

Laparoscopic sleeve excision of lesser curvature gastrointestinal autonomic nerve tumour

Ahmed P¹, Reddy A², Narayanan CD³

¹Dr Pervez Ahmed
MBBS, MS, Assistant Professor
dr_ahmed_pervez@yahoo.co.in
²Dr Anirudh Reddy
MBBS, Resident
anirudhreddy_89@hotmail.com
³Dr Narayanan Cunnigaiper
Dhanasekhar
MBBS, MS, FRCS, Professor
cdnarayan24@yahoo.com
^{1,2,3}Department of General
Surgery
Sri Ramachandra University
Chennai, India

Received: 24-01-2015
Revised: 25-02-2015
Accepted: 10-04-2015

Correspondence to:
Dr Pervez Ahmed
dr_ahmed_pervez@yahoo.co.in
(0)9791053520

ABSTRACT

Gastrointestinal autonomic nerve tumors (GANT) are uncommon stromal tumors of the intestinal tract. Their histological appearance is similar to that of other gastrointestinal stromal tumors. We report probably the first laparoscopic excision of this rare tumor from our geographical region and compare our findings with the available case reports in the medical literature. A 38 year old male patient undergoing a routine health check was diagnosed with an exophytic growth on the lesser curvature of stomach on computerized tomography and underwent an initial endoscopic biopsy which was inconclusive. He underwent a laparoscopic sleeve excision of the GANT on the lesser curvature. Radical surgical resection of gastrointestinal autonomic nerve tumors seems to be the only available curative approach and survival seems possible even in large tumors.

Keywords: Gastrointestinal Autonomic Nerve Tumour (GANT), lesser curve of stomach, left gastric artery branch, laparoscopic sleeve excision, endostapler

Introduction

Gastrointestinal Autonomic Nerve Tumours (GANTs) are rare, about 1% of all malignant gastrointestinal stromal tumors (GIST). GANTs were first described by Herrera et al in 1984. [1] It consists of a subgroup of gastrointestinal stromal tumors (GISTs) with a specific ultrastructural appearance, suggesting origins from neurons of the enteric plexus. [2] The term plexosarcoma was used to describe this entity. [1, 2] In the last decade several investigators have recognized the distinct characteristics of this type of tumor. [2, 3, 4, 5, 6]

GANTs are low-grade spindle-cell neoplasms but cannot be definitely

distinguished from stromal tumors or other neurogenic tumors by histological or immunohistochemical features. To date, the diagnosis of GIST is strongly suggested by immunostaining for the transmembrane tyrosine kinase receptor CD117 and c-kit gene mutations. [7, 8] GISTs have been defined as tumors of the interstitial cells of Cajal. [9] GANTs also showed CD117 staining, some considered GISTs to be identical to GANTs. Eyden et al. have studied 82 gastrointestinal stromal tumors in terms of cellular differentiation by using electron microscopy. [6] They demonstrated that electron microscopy was needed to diagnose GANTs and exclude other

gastrointestinal stromal tumors.

There is no clear mention of laparoscopic excision of these tumors especially on the lesser curvature in Indian literature. We report this case of laparoscopic excision of GANT.

Case report

A 38 year old male patient with no other symptoms excepting abdominal discomfort of 2 months duration came with an ultrasound report of an intra abdominal mass. He underwent a contrast enhanced CT which revealed an exophytic growth on lesser curvature of stomach with areas of necrosis. (Fig.1) He underwent an endoscopy which showed a mass on fundus, biopsy proved to be inconclusive. Laboratory parameters and CT thorax were normal.

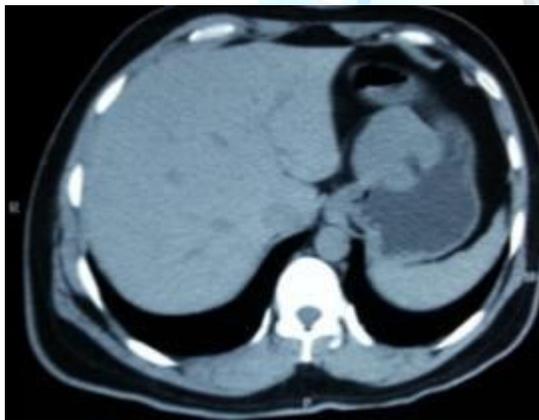


Fig. 1 CT scan of the tumor abutting the stomach

He underwent a diagnostic laparoscopy which revealed an exophytic mass 6 x 6 x 4cms probably a gastrointestinal stromal tumor from the lesser curvature of the stomach about 6cm distal to the oesophagogastric junction. There was no ascites. Liver, omentum, peritoneum and pelvis were normal. The patient underwent a totally laparoscopic

excision of the tumour along with part of the lesser curvature similar to a sleeve resection. Patient tolerated the procedure well and was extubated. Postoperative period was uneventful. Patient was discharged after 2 days.



Fig. 2 Laparoscopic stapling of the tumour on the lesser curve of stomach

Histopathology revealed that the tumour was a variant of a gastrointestinal tumor. It had a mitotic rate of 5/50 HPF, no necrosis, low grade, free margins, no nodes involved. The immunohistochemistry reports were Ckit negative, DOG1 negative, PDGFR negative and SMA negative. The tumor was S100 positive and had lymphoid aggregates in the serosa.

Discussion

The literature search yielded 120 published cases of GANTs. The age range was (10 to 85 years). Most patients were older than 50 years but our patient was younger. The tumors were found more often in males (62 males vs 58 females) similar to our patient. The reported clinical signs and symptoms were non specific. In most cases the common site of tumor was the stomach (similar to our case), duodenum, jejunum and ileum. On

imaging studies, the tumor often presents as a large and lobulated solid mass, with areas of necrosis. [4] It is locally invasive. Radiological techniques did not permit a distinction between GANTs and other GISTs. [5] The greatest dimension of the tumor was determined in 81 cases, and ranged from 1.5 cm to 40 cm (median 10.4 cm) with our patient being 6cm. These bulky tumors were predominantly cystic and necrotic just as our case.

Histologically, the tumors showed a variety of patterns. Most lesions were spindle cell tumors with features resembling either smooth muscle tumors or neurogenic tumors. Epithelioid cells were also detected. The cellular arrangement was whorled, patternless, fascicular, palisaded or storiform. The tumor cells had an eosinophilic cytoplasm. Pleomorphism was not significant and mitosis ranged from 1 to 23/10 high-power fields.

Results of immunohistochemical studies demonstrated that the tumor was most often reactive to vimentin and other markers of nerve tissues such as neuron-specific enolase (NSE), synaptophysin, S-100 protein, neurofilament, and chromogranin A. [9] Unlike our patient who was only S100 positive. These proteins are normally expressed by neurons from the autonomic enteric nerve plexus, supporting a histogenesis of GANTs from enteric autonomic plexuses of Meissner or Auerbach. The muscle marker desmin and muscle specific actin could not be demonstrated, although focal alpha SMA positivity was seen in 7 reported cases. Except for one study (2 cases) focal staining for cytokeratins

(CAM5.2) was found to be negative. GANTs usually lacked smooth muscle cell features. [2,6]

Ultrastructural studies of all the reported cases revealed features suggestive of myenteric plexus in origin (similar to our case). [3] The diagnostic features included 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. It should be noted that five reported GANTs showed weak signs of smooth-muscle morphology. Therefore, the presence of smooth muscle cell features might also suggest GANT [6]

The primary therapy in reported cases was surgical resection (similar to our patient), in 2 cases preceded by radiotherapy, and in nine cases preceded by chemotherapy (not done in our case as it was low grade). [10] Three patients received chemotherapy alone. There was no evidence of clinical response using chemotherapy or radiation.

Gastrointestinal autonomic nerve tumors occurred with an estimated frequency of 1% of all malignant gastrointestinal tumors and up to 25% of gastrointestinal stromal tumors. [6, 7] In retrospective studies of collected pathological specimens, several investigators found that light microscopic studies yielded ambiguous

results and ultrastructural examination was required in order to establish an accurate diagnosis of gastrointestinal autonomic nerve tumor. [7] Based on these observations we believe that GANTs are probably more common than previously thought. The rarity of GANT may therefore be a consequence of the unavailability of routine electron microscopic analysis. But it appears that, despite their low grade malignant histological appearance, most GANTs had an uncertain prognosis in case of metastases. [8]

The recent observation that most GANTs were 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. [10] Radical surgical resection appears to be the most promising and solely curative treatment for gastrointestinal autonomic nerve tumors. [10]

Conclusion

Laparoscopic surgery is feasible in such rare tumours located in precarious areas. Higher imaging modalities and equipment to do ultrastructural studies like electron microscopy are becoming necessary. One must always approach such tumours with caution and convert to open surgery without ego at the first sign of trouble.

References

1. Herrera GA, Pinto de Moraes H, Grizzle WE, Han SG. Malignant small bowel neoplasm of enteric plexus derivation (plexosarcoma). Light and electron microscopic study confirming the origin of the neoplasm. *Dig Dis Sci* 1984;29:275-284.
2. Herrera GA. Small bowel neoplasm. *Dig Dis Sci* 1985;30:698.
3. Walker P, Dvorak AM. Gastrointestinal autonomic nerve (GAN) tumor. Ultrastructural evidence for a newly recognized entity. *Arch Pathol Lab Med* 1986; 110:309-316.
4. Tortella BJ, Matthews JB, Antonioli DA, Dvorak AM, Silen W. Gastric autonomic nerve (GAN) tumor and extra-adrenal paraganglioma in Carney's triad. A common origin. *Ann Surg* 1987;205:221-225.
5. Dvorak AM. Gut autonomic nerve (GAN) tumors. In: Watanabe S, Wolf M, Sommers SC, editors. *Digestive disease pathology*. Vol 2. Philadelphia: WB Saunders; 1989. p. 49-66.
6. Eyden B, Chorneyko KA, Shanks JH, Menasce LP, Banerjee SS. Contribution of electron microscopy to understanding cellular differentiation in mesenchymal tumors of the gastrointestinal tract: a study of 82 tumors. *Ultrastruct Pathol* 2002; 26:269-285.
7. Miettinen M, Sobin LH, Sarlomo-Rikala M. Immunohistochemical spectrum of GISTs at different sites and their differential diagnosis with a reference to CD117 (KIT). *Mod Pathol* 2000;13:1134-1142.

8. Schmid S, Wegmann W. Gastrointestinal pacemaker cell tumor: clinicopathological, immunohistochemical, and ultrastructural study with special reference to c-kit receptor antibody. *Virchows Arch* 2000;436: 234-242.
9. Kindblom LG, Remotti HE, Aldenborg F, Meis-Kindblom JM. Gastrointestinal pacemaker cell tumor (GIPACT): gastrointestinal stromal tumors show phenotypic characteristics of the interstitial cells of Cajal. *Am J Pathol* 1998; 152:1259-1269.
10. Joensuu H, Fletcher C, Dimitrijevic S, Silberman S, Roberts P, Demetri G. Management of malignant gastrointestinal stromal tumours. *Lancet Oncol* 2002;3:655-664.

Cite this article as: Ahmed P, Reddy A, Narayanan CD. Laparoscopic sleeve excision of lesser curvature gastrointestinal autonomic nerve tumour. *Int J Med and Dent Sci* 2015; 4(2):856-860.

**Source of Support: Nil
Conflict of Interest: No**

