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Zollinger–Ellison Syndrome: A Critic Review

***Ananda Kudari*[1*], *Chetan Ramesh Sangati*[2]**
[1]*Associate Professor,* [2]*M.Sc. Nursing Scholar, Department of Medical Surgical Nursing, SDM Institute of Nursing Sciences, Sattur, Dharwad, Karnataka, India*
Email: *anandakudari@yahoo.co.in*
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Abstract

Zollinger-Ellison syndrome is a rare condition of gastrointestinal system, in which one or more tumors in the pancreas or the upper part of the small intestine (duodenum). These kinds of tumors are called gastrinomas, which secrete large amounts of the hormone called gastrin, which result in stomach to produce too much Hydrochloric acid (HCL). The excess HCL then leads to peptic ulcers (PU), as well as diarrhoea and other symptoms. Zollinger-Ellison syndrome (ZES) which affect all age groups as well both the genders. The disease may occur at any time in life, but it is more common in age group of 20 and 50 years. The treatment for the Zollinger Ellison syndrome is, Medications (H_2 receptor blockers) to reduce stomach acid and heal the ulcers of the stomach, duodenum, and surrounding area of the gastrointestinal system.

Keywords: *Gastrinomas, hydrochloric acid, H_2 receptor blockers, peptic ulcers, small intestine*

INTRODUCTION

When there is a tumor so called gastrinomas present in the pancreas and duodenum which secrets the hormone called as gastrin results in more production of HCL in the stomach this condition is called as zollinger-ellison syndrome (ZES) [1]. The acid plays a key role in digestion of proteins [2]. Gastrinomas occur as single tumors or several tumors. These tumors often spread to the liver and nearby lymph nodes which result in Zollinger Ellison syndrome. [3]. Experts do not know the exact cause of Zollinger-Ellison syndrome. About 20 to 35 percent of gastrinomas are witnessed by an inherited genetic disorder so called multiple endocrine neoplasia type 1 (MEN1).

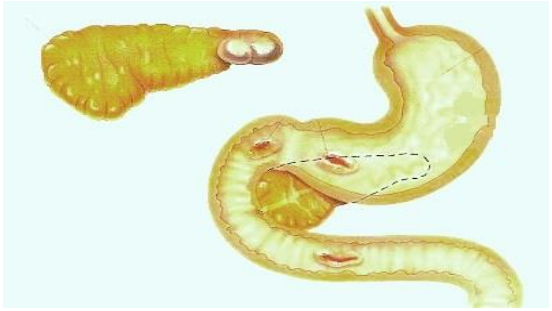

Figure 1: *zollinger-Ellison Syndrome*

DEFINITIONS

Zollinger-Ellison syndrome is a rare disorder characterized by the development of a tumor called a Gastrinoma found in the pancreas and/or duodenum which secrete excessive levels of gastrin, a hormone that stimulates production of acid by the stomach [4].

Zollinger-Ellison syndrome (ZES) is characterized by the development of a tumor (Gastrinoma) or tumors that secrete excessive levels of gastrin, a hormone that stimulates production of acid by the stomach [5].

INCIDENCES

Zollinger-Ellison syndrome is rare and only occurs in about one in every 1 million people [6]. Although anyone can get Zollinger-Ellison syndrome, the disease is more common among men 30 to 50 years old. A child who has a parent with MEN1 is also at increased risk for Zollinger-Ellison syndrome [7]. Only 0.1% to 1% of patients with peptic ulcer disease (PUD) have zollinger-ellison syndrome (ZES) [8,

9]. About 25% to 30% of patients with ZES have multiple endocrine neoplasia type 1 (MEN1) which is a group of hyperplasia and/or tumors of pituitary, parathyroid and pancreatic islet cells. Therefore, patients with ZES from MEN1 could cause hypersecretion of gastric acid from hypercalcemia [10, 11] and gastrinomas are found in every 0.1 to 3 persons per million [12].

CAUSES/ETIOLOGY

- Excess secretion of Hydrochloric acid (HCL)
- Neuroendocrine tumor(NET)
- Gastrinoma of the duodenal less frequently pancreatic in location
- Autosomal-dominant syndrome
- Hyperparathyroidism
- Pituitary adenomas
- Separate distinct syndrome
- Cushing's syndrome
- Carcinoid syndrome
- Insulinoma and parathyroid hormone-related protein secreting tumors
- Insulinomas
- Physiological-hypergastrinemia
- Chronic atrophic gastritis/pernicious anemia (hypochlorhydria),
  Chronic helicobacter infection of the oxyntic mucosa (hypochlorhydria). [13]

PATHOPHYSIOLOGY

Due to various etiological factors

⬇

Excessive gastric acid secretion

⬇

Result of gastin- producing gastrinomas from delta cells at found in the pancreas

⬇

Gastrin directly stimulates pancrietal cells secretion and also causes expansion if the mass of parietal cells

⬇

Increase in basal acid output and maximal acid output

⬇

Result in gastro esophageal reflex disease and damage to the mucosal lining of the GI tract causing peptic ulcer

⬇

Which contributes to diarrhea, steatorrhea, Malabsorption or lipid –soluble nutrients [14]

CLINICAL SYMPTOMS

- Peptic ulcer
- Increase in hydrochloric acid
- Malabsorption of nutrient components
- High serum gastin concentration
- Inhibit sodium and water reabsorption
- Severe diarrhea
- Abdominal pain
- Weight loss [15]
- Nausea
- Wheezing
- Vomiting blood
- Malnourishment
- Loss of appetite
- Steatorrhea [16]

Diagnostic Evaluation

The diagnosis of Zollinger–Ellison syndrome is paramount, the diagnosis is made through several laboratory tests and imaging studies [17]

Medical history: Doctor will ask about signs and symptoms and review medical history

Blood Tests: A sample of blood is analyzed to see whether have elevated gastrin levels. If there is an increased level of gastrin in the blood stream gives the clue for presence of tumors in the pancreas or duodenum.

Upper gastrointestinal endoscopy: Upper GI endoscope is very useful to look any ulcers in the stomach and duodenum. In this procedure, doctor may collect the tissue sample (biopsy) from stomach or duodenum to check the presence of gastrin-producing tumors.

Endoscopic ultrasound: In this procedure, doctor examines client stomach, duodenum and pancreas with an endoscope fitted with an ultrasound probe. By this procedure doctors look for any tumors present in it.

Imaging tests: Doctor may use imaging techniques such as a nuclear scan called somatostatin receptor scintigraphy. This test uses radioactive tracers to help locate tumors. Other helpful imaging tests include ultrasound, computerized tomography (CT) and magnetic resonance imaging (MRI) [18].

Predictive testing: For at-risk, asymptomatic, adult family members requires first finding the disease-causing mutation in the affected family member.

The Genetic Testing Registry: (GTR) provides information about the genetic tests for multiple endocrine neoplasia type 1 [19].

Complications

- Gastric outlet obstruction
- Bleeding and sever pain
- Gastric perforation [20]

Medical Management

Proton pump inhibitors: Proton pump inhibitors are the drugs which will decrease the secretion of gastric acid E.g. Omeprazole

H_2-receptor antagonists: Are the drugs which slow down the secretion of gastric acid E.g. Famotidine and Ranitidine [21].

Synthetic porcine secretin: For use in secretin stimulation testing for: Stimulation of pancreatic secretions to facilitate the identification of the ampulla of Vater and accessory papilla during endoscopic retrograde cholangio-pancreatography (ERCP) [22].

Chemotherapy: Chemotherapeutic agents are used to treat gastrinomas in the stomach and in duodenum. Commonly used single agent chemotherapeutic medication includes 5-Fluorouracil, Cisplatin, and Mitomycin.

Endoscopy: To determine the degree of ulcer and tissue specimen can be obtained for identification of H.pylori and to rule out gastric cancer.

- Anti-angiogenesis agent such as bevacizumab inbhit vascular endothelial growth factor activity, with the goal of regressing tumors by starvation.
- Radiation therapy is mainly used for palliation in patient with obsatction

Non-Invasive Test

- Serum and whole body test in particularly ,immunoglobulin G(Ig G)
- Urea breath test can determine the presence of active infection [18].

Surgical Treatment

The chief goal of surgery is to cure or control the tumor, and prevent metastases [23].

- **Laparotomy**, the stomach, small bowel, liver, hepatic portal area, gastro hepatic mesentery, greater omentum, and mesentery of the large and small intestines were thoroughly explored for primary and metastatic islet cell tumors. The peritoneum at the inferior and superior borders of the pancreas was incised to allow palpation of the body and tail of the pancreas. Any enlarged lymph nodes were excised. A proximal gastric vagotomy without a drainage procedure was performed with truncal vagotomy and

pyloroplasty because obesity made performing a proximal gastric vagotomy difficult [24].

- **Total gastrectomy** may be performed for a resectable cancer in the midportion or body of the stomach. The entire stomach is removed along with the duodenum, the lower portion of the esophagus, supporting mesentery and lymph nodes.
- **Essophagojunostomy** reconstruction of the GI tract is performed by anastosting the end of the jejunum to the end of the esophagus.
- **Radical subtotal gastrectomy** is performed for a resectable tumor in the middle and distal portion for a resectable tumor in the middle and distal portions of the stomch.
- **Billroth I**: involves a limited resction and offers a lower cure rate than the Billroth- II.
- **Billroth-II**: is a wider resction that invalves removing approximately 75% of the stomch and decrease the possibility of lymph node spread or metastic recurrence.

Nursing Management

Nursing Diagnosis

- Anxiety related to the disease and anticipated treatment.
- Imbalanced nutrition: less than body requirements related to early satiety or anorexia.
- Acute pain related to tumors mass.

Intervention

Reducing Anxiety

- A relaxed, nonthreatening atmosphere is provided so the patient can express fears, concerns and possibly anger about the diagnosis and prognosis.
- The nurse encourage the family to support patient.
- Offering reassurance and supporting positive coping measures.
- The nurse advices the patient about any procedure and treatment so that the patient knows what to expect.

Promoting Optimal Nutrition

- Encourage him to eat small, frequent portion of nonirritating foods to reduces gastric irritation.
- Food supplement should be in high in calorie.
- If the patient is unable to eat adequately prior to surgery to meet nutritional requirement, parenteral nutrition may be necessary.
- The nurses monitor the IV therapy and nutritional status and record intake, output and daily weight to ensure the patient is maintaining or gaining weight.
- The nurses assess for the dehydration, decrease urine output and reviews the result of daily laboratory studies to note any metabolic abnormalities

Relieving Pain

- The nurse administers analgesic agents as prescribed.
- A continuous IV infusion of an opioid or PCA pump set to infuse an opioid may be necessary to mitigate post-operative pain.
- The nurses routinely assess the frequency, intensify and duration of the pain to determine the effectiveness of the analgesic agent [18].

CONCLUSION

Zollinger-ellison syndrome is rare disorder of the gastrointestinal tract which result increased level of gastric acid in the stomach and causes peptic ulcer disease. This increased level of gastric acid is causes the number of symptoms of the disease. Certain types of medication can control the secretion of gastric acid and also surgical procedures also can be do for the Zollinger–Ellison syndrome, early

detection of the disease can be prevent the complication and save the client life.

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