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**Case Report**

# ZOLLINGER ELLISON SYNDROME IN A 12 YEAR OLD CHILD - A CASE REPORT

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**ABSTRACT**

The syndrome described by Zollinger and Ellison (ZES) in 1955 is a rare clinical entity which is even rarer in children. This report describes a 12 year old boy who presented with refractory peptic ulcer disease which was finally diagnosed to be due to a gastrinoma and was successfully treated.



## INTRODUCTION

The overall incidence of ZES is 0.1-3 per million and among these patients, only 2% belong to the pediatric population.[1] Most of these tumours are located in the duodenum but they are typically found in the well described Passaro's gastrinoma triangle.[2] We report a case of this rare entity which requires a high index of suspicion in cases of refractory peptic ulcer disease.

## CASE REPORT

A 12-year-old boy weighing 35 kg presented with history of severe epigastric pain, vomitting, and occasional loose stools for 1year with a weight loss of about 10kg. Physical examination showed epigastric tenderness but no other abnormalities. Acute pancreatitis was ruled out. With a provisional diagnosis of Peptic ulcer disease, H2 receptor blocking agents and proton pump inhibitors (PPI) were started. However, the patient was refractory to this treatment. Upper GI endoscopy showed severe esophagitis with gastric mucosal thickening with multiple mucosal ulcerations in duodenum. On further investigation, ultrasound of the abdomen showed a heterogenous, hyperechoic lesion in the head of pancreas with severe degree of gastric mucosal thickening. Further confirmation done with contrast enhanced CT abdomen showed 3.8X2.8 cm well defined moderately enhancing soft tissue lesion, in Passaro's triangle arising from head of the pancreas and anterolaterally reaching the lesser curvature of stomach. Laterally and posteriorly, the lesion was abutting the left lobe and caudate lobe of liver respectively. The gastric wall was significantly thickened. To confirm the diagnosis of a secreting tumour, fasting serum gastrin levels were sent which turned out to be high - 940 pg/ml. (N- 15-113pg/ml). Plasma chromogranin A levels were also high - 1310ng/ml (N<108ng/ml) suggestive of a neuroendocrine tumor. The tumour was located in the lesser sac at the superior border of head of pancreas supplied by a branch of superior pancreaticoduodenal artery and was excised in toto. It measured about 4X4X4 cm in size. There were no enlarged lymph nodes or peritoneal deposits. The post operative course was uneventful. Histolopathology confirmed the diagnosis of gastrinoma. Preoperative ultrasonography had shown gastric mucosal thickening of 2.5cm, which reduced significantly to 4mm postoperatively. The patient is pain-free without any medications at 2 months follow up and is under surveillance.

## DISCUSSION

Gastrinomas are the second most common pancreatic neuroendocrine tumours after insulinomas.[3] In the pediatric age group, they have an infrequent prevalence.[1] The youngest reported case was just under 2 years of age.[4]

The definitive diagnosis of gastrinoma requires demonstration of high levels of gastrin in fasting serum (>1000pg/ml). Our patient had similar elevated levels i.e. 940pg/ml. Another

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important diagnostic clue was the presence of thickening of the gastric mucosa seen on simple ultrasound imaging associated with a mass. This according to us provides corroborative evidence that the mass is secreting gastrin.

Pancreatic gastrinomas are usually large (mean 3.8cm) unlike duodenal tumours (mean 0.93cm)[5] as was the case with our patient but giant gastrinomas have also been reported[6,7]. These tumours are classically described in the Passaro's gastrinoma triangle (85-95%) but stray cases of other locations like lymph nodes[8], liver, bile duct and ovary[9] have been seen.

The use of gastric mucosal thickness on ultrasound as an indicator of hypertrophy of the parietal acid secreting cells of the stomach due to the effect of a gastrin secreting tumour has not been reported in literature. We think that this can be a very useful modality to increase the suspicion of a possible neuro endocrine tumour. It is especially relevant in the cases of small tumours (<1cm) which cannot be picked up on conventional imaging modalities like CT scan and Ultrasound.

These tumours can be benign or malignant. In children, most recent studies report a malignancy rate of close to 30 %[1]. The poor prognostic factors include liver and lymph node metastasis, large tumour size(>3cm), short history of disease, inadequate control of gastric hypersecretion and various histopathological and flow cyometric features among others.[5] Hence regular follow up and surveillance of these patients is mandatory.

## CONCLUSION

Gastrinomas should be suspected in children of refractory peptic ulcer disease. Gastric mucosal thickness on ultrasound or CT scan can serve as a good corroborative diagnostic as well as follow up modality in these patients.

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