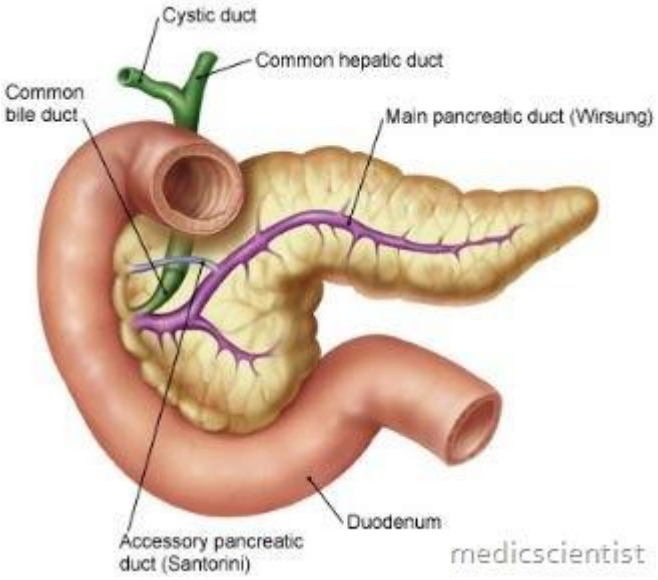
Fact Sheet
on
Somatostatinoma

Introduction

Somatostatinomas are rare tumours that can develop in the pancreas or small bowel. Pancreatic tumours most commonly develop in the head of the pancreas.

[Picture Credit: Pancreas]



Somatostatinomas develop in hormone producing cells in the pancreas called islet cells. There are different types of islet cells, each producing different hormones. Delta islet cells make the hormone somatostatin. Somatostatin cuts off production of other hormones by the pancreas and controls how the gut works. Somatostatinomas produce extra somatostatin, which eventually leads to symptoms.

Elangovan, A. & Zulfiqar, H. 2020.

"Somatostatinoma is a rare neuroendocrine tumor (NET) with an incidence of 1 in 40 million individuals and accounts for less than 5% of pancreatic NETs (pNETs). The tumor was first described in 1977 in a 46-year-old woman in the head of the pancreas. The tumor originates from the delta cells of the pancreas and predominantly contains somatostatin with trace quantities of other pancreatic hormones such as insulin, glucagon, gastrin, and vasoactive intestinal polypeptide. It remains localized to the pancreas in 56 to 70% of the cases, out of which 36% occur in the head, 14% in the pancreatic body, and 32% in the tail. Other common sites include duodenum (19%), ampulla of Vater (3%), and small bowel (3%). Reports exist of rare instances of extra-gastrointestinal primaries in the lungs, kidneys, and thyroid."

Incidence of Somatostatinomas

The National Cancer Registry of 2017 does not furnish any information regarding Stomatostatinoma.

Signs and Symptoms of Somatostatinoma

The following are typical signs and symptoms for Somatostatinoma:

- Excess somatostatin
- Mild hyperglycaemia
- Presence of gallstones
- Malabsorption problems
- Presence of excessive fat in faeces
- Weight loss
- Diagnosis of diabetes
- Cholestatic jaundice
- Presence of intestinal tumour
- Presence of a pancreatic tumour
- Reduced hydrochloric acid in digestive system
- Reduced gastric release

Martin, S., Fica, S., Parfeni, O., Popa, L., Manuc, T., Rizea, O., Lupescu, I., Gherghe, M., Becheanu, G. & Croitoru, A. 2020.

"Somatostatinomas are rare neuroendocrine tumors (NET) that arise in the gastrointestinal (GI) tract. Because of their insidious growth, they are usually asymptomatic until late stages, presenting as malignant disease. We report the case of a 50-year-old woman who presented with epigastric abdominal pain, diarrhea and significant weight loss in the last two years. On clinical examination the patient met the criteria for neurofibromatosis type 1 (NF1). Abdominal CT and MRI revealed an infiltrative duodenal mass, with pancreatic invasion, locoregional enlarged lymph nodes and disseminated hepatic nodules. Microscopy and immunohistochemistry uncovered a neuroendocrine tumor, staining positive for chromogranin A (CgA), synaptophysin and somatostatin, with a Ki67 = 1%. Somatostatin receptors (SSTRs) type 2 were negative and SSTRs type 5 were positive in less than 50% of tumoral cells. Our patient was classified as a T3N1M1 stage IV metastatic duodenal grade 1 somatostatinoma and treatment with somatostatin analogues and chemotherapy with capecitabine and temozolomide was started, with so far abdominal imaging follow-up showing stable disease. When a patient is diagnosed with a rare NET, such as a somatostatinoma, it is of utmost importance to determine if it is a sporadic tumor or just a feature of a genetic disorder."

Treatment of Somatostatinoma

Since somatostatinomas have a high malignant potential, aggressive intervention is warranted. Surgical excision usually offers the only opportunity for long-term cure.

About Clinical Trials

Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The South African National Clinical Trials Register provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

Medical Disclaimer

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