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# Gastric Neoplasm – A Diagnostic Puzzle

**Madhumita Mukhopadhyay[1], Binata Bandopadhyay[2], Chhanda Das[3], Ananya Ghatak[4], Subhamoy Saha[5], Rituparna Kayal[6]**

[1]Professor, Department of Pathology IPGME&R, Kolkata, West Bengal, India

[2,4,5,6]PGT, Dept. of Pathology, IPGME&R, Kolkata, West Bengal, India.

[3]Associate Professor, Department of Pathology, Burdwan Medical College, Burdwan, West Bengal, India

**ABSTRACT**

**Introduction:** Paragangliomas are non-epithelial neuroendocrine tumors originating from neural crest derived paraganglion cells situated in the region of the autonomic nervous system ganglia and accompanying nerves. Very few cases of primary gastric paraganglioma have been reported in the literature till date. We report a rare case of primary gastric paraganglioma.

**Case Report:** A 38 year old woman presented with history of melena and dyspepsia. Physical examination revealed mild tenderness in the right hypochondrium, and she was diagnosed to have gall stone disease. However, upper gastrointestinal endoscopy and contrast enhanced computed tomography whole abdomen revealed presence of a small elevated intraluminal lesion at the body of stomach along greater curvature and clinically diagnosed as gastrointestinal stromal tumor(GIST). Endoscopic biopsy report revealed a diagnosis of submucosal Brunner's gland hamartoma. Laparoscopic cholecystectomy and laparoscopy assisted wedge resection of the stomach mass was done and the specimens were sent for histopathological examination. The stomach mass was diagnosed as paraganglioma which was confirmed by immunohistochemistry.

**Conclusion:** Primary gastric paraganglioma is a very rare tumor. They are more difficult to diagnose if they are non-functional. Complete surgical excision is the first line treatment that can be performed with open surgical resection or laparoscopic technique.

**KEYWORDS:** gastrintestinal stroma tumor, incidentaloma, paraganglioma.

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## INTRODUCTION

Paragangliomas are rare neuroendocrine neoplasms that arises from embryonic neural crest [1-3]. They can be located anywhere in relation to the autonomic nervous system (ANS). Paragangliomas are usually sporadic but can be associated with genetic syndromes. They are of two types- sympathetic ( arising from sympathetic ganglia distributed along prevertebral, paravertebral and paraaortic sympathetic chains and sympathetic nerves innervating the retroperitoneum, thorax and pelvis), parasympathetic ( arising from the head neck paraganglia which includes the carotid body, jugulotympanicum, vagus nerve and larynx).Extra adrenal paragangliomas represent only 5 to 10%, located from the upper cervical region to the pelvis, in association with ANS[2]. Visceral paragangliomas are rare, can develop in larynx, interatrial septum of the heart, bladder, liver, gallbladder[4]. Gastric cases are very rare.

Paragangliomas may be asymptomatic and diagnosed on routine investigations and that's why they are termed as "incidentalomas"; clinical features depend on mass effect, catecholamine secretion or malignant behaviour [3,5.6].

As per our knowledge, very few cases of gastric paragangliomas have been reported [2-4, 7-11]. Only 10 cases of gastric paraganglioma have been reported.[11].

Here we are presenting a very rare case of primary gastric paraganglioma in a 38 years old female which was initially misdiagnosed as submucosal Brunner's gland hamartoma on histopathological examination of upper gastrointestinal endoscopic biopsy and clinically mistaken as gastrointestinal stromal tumor(GIST).

## CASE REPORT

A 38 years old female presented in the surgical out patient department (OPD) with a three months history of melena and

dyspepsia since four to five months. The patient was apparently well three months back when she developed acute onset pain in the abdomen over the epigastric region, which was dull aching in nature and insidious in onset, and it gradually progressed to involve the entire abdomen. There was no history of fever or loss of appetite, weight loss, vomiting, jaundice or hematemesis.

She complained of melena for one week three months ago. She is a known case of diabetes mellitus since last one year. Past surgical history revealed she underwent three cesarean sections.

During her first visit to the surgical OPD, her general condition was good. She had mild tenderness in the right hypochondrium. Baseline blood investigations and ultrasonography whole abdomen( USG W/A) along with upper gastrointestinal endoscopy (UGIE) were advised.

Blood investigations revealed Hemoglobin- 10.4g% , rest were under normal limits. USG W/A revealed a solitary calculus of 2.32cm in the fundus of gallbladder. UGIE revealed a 2cm sized overelevated submucosal lesion with a central ulcer seen at the midbody of the stomach along the greater curvature ( Fig 1. ). Endoscopic biopsy of the stomach lesion done. Then contrast enhanced computed tomograpgy whole abdomen (CECT W/A) were then advised. Endoscopic biopsy report revealed the diagnosis of submucosal Brunner's gland hamartoma at the body of the stomach along the greater curvature. CECT W/A revealed partially contracted gallbladder with slightly thickened wall, hepatomegaly and a small luminal polypoidal lesion (2.6cm x 1.9cm) on the greater curvature of the body of stomach ( Fig. 2 ). No metastasis or tumor in any other site noted.

She was planned for laparoscopic cholecystectomy with wedge resction of the mass in the stomach. Laparoscopic cholecystectomy was done. However, the mass in the stomach could not be delineated via laparoscopy hence laparoscopy was converted to Laparoscopy assisted surgery and the mass was found in the greater curvature of the stomach. Wedge resection of the stomach mass was then done. Both the specimens of resected gallbladder and the stomach mass were then sent to the department of Pathology for histopathlogical examination (HPE).

The specimen of stomach mass was labelled as "Gastric GIST obtained by wedge resection". On gross examination it was found to be a globular tissue piece which measured 3cm X 2cm X 1cm. The cut section was whitish homogenous ( Fig. 3 ).

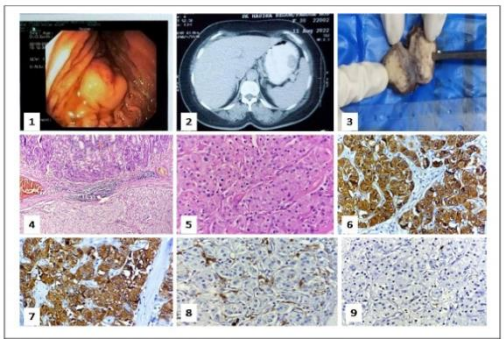

**Fig 1**. Upper GI Endoscopy showing a lesion along the greater curvature of stomach.
**Fig 2**. CECT abdomen showing a small intraluminal polypoidal lesion on the greater curvature of the body of stomach.
**Fig 3.** Gross picture showing a globular soft tissue mass measuring 3x1x1cm with whitish homogenous cut surface.
**Fig 4.** HPE on 400X showing histological features of Gastric paraganglioma.
**Fig 5.** HPE on 100X showing histological features of Gastric paraganglioma.
**Fig6.** Showing diffuse cytoplasmic positivity for Synaptophysin in Gastric paraganglioma(400X).
**Fig 7.** Showing diffuse cytoplasmic positivity for Chromogranin in Gastric paraganglioma(400X).
**Fig 8.** Sustentacular cells showing S100 positivity in Gastric paraganglioma(400X).
**Fig9.** Showing CD117 negativity in Gastric paraganglioma(400X).

Later HPE revealed a circumscribed cellular tumor with a zellballen pattern, composed of two cell types-Chief cells with abundant eosinophilic granular cytoplasm, bland looking round to oval nuclei with salt and pepper chromatin arranged in nests, Sustentacular cells which are slender, spindle shaped cells located peripherally around the chief cell nests. Mitotic count was low. There was no significant nuclear or cytological atypia. Necrosis was absent ( Fig. 4& 5 ). The histological features were suggestive of Primary Gastric Paraganglioma.

The diagnosis was confirmed by immunohistochemistry (IHC) which revealed that the chief cells in nests were positive for Synaptophysin and Chromogranin A ( Fig.6 & Fig. 7 ) the sustentacular cells around the chief cell nests were positive for S100 ( Fig. 8 )

**DISCUSSION**

Paragangliomas are neural crest derived neuroendocrine neoplasms, which are either functional or non functional. They account for 10% of catecholamine secreting tumors with an annual incidence of 1/100000[3,5]. Males are slightly more commonly affected than females[11]. Depending on their location into the ANS, paragangliomas may be classified into two groups- sympathetic and parasympathetic. Sympathetic paragangliomas usually secretes catecholamines while majority of the parasympathetic paragangliomas are non functional. Norepinephrine secreting paragangliomas clinically presents with arterial hypertension, sweating ,palpitation, headache while those not secreting catecholamines presents with vague symptoms like weight loss, anxiety, facial pallor, polyuria/polydipsia, stroke, hyperglycemia, secondary erythrocytosis,cardiomyopathy[3].

However, Paragangliomas may be asymptomatic, as in our case and are incidentally detected, those tumors are called "incidentalomas" [3].

In our case, a small mass with central ulcer was found in the mid portion of the body of stomach along the greater curvature. It was non functional and misdiagnosed as submucosal Brunner's gland hamartoma on UGI endoscopic biopsy. Primary gastric paraganglioma is a very rare tumor and non functioning ones are more difficult to diagnose clinically or even by imaging modalities. The pre operative differential diagnoses includes GIST, schwannoma, even in our case the post operative specimen was sent as "Gastric GIST". R.Bura et al. Also found a 2 cm. In the greater curvature in the body of stomach near greater curvature. It was clinically diagnosed as acute appendicitis .After histopathological examination and IHC it was diagnosed as gastric paraganglioma.

Computed tomography (CT) and Magnetic resonance imaging (MRI) are useful in determining various tumor features like the site, the number, the malignant behaviour or association with any other neoplasm[3,11]. In our case CECT W/A revealed the presence of a small intraluminal polypoidal lesion (2.6cm x 1.9cm) on the greater curvature of the body of the stomach, no signs of metastasis or any tumor in any other site noted.

Diagnosis of paraganglioma is done by HPE of the resected specimen and confirmed by IHC. The most characteristic feature is the presence of nests of uniform looking cells with bland round to oval nuclei and salt and pepper chromatin and abundant eosinophilic granular cytoplasm surrounded by spindle shaped sustentacular cells. IHC shows diffuse cytoplasmic positivity for synaptophysin and chromogranin A with S100 positive sustentacular cells. In our patient both the histological and immunohistochemical features were consistent with paraganglioma. CD117 was negative which ruled out GIST.

The biological behaviour of paragangliomas can not be predicted by microscopical appearance of the cells as sometimes atypia is also present in benign lesions.[2]. All tumor irrespective of whether they are functional or not, have metastatic potential. So, paragangliomas are often referred to as metastatic or non metastatic instead of benign or malignant.

The treatment depends on several factors-location, size, extension, symptoms, malignancy and status of somatostatin receptors[3]. The first line treatment for resectable tumors is complete surgical resection of the tumor either with open surgery or laparoscopy. However, several studies have revealed laparoscopic treatment for extra-adrenal intra abdominal paragangliomas when the tumor size is less than equal to 4cm[3]. In our case the patient underwent laparoscopic cholecystectomy with lap assisted wedge resection of the stomach mass.

## CONCLUSION

Primary gastric paraganglioma is a very rare tumor and more difficult to diagnose if they are non functional. The main differential diagnoses includes gastrointstinl stromal tumor and schwannoma. Complete surgical excision is the first line treatment and can be done by open surgical resection or laparoscopy.

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