ISSN: 0976-3031

Available Online at http://www.recentscientific.com

CODEN: IJRSFP (USA)

International Journal of Recent Scientific Research
Vol. 15, Issue, 06, pp.4789-4793, June, 2024

International Journal of Recent Scientific Research

DOI: 10.24327/IJRSR

Research Article

A RARE CASE REPORT OF GASTROINTESTINAL AUTONOMIC NERVE TUMOUR

Dr.D Mohit Rao[1]., Dr Pranisha Yalala[2]., Dr. Sandeep Madineni[3]., Dr.G.Ramakrishna Reddy[4] and Dr.K. Venkat Ram Reddy[5]

[1,2]Resident, [3]Associate professor, [4] Professor and HOD and [5]Professor
Sri Venkata Sai Medical College, Mahabubnagar, Telangana, India

DOI: http://dx.doi.org/10.24327/ijrsr.20241506.0898

ARTICLE INFO

Article History:

Received 14th May, 2024
Received in revised form 20th May, 2024
Accepted 15th June, 2024
Published online 28th June, 2024

Keywords:

GANT, GIST, Carcinoma Stomach, Neurofibroma

ABSTRACT

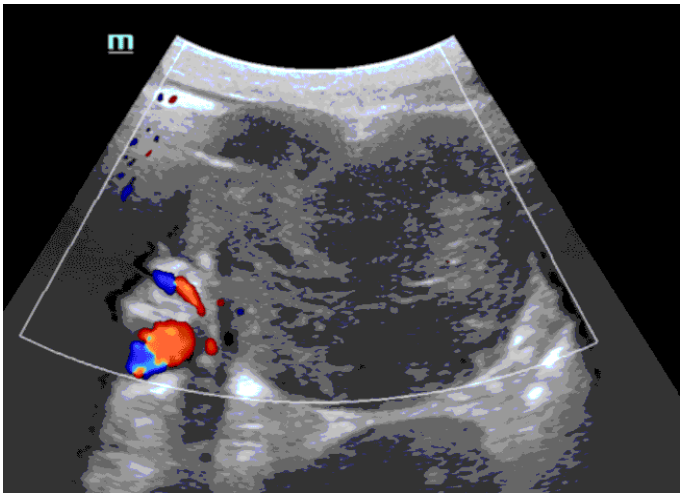
Gastrointestinal autonomic nerve tumors (GANT) are rare tumors linked to the autonomic nervous plexuses in the gastrointestinal tract. Distinguishing them from s tromal tumors involves examining their unique ultra structural features, often through electron microscopy and immunohistochemical analysis. Despite their slow growth, they tend to have an aggressive clinical course and are often associated with a poor prognosis, frequently leading to fatalities. Here we present a case of 35 year old female presenting in the pain abdomen, loss of weight and appetite.

INTRODUCTION

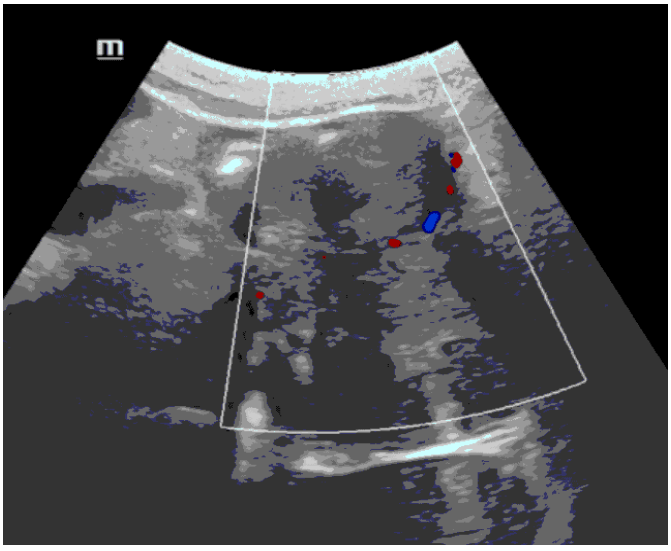
Gastrointestinal autonomic nerve tumors (GANTs) are exceptionally rare growths that constitute a distinct subgroup within gastrointestinal stromal tumors (GISTs). While their radiological characteristics may lack specificity,the presence of particular morphological features could signal the diagnosis.

CASE REPORT

A 35 year old female patient came with the complaints of pain abdomen, weight loss and loss of appetite since 1month.

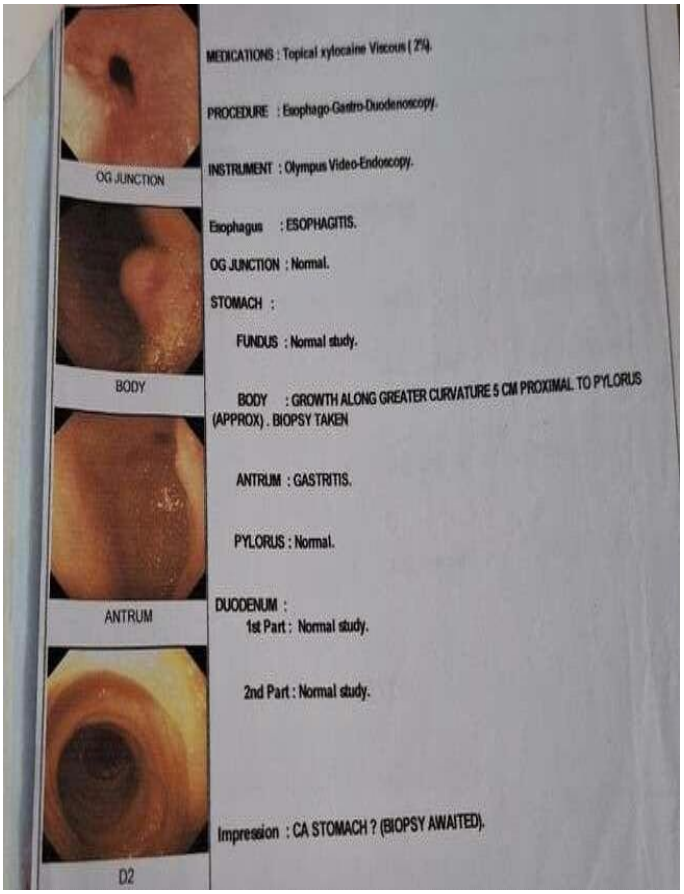


She underwent an ultrasound scan as suggested by her treating surgeon which revealed a large heterogeneously hypoechoic mass in the right hypochondriac region measuring 7x7cms with minimal flow on application of color doppler. Differential diagnosis of gastric neoplasm or pancreatic mass was given. The pateint was advised Upper GI Endoscopy and CECT Abdomen.

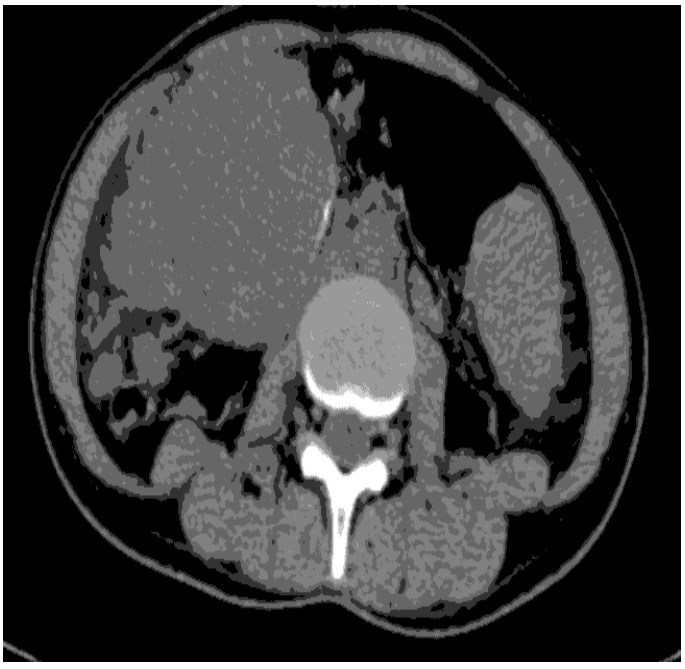


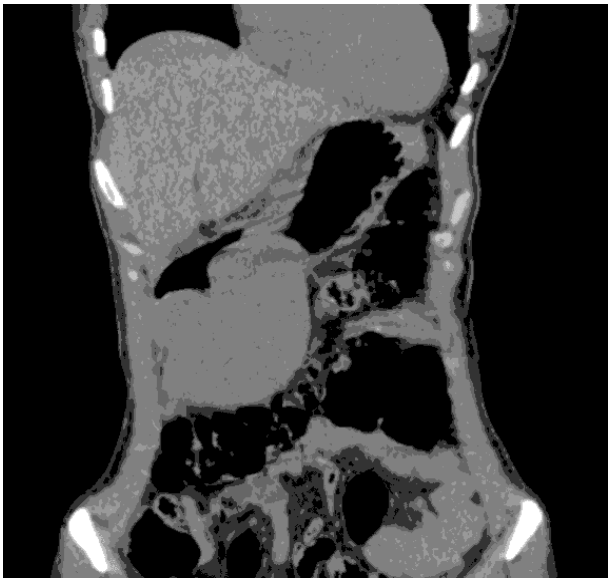
Endoscopy revealed a growth along the greater curvature 5cms proximal to pylorus.

Corresponding author:* **Dr.D Mohit Rao
Sri Venkata Sai Medical College, Mahabubnagar, Telangana, India

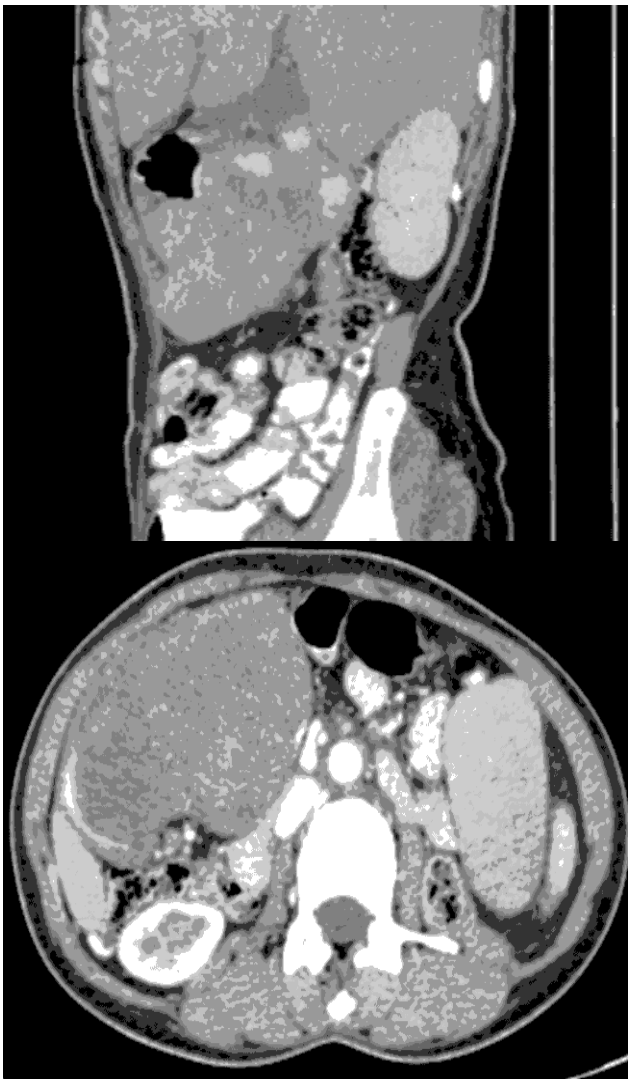


On further imaging with CECT abdomen, a single large lobulated predominantly exophytic soft tissue dense mass lesion arising from the greater curvature of stomach in the pyloric antral region with few peripheral calcifications and internal hypodense components. The lesion measures 8.5x8.3x7.2cms (APxCCxTr) with CT attenuation value of 30-50HU. Asymmetrical wall thickening noted involving pylorus and antrum of stomach with no intraluminal extension or desmoplastic reaction.





On post contrast, minimal heterogeneous enhancement with CT attenuation value of 80-90 HU with few peripheral non enhancing necrotic areas noted on arterial phase. On delayed phase, intense heterogeneous enhancement noted with CT attenuation value of 130-140HU with few non enhancing necrotic areas.

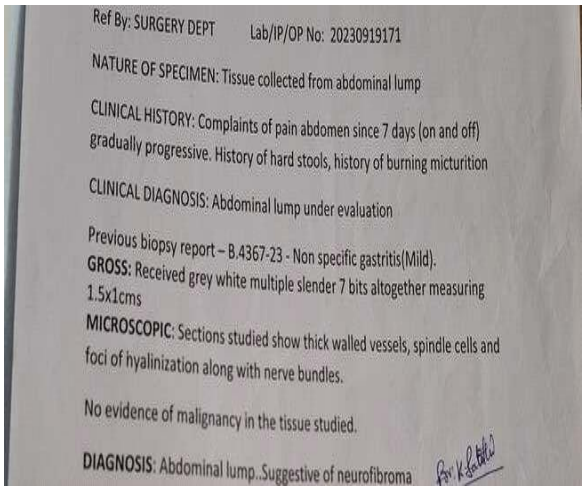


Multiple homogenously enhancing peripancreatic, mesenteric, paracaval lymph nodes noted.

Differential diagnoses were given as

1. GIST
2. Adenocarcinoma of stomach
3. GANT

On needle biopsy of the lesion, sections showed thick walled vessels, spindle cells and foci of hyalinization along with nerve bundles which were suggestive of Neurofibroma.



Ref By: SURGERY DEPT Lab/IP/OP No: 20230919171

NATURE OF SPECIMEN: Tissue collected from abdominal lump

CLINICAL HISTORY: Complaints of pain abdomen since 7 days (on and off) gradually progressive. History of hard stools, history of burning micturition

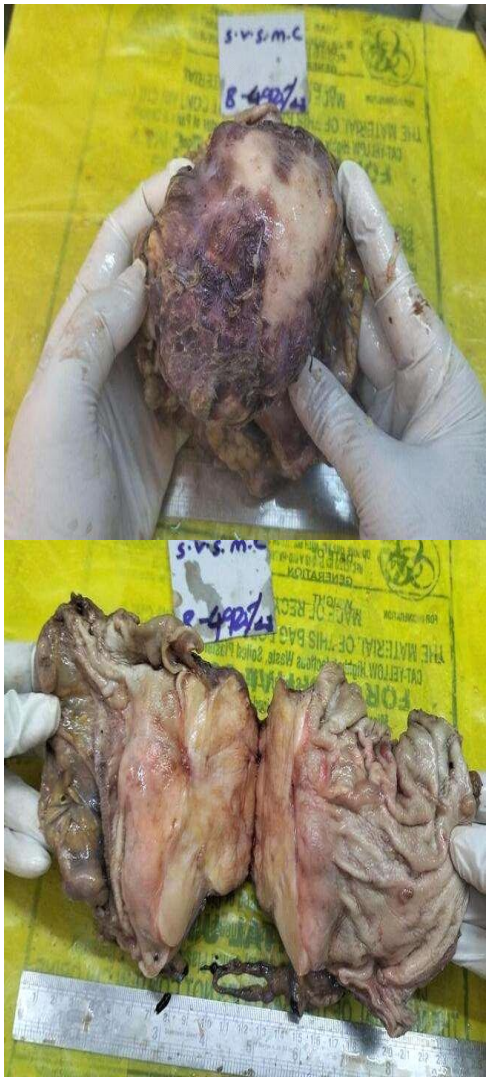
CLINICAL DIAGNOSIS: Abdominal lump under evaluation

Previous biopsy report – B.4367-23 - Non specific gastritis(Mild).
GROSS: Received grey white multiple slender 7 bits altogether measuring 1.5x1cms
MICROSCOPIC: Sections studied show thick walled vessels, spindle cells and foci of hyalinization along with nerve bundles.

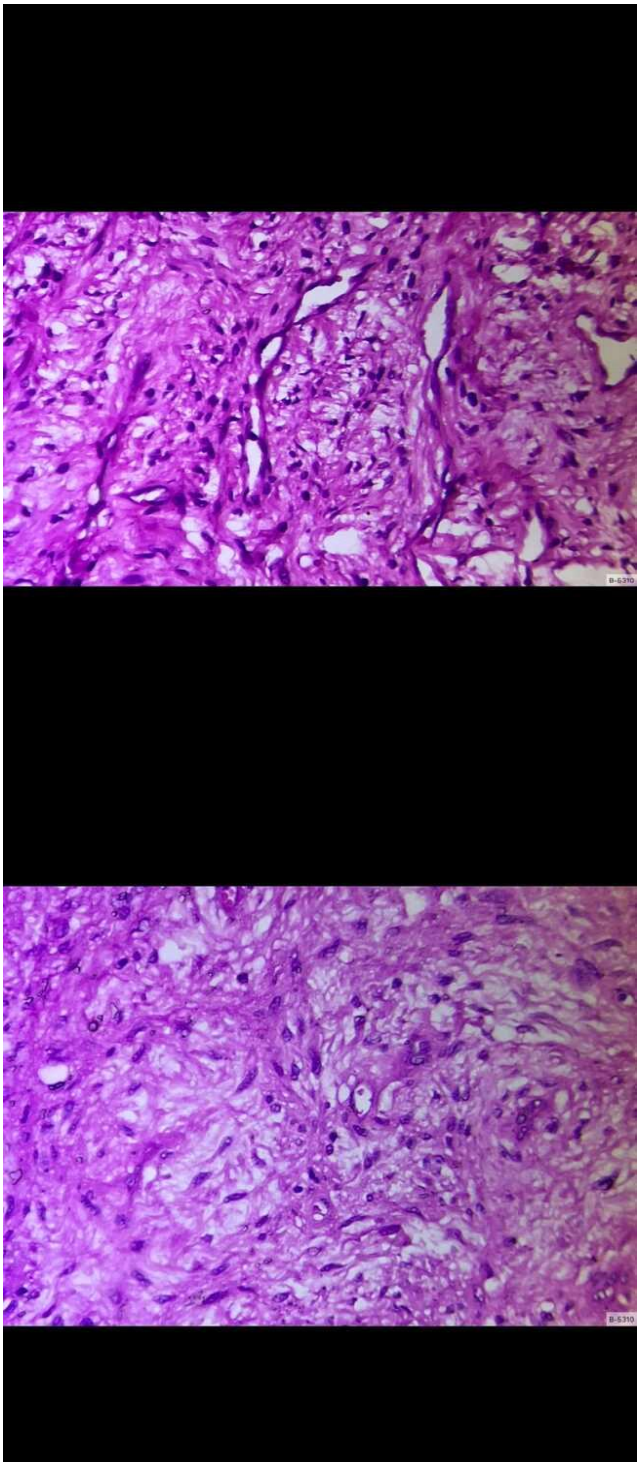
No evidence of malignancy in the tissue studied.

DIAGNOSIS: Abdominal lump..Suggestive of neurofibroma

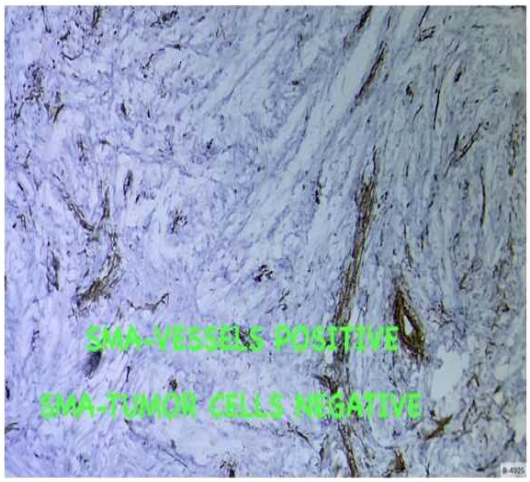
Patient then underwent surgery (distal partial gastrectomy) and the mass was resected en bloc which was sent for histopathological diagnosis.

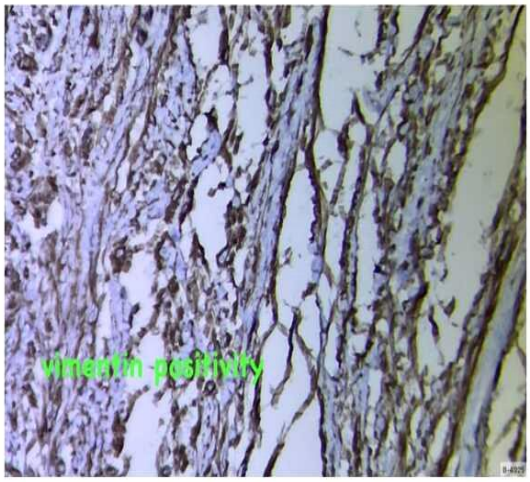


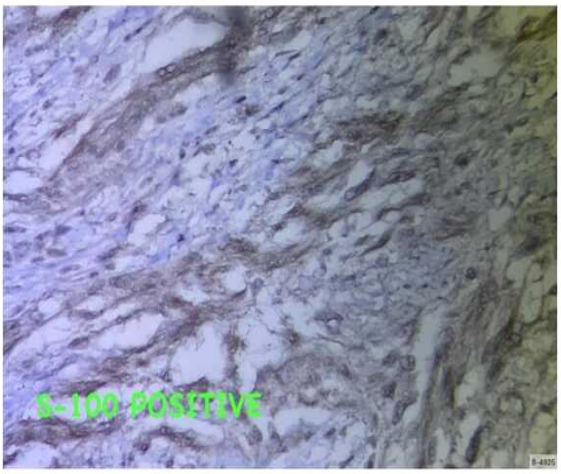
On HPE, the images revealed plump spindled cells in fascicular pattern with lymphocytes in a background of fibrosed to edematous stroma. A fewer than 5 Mitotic figures are seen. H&E 400x

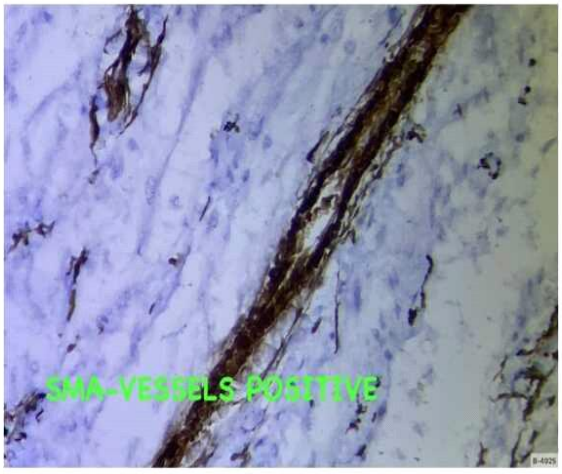


On Immuno histochemistry (IHC) analysis,the lesion was vimentin, S-100, SMA vessels positive and negative for SMA tumor cells, CD34, KIT 117 and desmin which are consistent with Gastrointestinal autonomic nerve tumour (GANT).









Patient was followed up after 6 months with USG, which showed no recurrence.

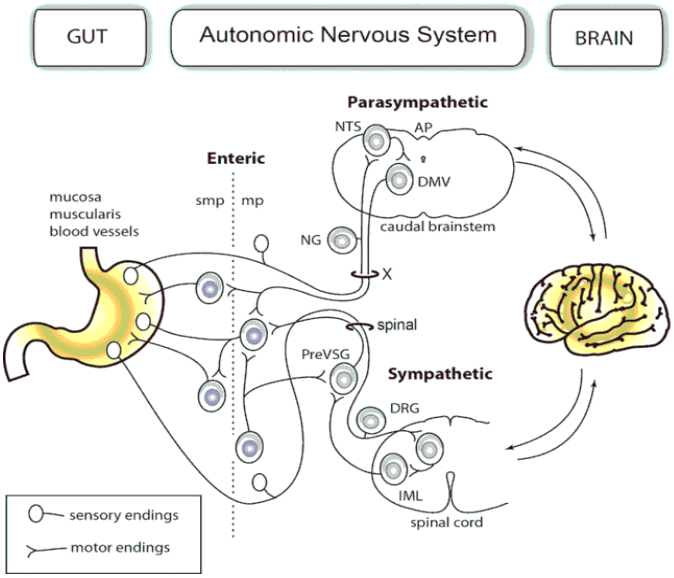
DISCUSSION

Initially termed plexosarcomas, Gastrointestinal Autonomic Nerve (GAN) tumors are now recognized as a distinct entity separate from other gastrointestinal stromal tumors (GISTs). These tumors are exceptionally rare, comprising only 0.1-0.5% of all malignant gastrointestinal neoplasms. They predominantly affect males and typically manifest in adults over 40 years old, with the most common sites being the jejunum, ileum, and stomach. While GAN tumors may occasionally arise in the retroperitoneum, unlike other GISTs, none have been observed in the esophagus or large intestine.

Innervation of the stomach involves the autonomic nervous system: Parasympathetic innervation originates from the anterior and posterior vagal trunks, which are branches of the vagus nerve. Sympathetic innervation arises from the T6-T9 spinal cord segments and travels to the coeliac plexus through the greater splanchnic nerve.

Clinically, gastrointestinal autonomic nerve tumors typically present with nonspecific symptoms such as palpable abdominal masses, abdominal pain, malaise, nausea and vomiting, or

anemia. Distinguishing these tumors from other gastrointestinal stromal tumors based solely on conventional pathological techniques poses challenges. Gross examination often reveals well-defined, lobulated tumors with areas of focal hemorrhage. Diagnosis relies on a combination of light microscopic, ultrastructural, and immunohistochemical analyses. Light microscopic features include arborizing plexiform vasculature dividing tumor cells, characterized by round or short spindle cells with plump round or ovoid nuclei, indistinct cell borders, and bubbly cytoplasm.



Immunohistochemical staining for vimentin, neuron-specific enolase, synaptophysin, and neurofilaments is commonly positive in gastrointestinal autonomic nerve tumors, indicating their origin from the enteric autonomic plexuses of Meissner or Auerbach. Ultrastructural examination reveals neuronal differentiation, evident through well-developed neuritic processes, dense-core neurosecretory-type granules, and synapse-like structures.

Plexosarcomas demonstrate aggressive behavior, often characterized by a significant likelihood of local recurrence and hepatic metastasis. Factors predicting their biological behavior include tumor location, size (greater than 10 cm), and level of mitotic activity. Specific associations with stomach GAN tumors include Von Recklinghausen disease or the Carney triad, which comprises non-epithelial gastric tumors, lung chondromas, and extra-adrenal paragangliomas.

Chemotherapy typically produces limited responses in GAN tumors, making radical excision the sole treatment option associated with favorable outcomes. Due to the difficulties in diagnosing GAN tumors clinically and pathologically, imaging methods could prove invaluable. Although radiological techniques may not distinguish GAN tumors from other gastrointestinal stromal tumors, radiologists should include them in the list of possible diagnoses for stomach and small intestine tumors displaying particular features. They can then suggest specific immunohistochemical techniques to ensure an accurate diagnosis.

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. GANTs are extremely rare tumors that arise from the autonomic nerve plexuses. The diagnosis is based on light microscopic, ultrastructural, and immunohistochemical analyses.

References

1. Herrera GA, DeMoraes HP, Grizzle WE: 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. 10.1007/BF01296263.
2. Marjanovic, Zoran, Slavkovic, Andjelka, Krstic, Marijana, Djodjevic, Ivona and Dimov, Dragan. "Surgical treatment of gastrointestinal autonomic nerve tumors (GANT) in children-2 case reports" Open Medicine, vol. 6, no. 4, 2011, pp. 395-399.
3. Thomas JR, Mrak RE, Libuit N (1994) Gastrointestinal autonomic nerve tumors presenting as high-grade sarcoma. Case report and review of the literature. Dig Dis Sci 39 (9): 2051±2055
4. Lauwers GY, Erlandson RA, Casper ES, Brennan MF, Woodruff JM (1993) Gastrointestinal autonomic nerve tumors. A clinicopathological, immunohistochemical and ultrastructural study of 12 cases. Am J Surg Pathol 17 (9): 887±897
5. Meshikhes, Abdul-Wahed. (2015). Gastrointestinal Autonomic Nerv Tumors: A Clinical Review. Journal of Gastrointestinal Surgery. 19. 10.1007/s11605-015-2798-z.

How to cite this article:

D Mohit Rao., Dr Pranisha Yalala., Sandeep Madineni., G.Ramakrishna Reddy and K. Venkat Ram Reddy. (2024). A rare case report of gastrointestinal autonomic nerve tumour. *Int J Recent Sci Res.*15 (06), pp.4789-4793.

****