

## A rare cause of gastrointestinal bleeding, “GANT”

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### ABSTRACT

Acute gastrointestinal hemorrhage is frequently seen in emergency surgical conditions. Benign pathologies are generally responsible in etiologic factors. Our case is a GANT. GANT is a mesenchymal tumor originated from Cajal cells or precursors of them. Cajal cells are pacemaker cells in the intestinal wall. Because GANT is a very rare tumor, we would like to report our case to the medical literature. We think very rare cases and treatments of its as like our case create view point states in etiology of acute gastrointestinal hemorrhage.

**Keywords:** Gastrointestinal bleeding, duodenal, shock

### INTRODUCTION

Gastrointestinal stromal tumors are a rare mesenchymal tumors which are thought to be originated from neoplastic transformation of pacemaker cells (interstitial Cajal cells) or precursors of them in the intestinal wall (1). Gastrointestinal autonomic nerve tumors (GANTs) were first described by Herrera *et al.* in 1984 (2). The authors used the term plexosarcoma to describe this entity (1). GANTs, also termed „plexosarcomas“, arise from autonomic nervous system plexuses of the gastrointestinal tract. Recent studies indicates GANTs, like GIST tumors, express KIT mutation. GANTs share numerous morphologic and genetic features. For this reason, they are classified as a subtype of GIST tumors, instead of being regarded as a different entity. GANTs are fatal and must be considered malignant. They need radical surgical resection. Radical surgical resection of gastrointestinal autonomic nerve tumors seems to be the only available curative approach to date, and long term survival is possible even in large metastasized tumors. To date, the diagnosis of GIST is strongly suggested by immunostaining for the transmembrane tyrosine kinase receptor CD117 and c-kit gene mutations (3,4).

We aimed to present a case who admitted to our hospital with gastrointestinal bleeding and treated by resection of the bleeding tumoral mass located in the third

segment of duodenum. The tumor was diagnosed as “GANT” later on by histopathologic examination.

### CASE PRESENTATION

A 73-year-old female admitted to our hospital with melena and hypotension. In endoscopic examination there was a bleeding tumoral mass in the third segment of duodenum. Immediate replacement therapy with blood transfusion and intravenous crystalloid fluids was started. Vital signs were monitored. Once the patient has been stabilized, she was operated on. A 3\*3 cm intramural mass in the third segment of duodenum has been observed. Resection of the involved segment and then duodenostomy and jejunostomy were performed. Drainage tube was placed in the abdomen and the operation was ended.

The patient had an uncomplicated postoperative course, was started on clear liquids on postoperative day 3, the drainage tube in the abdomen was removed on postoperative day 4. She was discharged on postoperative day 7.

Histopathologic examination of 3\*3 cm tumoral mass revealed NSE; (+), CD 117:diffuse (+), Vimentine: diffuse (+), Ki67: %1-2 (+), Pycnosis (+) gastrointestinal autonomic nerve tumor (GANT) (Figure 1).

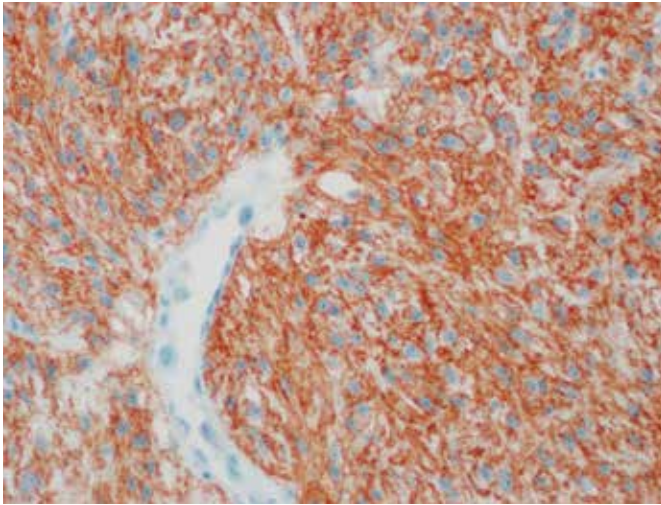
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**Figure 1.** Histopathological appearance of the GANT

## DISCUSSION

GISTs are a rare entity consisting of the 1% of the primary tumors of gastrointestinal system (5). They can be seen anywhere along the gastrointestinal tract. They most commonly seen in the stomach (60%) and the small intestine (20-30%). Colon involvement is 10% and esophagus involvement is 5% of cases (6). Lesions which cannot be differentiated on morphological and immunophenotypical grounds from GISTs have been reported in localisations other than gastrointestinal tract such as mesentery, omentum and retroperiton (6). GANT could be seen in a wide range of age with a slight male predominance (7,8). It most commonly involves small intestine, however, less commonly, stomach, omentum, peritoneal surface, esophagus could be affected. Other neurogenic tumors such as neuroblastoma and paraganglioma should be considered in the differential diagnosis. Ultrastructural studies of all the reported cases revealed features suggestive of myenteric plexus in origin. The diagnostic ultrastructural features included the presence of long, closely opposed cell processes containing intermediate filaments, dense-core neurosecretory granules, microtubules, and synapselike structures with variable numbers of neurosecretory granules and small vesicles. The essential ultrastructural criteria applied for the diagnosis of GANT in all reported cases included neurosecretory granules and intermediate filaments (9). GANT has malign potential and local recurrence after resection is possible (10,11). aggressive tumor debulking without any further treatment could also be considered suitable in terms of prolonged survival and quality of life. It is difficult to differentiate subtypes of GIST tumors. light microscopic studies yielded ambiguous results and ultrastructural examination was required in order to establish an accurate diagnosis of gastrointestinal autonomic nerve tumor (12,13). Muscle, neuronal, Cajal, Schwannian cell differentiation is common among these tumors. Sometimes tumoral mass could be a combination of these subtypes. They are CD117 positive. CD117 positive could have an important clinical impact, since tyrosine kinase inhibitors have

yielded good responses in other CD117 positive tumor entities. To date there are no reports on the use of tyrosine kinase inhibitors in CD 117 positive GANTs, but promising data for c-kit positive GISTs which were successfully treated with tyrosine kinase inhibitors have recently been published (14). Even in cases of large tumor masses, non-curative, aggressive surgical tumor debulking is potentially useful to improve the patient's quality of life. Stromal tumors express smooth muscle differentiation in such gastrointestinal segments as esophagus and rectum where stromal tumors are subject to more powerful contractile forces. In small bowel segments, on the other hand, where coordinated activity is required, GANTs are common (15-17).

Treatment modality for GANT is surgical resection of the tumor with clear margins. In patients in whom there remains residual tumoral tissue or the properly resected tumor shows malignant characteristics, imatinib treatment leads successful cure. Although rarely seen, in the differential diagnosis of gastrointestinal bleeding GANT should be kept in mind.

**Conflict of Interest:** No conflict of interest was declared by the authors.

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