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Ganglioneuroma: A Rare Incidentaloma

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ABSTRACT:
An uncommon, benign, well-differentiated tumor that arises from neural crest cells specifically in the sympathetic nervous system is called ganglioneuroma. This kind of tumor, which originates from the autonomic ganglia, usually consists of mature ganglion cells, Schwann cells, and nerve fibers. The posterior mediastinum, retroperitoneum, and adrenal gland are the most prevalent locations for ganglioneuromas, with the cervical region being less common.This is a study of a recent case of a 35-year-old young woman with a history of weight loss along with gastroesophageal reflux symptoms. An incidental abdomen mass was picked up on CT study as a ganglioneuroma. The tumour was removed safely without damaging the surrounding structures. The confirmation of ganglioneuroma was done with the help of histopathological examination. It was confirmed to be a benign tumour.The purpose of this abstract is to give a brief introduction to ganglioneuroma, emphasizing its clinical manifestation, method of diagnosis, and approaches to treatment.

I. CASE REPORT:

A 35-year-old woman with no known co-morbidity presented with a history of progressive unintentional weight loss of 6 kg in 6 months, with symptoms of gastroesophageal reflux disease (heartburn and non-cardiac chest pain) for 3 months intermittently.Additionally, there was no history of dysphagia or odynophagia**.** There were two episodes of colicky lower abdominal pain in the preceding 2 months, requiring hospitalisation. However, no significant precipitating factors, such as abdominal distension were noted. Both the pain episodes were relieved with frequent passage of small /large quantities of stools semi-formed to watery, with no blood or mucus. Bowel movements were otherwise normal with a Bristol score of 4.

Baseline laboratory investigations were non-contributory. Both upper endoscopy and colonoscopy were normal. On Contrast Enhanced Computed Tomography (CECT) with Enterography, there was a lobulated non-enhancing retroperitoneal soft density mass lesion 5.0 x 5.0 cm in the left upper para-aortic region, encasing the origin of the coeliac and superior mesenteric artery. The rest of the viscera liver, spleen gall bladder, kidney and adrenal glands were normal, small intestine was normal. A CT-guided biopsy of the retroperitoneal mass showed the presence of spindle cells and focal areas of ganglioneuromatous proliferation against a backdrop of fibrocollagenous tissue.

Management: The patient was taken up for laparotomy. An L - shaped incision was made on the left upper quadrant of the abdomen. A well-defined lobulated non-enhancing soft tissue lesion of approximate size of 5.0 x 5.0 cm was seen in the left side of the retroperitoneum that was adherent to the left adrenal gland and the left renal artery, superior mesenteric artery, and celiac artery. There were no satellite deposits.

Surgical resection: The left paracolic gutter was mobilised and the flap was raised with the spleen,

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pancreas, and left kidney, and the entire complex was shifted to the right side. The tumour was mobilized releasing the retroperitoneal adhesions; mobilization of the lateral, posterior, and anterior surface of the tumour was completed. The medial surface adhered to the aorta, with a tongue-like extension between the celiac and superior mesenteric vessels. A small segment of the upper polar left renal artery was passing through the lesion and was transected and anastomosed with 6'0 prolene sutures. The adrenal gland was normal. The entire tumour was removed after ligating the feeding vessels arising directly from the aorta. Doppler study of the aorta was done to ensure adequate blood flow was present to the liver, small bowel, and left kidney. The anterior abdominal wall's rectus sheath and muscle layer were opposed with one loop Ethilon and skin sutured with staples. The postoperative period was uneventful. The histology of the resected specimen confirmed a benign ganglioneuroma.

II. DISCUSSION:

Ganglioneuromas are fully grown tumours originating from the sympathetic nervous system and are usually not metabolically active. They come from the neural crest and are classified as neuroblastoma tumours. (4) Although these tumours are neither gender- nor age-specific, children older than ten are typically diagnosed with them. Oftentimes, ganglioneuromas are found accidentally by certain imaging features and do not cause any symptoms (4).

Different biological behaviours are exhibited by three forms of neurogenic tumours: ganglioneuromas, ganglioneuroblastomas, and neuroblastomas. It is thought that sympathetic ganglia are the source of ganglioneuromas, and their histological characteristics distinguish them from other neurogenic tumours like neuroblastomas. (1) First of all, the majority of ganglioneuromas, which fall under the category of neuroblastic tumours, affect youngsters. Although these tumours usually manifest as isolated masses, they occasionally have the potential to spread to lymph nodes. (2)

Theoretically, GNs can form at any point along the sympathetic nervous system. Adrenal glands (21%) the retroperitoneum (37.5%) and the posterior mediastinum (41.5%) are common sites for these tumors. Less frequently, they can also happen in the colon, skin, cervical area, and along the trigeminal nerve. (3). of all primary tumours detected in the retroperitoneal region, 0.72 to 1.6% is retroperitoneal ganglioneuromas. (5)

Ganglioneuromas (GNs) typically show little to no symptoms and have a good prognosis, particularly if they are unintentionally found using radiological imaging. Immunohistochemistry and histological analysis are used to confirm the diagnosis. (3)

The removal of the tumour surgically is the mainstay of treatment for ganglioneuromas. However, because these tumours are often positioned close to major arteries, there is a considerable risk of morbidity and death associated with this method. (4). Clinical settings often do not explore the use of adjuvant systemic chemotherapy or local radiation due to the benign nature of ganglioneuromas (GN). Currently, there is no better way to treat GN than surgical resection, which eliminates the need for radiation or chemotherapy. (3).

Even though Ganglioneuromas (GNs) often advance slowly, quiescently, and indolently, it is recommended to have routine, long-term follow-ups to closely monitor for any local recurrence. (6)

In our case, initially, we anticipated challenges in tumour resection due to the involvement of adjacent structures and vessels close to the tumour. However, these concerns were not encountered during the surgery. Intra-operatively, we were able to separate the tumour without harming the architecture of the surrounding structures.Our patient was on regular follow-up, has shown improvement in weight gain along with no clinical GI issues and is stable with no significantgastrointestinal complaints.

III. CONCLUSION:

Therefore, ganglioneuromas are an intriguing subgroup of neurogenic tumours that are distinguished by their rarity and frequently coincidental findings. Even though they are picked up during imaging scans and pathological assessment a thorough review is necessary to make the diagnosis to distinguish them from other, more aggressive neoplasms. The mainstay of care is still surgical resection, which provides the patient with excellent long-term results. However, in certain situations where complete resection is not achievable, surveillance and follow-up may be recommended.

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NOMEN CLATURE:

1. GN – Ganglioneuroma
2. CECT - Contrast Enhanced Computed Tomography

INTRAOPERATIVE IMAGES:

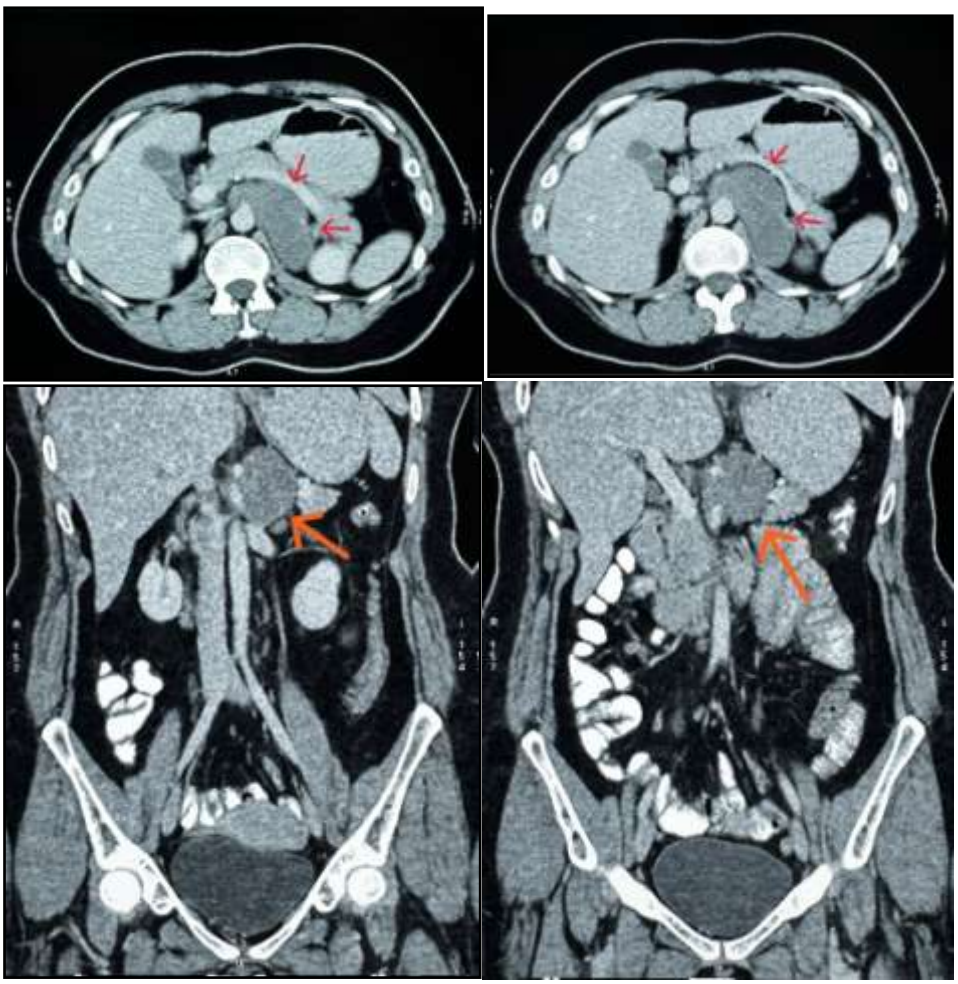

Fig. 1 – CT scan showing the retroperitoneal tumour marked by arrows.

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Figures: CT-guided biopsy of the retroperitoneal mass revealed spindle cells and focal ganglioneuromatous proliferation on a background of fibro collagenous tissue, (Fig 2 and 3)

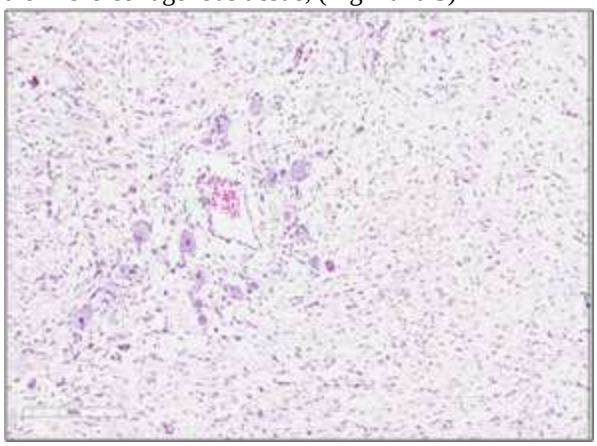

Fig.2- 100 x magnification: showing tumour with wavy spindle cells

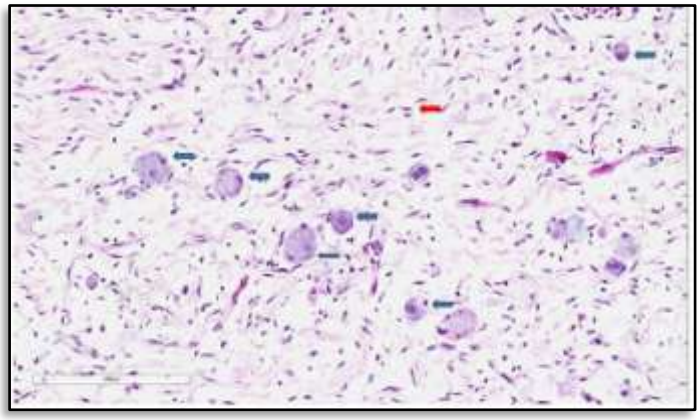

Fig. 3 – 100 x Magnification showing spindle cells (Red arrow) with scattered ganglion cells (Blue arrows)

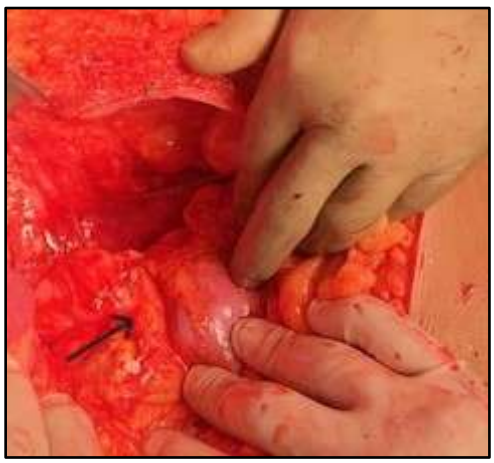

Fig.4: Intra-operative: Image showing the tumour indicated by the arrow.

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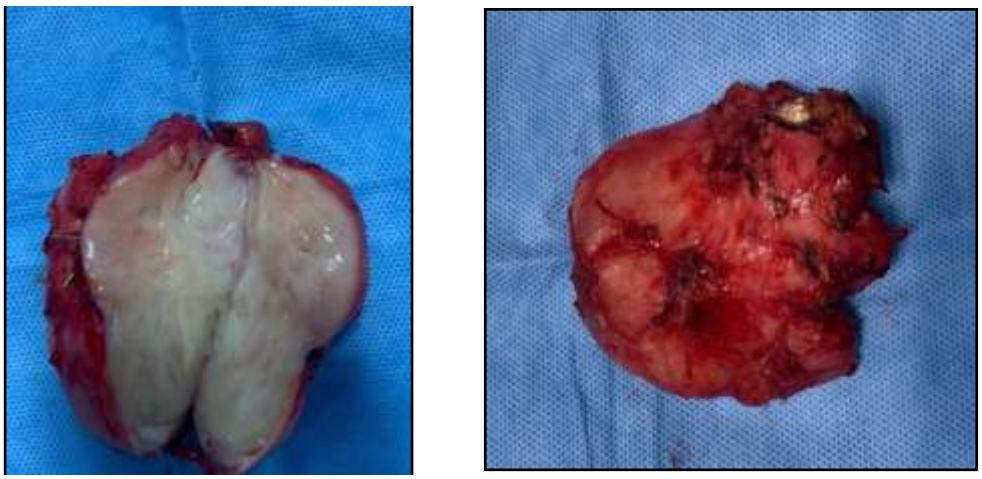

Fig.5: Resected tumour