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Case Report

Artery first approach for resecting ganglioneuroma encasing superior mesenteric artery

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Introduction

Ganglioneuromas are rare and account for 1% of primary retroperitoneal tumors.^{1,2} Their origin from sympathetic chain predisposes them to be inconveniently abutted to major vessels, rendering their excision exigent.^{1,2} We present a case of ganglioneuroma which had encased the superior mesenteric artery (SMA), which was excised, successfully, using the artery first approach.

Case Report

An 44 years old female patient presented with dull pain in upper abdomen and vomiting of 2 months duration. Laboratory

work up was normal. Computerized tomography (CT) showed a 7.2 cm × 6.7 cm × 5.6 cm, well-defined uniformly hypo-dense mass in upper retro-peritoneum, posterior to the head of the pancreas and left lobe of liver. It was wedged between the inferior vena cava (IVC), aorta, and right renal vessels – posteriorly; and splenic and portal vein – anteriorly. The lesion was indenting the porta and posterior aspect of left lobe of liver. Duodenal C-loop was widened by the mass, causing luminal narrowing. The hepatic artery was marginated and SMA was totally encased by the mass for a cranio-caudal distance of 4.3 cm from its origin (Figs. 1–3). Roof top incision was used for access. Cattell–Braasch maneuver was performed, lesser sac was entered and the pancreas was exposed. Common hepatic artery was looped in the supra-colic compartment, whilst SMA was looped in the infra-colic compartment, for vascular control. The left and right hemicircumference of SMA were dissected free off the tumor, in sequence, caudo-cranially, using the infracolic mesenteric approach. A firm, well circumscribed tumor, conforming to the above description, was successfully dissected off its surrounding vital structures, without collateral damage (Fig. 4). Histopathology showed an encapsulated tumor, arranged in fascicles. Tumor cells were spindle shaped with scant eosinophilic cytoplasm and wavy elongated buckled hyperchromatic nuclei with pointed ends (Fig. 5). Interspersed amongst them were numerous ganglion cells, few showing multinucleation (Fig. 6). No neuroblastic elements were seen, confirming the diagnosis of ganglioneuroma. Patient is well on a year's follow up, with no recurrence.

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Figs. 1-3 – CECT abdomen and pelvis in arterial and portal venous phases (sagittal and axial image) showing a well-defined hypodense mass in the retro-peritoneum, posterior to the head of the pancreas and left lobe of liver, wedged between the IVC posteriorly and splenic vein anteriorly. The SMA is embedded within the mass.

Discussion

Ganglioneuromas are benign neurogenic tumors originating from ganglion cells, especially sympathetic ganglia.^{1,2} Sympathetic outflow being thoraco-lumbar, they are common in retroperitoneum (32–52%) and posterior mediastinum (39–43%) followed by cervical region (8–9%).¹ In the abdomen the tumors are either adrenal (41%) or extra-adrenal (59%), of which paravertebral location is commonest, followed by bowel, bladder trigone, gallbladder and abdominal wall – paralleling the distribution of neural crest cells.¹ The paraspinal location predisposes these tumors to closely abut major vessels, but mercifully, not narrow or compromise their lumen.^{1,2} The completeness and length of envelopment of SMA, seen in our case, is very rare.

Unlike pediatric prevalence in other ganglion cell tumors (neuroblastoma/ganglioneuroblastoma) the ganglioneuromas are seen in young adults with slight female predominance.¹ The tumors are sporadic and mostly asymptomatic even when

large, hence, are often incidentally detected on imaging or on post-operative histopathologic examination.³

The symptoms when present are vague, with abdominal pain and palpable lump being the commonest. However, a minority do exhibit symptoms such as diarrhea, virilization and hypertension pertaining to vasoactive intestinal polypeptide, androgen and catecholamine secreted by some tumors.^{1,2} Syndromic association with multiple endocrine neoplasia II and Turner's syndrome is also documented as is their maturation from ganglioneuroblastoma/neuroblastoma.^{1,2}

CT forms the backbone in detection/imaging of these tumors which are typified by their location, well circumscribed spherical/lobulated shape, homogeneous appearance, punctate discrete calcification (in 20%) and delayed heterogeneous contrast uptake.¹ In addition to the diagnosis, CT aids treatment planning, which is vital to prevent collateral damage.

The non-malignant/non-infiltrative nature makes radical resection (en-bloc with surrounding structures) unnecessary.^{1–3} However, the hazardous location of these tumors

Fig. 4 – Operative photo showing the mesenteric approach by which the SMA is being dissected off the tumor.

renders their excision difficult, as evident from our case. Dissection of tumor away from SMA was the crucial step in the surgery. Of the six artery first approaches,⁴ the superior and posterior approaches were not feasible due to 2 reasons – the tumor was enveloping the origin of SMA and it was encasing it all around for >4 cm, hence, the artery could neither be looped at its origin nor could it be dissected in an antegrade manner. The length and completeness of arterial encasement rendered the uncinate and the left posterior approaches impossible. Anterior approach necessitates division of pancreas, which was needless, hence was not adopted. The best approach suited for the tumor was the infracolic mesenteric approach as the SMA in the infracolic compartment was free and could be controlled, easily. Although the tumor was enveloping the artery completely, the predominant tumor bulk was to the right of the artery, hence, the tumor could be dissected away from the artery by sticking close to its adventitia and dissecting its left hemi-circumference free till its origin from the aorta (Fig. 4). Since, there are no branches originating from the left of SMA, till it reaches the infracolic compartment

Fig. 5 – Histopathology picture of ganglioneuroma. Hematoxylin-Eosin stain. 400× magnification.

Fig. 6 – High power view of the tumor.

(Fullen's zone 4), where the first jejunal branches originate, the dissection was safe and bloodless.⁵ Once, the left hemi-circumference of the artery was free, it could be looped at its origin as well, by sticking close to the adventitia. This in addition to the control of the common hepatic artery rendered the entire field bloodless which facilitated further dissection of the tumor away from the right circumference of the SMA.

The prognosis following total excision is excellent with low recurrence, however, long term follow up with imaging is recommended in view of reports of rare malignant transformation.^{1,2}

Summing up, retroperitoneal ganglioneuroma are rare benign tumors with propensity for enveloping major blood vessels. Their resection is challenging, but, adopting the right approach makes it doable. The good prognosis, with low recurrence, makes the effort worthwhile.

Conflicts of interest

The authors have none to declare.

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