Case Report

Gastrointestinal autonomic neural tumour of colon: a case report of rare unusual tumour of colon

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ABSTRACT

Gastrointestinal autonomic nerve tumours (GANTs) are uncommon stromal tumors of the intestinal tract. They can involve any part of the gastrointestinal system, but are very rarely seen in the Colon. We report a unique case of Gastrointestinal Autonomic Neural Tumour (GANTs) of colon extending from ascending colon to 2/3rd of the transverse colon. A 50-year-old male patient was admitted with complaint of pain in right side of abdomen. USG revealed t/s/o lymphoma/Carcinoma Colon. CECT Abdomen revealed? Lymphoma? Carcinoma extending from ascending colon to 2/3rd of the transverse colon with Lymphadenopathy. Exploratory Laparotomy was performed followed by Right Hemicolectomy. Histopathology and Immunohistochemistry confirmed the colonic lesion to be a Gastrointestinal Autonomic Neural Tumour. Literature review of the few reported cases has suggested radical surgical excision to be the best approach. Prognosis tends to be favourable after resection.

Keywords: Gastrointestinal autonomic neural tumour, Tumour of colon, GIST, GANT

INTRODUCTION

The gastrointestinal autonomic nerve tumours (GANTs) were first described and defined by Herrera et al, in 1984. GANTs are uncommon stromal tumours accounting for 0.1% of benign tumours of the gastrointestinal tract. It is a subgroup of gastrointestinal stromal tumours (GISTs) with specific ultrastructural differences; suggesting its origin from the myenteric plexus. Schwannomas belong to this group and may develop practically in any anatomic region. Without Immunohistochemical studies, Schwannomas are often misdiagnosed as leiomyomas or leiomyosarcoma and generally present as an asymptomatic mass and/ or with non-specific symptoms of fatigue and prolonged pain as well as signs of low grade pyrexia, anemia and hemorrhage. Conventional pathological techniques are usually not diagnostic; electron microscopy often being required to establish the diagnosis of GANTs and to exclude them from other gastrointestinal tumours.

Common sites for GANTs include the stomach, duodenum, jejunum, ileum and to date literature search has revealed only twenty cases of colonic schwannomas and only four reported cases of rectal schwannomas.

We report rare GANTs of colon.

CASE REPORT

A 50 years old male came to surgery opd with complaint of Pain in Right lower abdomen since 2 months and history of malena since 2 months. Pallor was present. Blood investigations Hb was 4.2 gm %. All other parameters were within normal limits. USG Abdomen suggested thickening of ascending and transverse colon s/o Lymphoma or carcinoma of colon with ascites. CECT Abdomen shows evidence of ill defined heterogeneously enhancing diffuse circumferential thickening involving whole of ascending colon and 2/3rd of transverse colon,
lesion is causing narrowing of lumen, there is also a homogeneously enhancing circumferential thickening of pyloric canal approx 1.5 cm with centimetric and subcentimetric Para-aortic and bilateral lymphadenopathy s/o? Lymphoma or Carcinoma extending from ascending colon to 2/3rd of Transverse Colon and similar lesion seen in pyloric canal with lymphadenopathy and moderate ascites (Figure 1 and Figure 2).

![Figure 1: CECT abdomen revealed thickening of ascending and transverse colon.](image1)

On basis of USG and CECT Abdomen patient’s elective Laparotomy was planned. On pre-operative day patient developed pain in abdomen, X-ray abdomen S/o Perforation of abdomen. Emergency Exploratory Laparotomy was performed and there was perforation at Caecum. Caecum, ascending colon and proximal 2/3rd transverse colon was thickened and dilated.

Right Hemicolecetomy was performed and segment of ileo-caecal junction, transverse colon along with tumour mass was resected (Figure 3).

Resected specimen was sent for histopathology. Section from tumour mass shows ulceration and superior erosion of mucosa, non specific inflammatory infiltrate of sub-mucosa and effacement of architecture of deeper tissues (muscularis and Serosa). Gastrointestinal Autonomic Neural Tumour (GANTS) of colon was given as histological diagnosis (Figure 4 and Figure 5).

![Figure 3: Resected specimen pf colon shows site of perforation along with thickened mass.](image2)

Immunohistochemistry was done for confirmation. Immunohistochemistry staining shows positive for vimentin, S-100, CD117 and neuron specific enolase (NSE).
DISCUSSION

Gastrointestinal autonomic nerve tumors represent a distinct but rare subcategory of gastrointestinal stromal tumors accounting for 1% of all malignant gastrointestinal tumors and up to 25% of gastrointestinal stromal tumors. Initially was described as plexosarcomas, these tumors have been reported to be more common in males and have a wide spectrum of age range. Literature review has shown rare association of GANTs with neurofibromatosis and adrenal ganglioneuroma. Schwannomas are types of gastrointestinal tract autonomic tumours and amongst this group, rectal schwannomas are very rare. Conventional imaging modalities such as barium enema, colonoscopy, endoscopic ultrasound, computerized tomography and MRI have been used to investigate these patients but there are no definite radiological criteria to differentiate benign from malignant stromal tumor. Recently Levy et al. have reported that CT imaging may help to differentiate GANTs from Gastro-intestinal stromal tumours (GISTs). Low attenuation features (indicative of haemorrhage, necrosis and degeneration typically found in the centre of the GISTs) were not seen in GANTs. Endoscopic ultrasound has been suggested to be reliable in predicting malignancy and the predictive features being irregular margins, depth of penetration, cystic spaces and lymph nodes with a malignant pattern. Endoscopic ultrasound guided fine needle aspiration with Immunohistochemical analysis may be useful in the preoperative diagnosis of GIST. The differentiation of schwannomas from other stromal tumours is important because the latter group has high-risk of malignant behaviour. Immunohistochemical studies of GANTs have usually demonstrated positivity to vimentin, CD34 and CD117. Positive reactivity with neuron specific enolase (NSE), S-100 protein, synaptophysin, and chromogranin A (proteins expressed by neurons from the autonomic nerve plexus) have also been reported and this support the histogenesis of GANTs from the autonomic plexus of Meissner or Auerbach. GANT is an ultrastructural variant of GIST, based on its consistent CD117 positivity and the presence of GIST-specific c-kit gene mutations has shown in significant number of cases. Immunohistochemistry is only test for definitive diagnosis of GANTs. The ultrastructural criteria that suggest origin from myenteric plexus are neuron-like cells with long cytoplasmic processes containing microtubules, bulbous synapocrine structures with dense core neurosecretory type granules and empty vesicles though Ultra structural examination is not available at most pathological units (including ours) and it is suggested that representative samples should be sent to centres having electron microscopic facilities. Although GANTs are generally considered benign, Lauwers et al reported that 30% of these patients developed local recurrence and as a result, resection or debulking is the optimal treatment. There was no case which gives evidence of clinical response on chemotherapy or radiation. The recent finding of CD117 receptors in GANTs combined with technological advances has led to the development of anti-tumour agents. Treatment of such CD117 positive GANTs with tyrosine kinase inhibitors have been shown to be beneficial and could in future, represent an appropriate form of palliative therapy in those patients with unresectable as well as metastatic tumors.

CONCLUSION

Literature review has suggested Immunohistochemistry is the best to establish the diagnosis and based on which, surgical resection or debulking the best approach. Prognosis tends to be favorable after resection.

ACKNOWLEDGEMENTS

It is acknowledgements to who so ever concerned this case study has been done on individual basis and has not been supported by any grant from any source.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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