Ganglioneuromas are well differentiated benign tumors of the sympathetic nervous system arising from the neuroectodermal cells. Its reported incidence is one per million populations. (1) We report an incidentally detected, histopathologically proven ganglioneuroma of the retroperitoneum in a 24 year old male patient with the preoperative diagnosis of duodenal GIST.

**Keyword**: ganglioneuroma, retroperitoneum, neuroectodermal cells

**CASE REPORT:**
A 24 yr male patient presented with significant loss of weight, episodic upper abdominal pain, nausea & hiccough for 45 days. Physical examination revealed a non tender, smooth, firm mass in right hypochondrium of size 8*6 cm which doesn't moves with respiration and doesn't fall forward. Baseline investigations are within normal limits. Ultrasound showed large exophytic mass arising from second part of duodenum sandwiched between liver and right kidney in the subhepatic region of size 12*9.5 cm with no invasive features or regional lymphnodes. MRI showed well defined iso to hypotense lesion of size 11.5*11.2*8.5 cm exophytically between liver and right kidney in the sub hepatic and precaval regions abuting the second part of duodenum causing mass effect displacing the IVC posteriorly and compressing it displacing the gallbladder, portal vein anterosuperiorly, pancreas towards the midline suggestive of duodenal GIST.

**MRI ABDOMEN**
- CEA-2.2 ng/ml (Ref.range <5ng/ml)
- CA19.9-3.6U/ml (R.range<30.9U/ml)
- CA-125-6.3U/ml(R.range<6.3U/ml)

Upper GI scopy and the colonoscopy are normal. Hypotonic duodenography is done which showed no mucosal irregularity or luminal narrowing.

HYPOTONIC DUODENOGRAPHY
MDCT –abdominal angiogram showed Features suggestive of exophytic growth measuring 11.2*10*8.6cm arising from the second part of the duodenum extending between liver and the right kidney displacing adjacent organs.

**HYPOTONIC DUODENOGRAPHY**

On exploration, retroperitoneal mass of size 15*10*12 cm was found compressing and displacing the IVC posteriorly, gallbladder aand portal vein anterosuperiorly, pancreas and the II part of duodenum laterally and the mass is between liver and the IVC. There was no evidence of invasion of the adjacent organs.

The mass is resected in mass by finer dissection from the surrounding structures. The resected specimen was pale yellow in color with firm in consistency with gritty sensation. The histological examination of the mass revealed a mixture of immature and mature ganglion.
Ganglioneuroma is a tumor of the sympathetic nervous system that arise from the neuroectodermal cells derived from the neural crest cells. Neuroblastoma tend to be aggressive and occur in younger patients(2). Ganglioneuroma occur in older children (2). The most common localization is the posterior mediastinum followed by the retroperitoneum.(3)

Grossly, they are large, encapsulated masses of firm consistency with an homogenous, solid, grayish white cut surface. Areas with different color or consistency should be sampled for microscopic examination with the suspicion of less differentiated foci.(4) They can be multiple and or associated with other independent types of neurogenous neoplasms such as neuroblastoma and pheochromocytoma.(4)

Microscopically, it consists of a spindle cell tumor resembling a neurofibroma but shows numerous ganglion cells. Microscopically ganglioneuromas have consists of a spindle cell tumor composed of neuritic processes, Schwann cells and perineural cells and show numerous ganglion cells.

**HISTOPATHOLOGY:**

Retroperitoneal ganglioneuromas are usually non-functioning and asymptomatic until they reach large sizes in which case they cause symptoms due to local expansion and pressure on adjacent structures. (5) Symptoms of autonomic dysfunctions are seen in patients with hormone secreting ganglioneuromas, may also be seen in patients with paravertebral ganglioneuromas compressing the autonomic fibers of the lumbosacral plexus.(6)

Functioning ganglioneuromas that were found to release peptides such as vasoactive intestinal peptides (VIP), somatostatins and Neuropeptide Y (NPY). These tumors may cause some symptoms like diarrhea(7), sweating and hypertension(8) related to those peptides. Ganglioneuromas may release catecholaminergic peptides, surgeons should be aware of the possibility of hypertensive crisis during the surgery.(9)

Radiological examination also has no diagnostic value in most cases. Because of the rarity of retroperitoneal ganglioneuromas and absence of any characteristic radiologic features, imaging of these tumors is not reliable and diagnostic.(10) Preoperative diagnosis of retroperitoneal ganglioneuroma is often difficult and the diagnosis is usually based on histopathological findings after surgical excision of the tumor.(11,12)

Ganglioneuromas are typically slowly growing, benign tumors and have a tendency to remain clinically silent for a considerable time(13). Most patients have prolonged survival without any evidence of
progression, surgical excision is sufficient for the treatment.(14)

Chemotherapy or Radiotherapy have no value in the treatment except it was associated with ganglioneuroblastoma changes when there might be some role of chemotherapy.(15)

CONCLUSION

Ganglioneuromas are rare benign tumours arising from sympathetic ganglion and affects children more often than adults. It is a slow growing tumor and preoperative diagnosis is difficult due to lack of specific radiological findings. As it is a slow growing tumor, total surgical excision with preservation of organ functions is a feasible surgical option

BIBLIOGRAPHY


