# Solitary Gastric Extramedullary Plasmacytoma Present as Subepithelial Tumor: A Case Report

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### ABSTRACT

Extramedullary plasmacytoma (EMP) is a plasma cell neoplasm, without systemic involvement. EMP involving gastrointestinal is rare. We report a case of a 64-year-old women diagnosed with a solitary extramedullary plasmacytoma of stomach. In our case, the computed tomography (CT) finding of EMP in stomach is like subepithelial tumor. Not yet reported this feature of EMP associated with the stomach.

Keywords: Solitary extramedullary plasmacytoma, stomach, Computed tomography

#### Introduction

Extramedullary plasmacytoma (EMP) is a plasma cell neoplasm that present as a solitary lesion in the soft tissue, without systemic involvement. EMPs are rare tumors that from 3-4% of all plasma cell neoplasm (Nolan *et al.*, 2005). EMP is mostly observed in the upper aerodigestive passage. Solitary extramedullary plasmacytoma is an extreamely rare form of EMP in stomach, with only approximately 100 cases since 1928 (Alexiou *et al.*, 1999). Therefore, we report a rare case of solitary extramedullary plasmacytoma presenting gastric subepithelial tumor on abdomen computed tomography (CT).

#### **Case Report**

A 63-year-old women with hepatitis B and liver cirrhosis was refered to our hospital due to gastric tumor identified during screening esophagogastroduodenoscopic (EGD) and endoscopic ultrasound (EUS) examination, which revealed a smoothly elevated lesion, 1cm in diameter occupying the posterior wall of stomach. Biopsy showed chronic gastritis. After 19 months, EGD and EUS was done. Previous smoothly elevated lesion occupying the posterior wall of gastric lower body grew to 1.5cm in size (Fig. 1). No ulcerative change was observed on the mucosal surface, the patient was asymptomatic and laboratory study finding was all within normal limits. Liver CT scan was performed for LC evaluation. CT revealed

1.5cm sized well-defined soft tissue density mass at the gastric lower body and intraluminal location on non-enhanced image. This mass showed early enhancement on early phase images and progressive enhancement on portal venous phase images. On delayed phase images, this mass showed homogenous enhancement and was more enhancement than the gastric wall. No evidence of perigastric infilteration or lymphadenopathy was seen (Fig. 2). Considering the EUS and CT findings, subepithelial tumor was suggested but ectopic pancreas cannot be excluded.



**Figure 1:** Endoscopy shows a 1.5cm protuberant lesion on the posterior wall of gastric lower body (a). Endoscopic ultrasonography shows a homogenous hypoechoic smooth margined mass deriving form submucosal layer (b).



**Figure 2:** The unenhanced axial image (a) shows a well-defined iso density mass (white arrow) in lesser curvature side of stomach lower body. After contrast administration, the mass is showed early enhancement compared with normal stomach wall on arterial phase (b) On portal phase, the mass is showed progressive enhancement (c) and homogenous enhancement on delayed phase axial and coronal images (d,e).

Laparoscopic wedge resection was performed. On pathology evaluation, a 1.5 cm-sized submucosal mass consisted of uniform population of discohesive cells with abundant eosinophilic cytoplasm and eccentric nucleus typical charateristics of plasma cells. These cells infiltrated along the gatric submucosa

and remaining mucosa associated lymphoid tissue (MALT) (Fig. 3a). These findings led to the necessity to rule out EMP and MALT lymphoma with plasma cell differentiation. On the high power view, marked atypical nuclei were identified with pleomorphism suggesting neoplastic condition (Fig. 3b). diagnosis was a extramedullary plasmacytoma. The mass was atypical infilteration of plasma cell in submucosa layer (Fig. 3). On the immunohistochemical study, these atypical cells were diffusely positive for CD138 indicating plasma cell and strong Kappa light chain positivity but Lambda light chain negativity of this plasma cells led to the conclusion of plasmacytoma (Fig. 4). To confirm the tumor monoclonality again, Immunoglobulin heavy chain gene rearrangement test was performed and monoclonality was identified in molecular level.



**Figure 3:** Atypical infiltration of plasma cells in submucosa presenting submucosal tumor. (a) Numerous aggregation of discohesive cells showing abundant eosinophilic cytoplasm and eccentric nuclei around the remnant lymphoid follicle (asterix) (H&E, x40; inlet x1). (b) The uneven sized and shaped dysplastic nuclei with prominent nucleoli indicating tumourous condition (H&E, x400).



Figure 4: The immunohistochemical stain results supporting plasama cell neoplasm. (a) The diffuse and strong CD138 positivity of tumour cells indicating plasma cell. (b) The pridomenent kappa light chain expression of plasma cells indicating monoclonality of the tumour cells.

Positron emission tomography/computed tomography (PET-CT) showed no other lesion. And serum and urine protein electrophoresis yielded normal result. Bone marrow examination showed no abnormalities.

Given the laboratory results listed above, the lesions were diagnosed as solitary extramedullary pleasmacytoma of the stomach.

## Discussion

Plasma cell neoplasms are classically categorized into four groups; multiple myeloma (MM), plasma cell leukemia, solitary plasmacytomas of the bone (SPB) and extramedullary plasmacystoma (EMP) (Alexiou *et al.*, 1999). The diagnosis of EMP requires demonstration of a histologically confirmed single lesion comprising monoclonal plasma cell infilteration, a negative skeletal survey, and no evidence of tumor in the bone marrow (Krishnamoorthy *et al.*, 2010).

Eighty to ninety percent of EMPs arise in the upper aerodigestive tract such as head, neck, upper respiratory tract (Nolan *et al.*, 2005). These are typically found in males in their sixties. Gastrointestinal involvement of all EMPs is very rare, representing less than 5% (Alexiou *et al.*, 1999). Gastrointestinal tract can be involved by EMP, which the small intestine being the most common, followed by stomach, colon and esophagus (Krishnamoorthy *et al.*, 2010). Symptom of GPs are non-specific symptom including anorexia, weight loss, epigastric discomfort or gastrointestinal bleeding (Nolan *et al.*, 2005).

Pathologically, gastric plasmacytoma (GP) has been hypothesized to originate from lymphoid follicles in the submucosa or from plasma cell in the lamina propria (Yoon *et al.*, 1999). Because primary GP originated from submucosa or lamina propria, EUS showed hypoechoic mass arising from the submucosal layer. Fukuhara *et al.*, (2016) reported hypoechoic mass in the submucosal layer on EUS. Also in our case, gastric GP showed homogenous hypoechoic mass deriving form submucosal layer.

A few radiologic finding of gastric GP reported. Yoon *et al.*, (1999) reported two case of primary GP in stomach, which presented polypoid mass in stomach on upper gastrointestinal series and gastric wall thickening on enhanced CT. but one of these case is homogenous enhancement but other case is poor enhancement pattern on enhanced CT images. And Fukuhara *et al.*, (2016) reported also focal wall thickening with hyperenhancement on the greater curvature of the stomach on CT scan images. But Zhao *et al.*, (2014) reported a case of primary GP that homogenous enhancing extraluminal mass in gastric mid body on CT images. Also primary GP of our case presented extraluminal mass with homogenous gradual enhancement pattern like a SET. Based on previous cases and our case, the CT findings of gastric GP can show various enhancement pattern. Primary gastric GP differentiate from gastric carcinoma, lymphoma or SET such as gastrointestinal stromal tumor (GIST), ectopic pancreas, It is difficult to distinguish gastric GP by CT finding only. CT, endoscopic finding and biopsy should be considered.

Treatment of EMP is no general guideline. Generally treatment is surgery or irradiation, with or without chemotherapy. After treatment of EMP in non-upper aerodigestive regions, 21.2% of patients had

recurrence and 14.1% of them converted to multiple myeloma (Krishnamoorthy et al., 2010).

In conclusion, gastric GP is rare, making it difficult to diagnose. In most cases, gastric GP presents focal or diffuse wall thickening or extraluminal mass of stomach on CT. Unlike the case reported previously, there was no case that could be mistaken for SET on CT like our case. And CT finding that looks like a SET has not been reported yet.

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