Abstract:
Ganglioneuromas are benign tumors of the sympathetic nervous system that rarely arise in the adrenal gland. Majority of cases are detected incidentally since they are usually asymptomatic. Ganglioneuromas are commonly seen in childhood. They are highly differentiated benign tumors and are compatible with long-term disease free survival. Retropertitoneal localization is relatively frequent for these tumors. Case Report - A 22 year old female with history of right loin pain since 4 months, on physical examination patient general condition good, mass palpable 6 cm below the right costal margin. CECT and MRI showed 14X11 cm right adrenal mass with inferior displacement of right kidney and encasement of surrounding vascular structures, preoperative usg guided biopsy suggestive of ganglioneuroma. Exploratory laparotomy was done and right adrenal tumour excision done. Post operative histopathologic examination again consistent with ganglioneuroma. Conclusions - Ganglioneuroma occurs rarely in adrenal gland and preoperative diagnosis is difficult since symptoms are usually nonspecific. Due to widespread utilization of abdominal imaging, however, it should be included in differential diagnosis of adrenal or retroperitoneal mass. Histopathologic examination is currently the mainstay of diagnosis.

Keyword: Ganglioneuroma, retroperitoneum, adrenal gland, excision

Introduction
Ganglioneuromas are tumors of the sympathetic nervous system that arise from the neuroectodermal cells. Neuroblastoma, ganglioneuroblastomas and ganglioneuromas are tumours within the spectrum of its disease continuum. Ganglioneuromas are fully differentiated benign tumours. The reported incidence is one per million population. The most common localization is the posterior mediastinum followed by the retroperitoneal space. Although catecholamine synthesis is an almost constant feature of all the neurogenic tumors, ganglioneuromas rarely lead to symptoms. Currently,
histopathologic examination is the only tool to diagnose ganglioneuroma and to differentiate it from other neural crest tumors.

Case report

We report an incidental detection of retroperitoneal tumour in a 22 year old female who presented to the urology department with the complaint of right loin pain since four months, dull aching, not radiating, G C good, afebrile, vitals: stable, P/Absence-soft, mass palpable 6 cm below the right costal margin, firm, nontender. Her laboratory investigations for complete blood count, serum electrolytes and urine were normal. Plain CT scan of the abdomen showed a heterogenous mass in the region of the right adrenal gland extending below the inferior vena cava with areas of calcification as shown in fig.1. Contrast CT showed adrenal mass with anterior displacement of Inferior vena cava as shown in fig.2. MRI of abdomen also done to further characterize the lesion showed inferior displacement of right kidney as shown in fig.3. A clinico-radiological diagnosis of adrenal tumor was made. Preoperative endocrine evaluation was within normal limits. Preoperative Ultrasound (USG) guided needle biopsy was done and Microscopic examination revealed Neoplasm composed of spindle shaped cells arranged in sheets, short fascicles and with cells having serpentine nuclei admixed with ganglion cells suggestive of ganglioneuroma as shown in fig.4. Immuno-histochemistry showed neuron specific enolase: positive in ganglion cells, S-100: positive in neuronal cells, Vimentin: positive in neuronal cells - features consistent with ganglioneuroma. The patient underwent exploratory laparotomy for removal of the tumour. Intraoperatively, the tumors were found encasing right renal hilar vessels, IVC, extending to the left renal hilum and was difficult to separate off the surrounding structures and in the process of adrenal mass excision right renal pedicle was inseparable and hence proceeded with right nephrectomy further tumour dissection done and the entire adrenal mass was excised as depicted in FIG 5. Grossly the tumor measured 14*10 cm. It was capsulated, firm in consistency with a homogenous, solid, grayish white cut surface and gritty sensation. fig 6A, FIG 6B. Histopathologic examination after a complete excision was again consistent with the diagnosis of ganglioneuroma.
Ganglioneuromas (GNs) are extremely rare benign neuroectodermal neoplasms that are composed of ganglion and Schwann cells. 21% of cases arose from the adrenal gland itself. Other sites of origin included the mediastinum (39%), retroperitoneum (31%), and isolated cases in the pelvis and neck. (Enzinger and Weiss, 1995) Predilection for the young, with only 20% of cases affecting individuals over age 40, and some 50% of cases found in patients between 10 and 29 years. Females are more prone to be affected than males. Clinical symptoms of ganglioneuromas are non-specific, mostly hormonally silent and related to their size/locations. Despite their generally benign nature, GNs may come to attention due to compression of their neighboring structures. Approximately up to 30% of patients were found to have elevated plasma and urinary catecholamine but they rarely develop symptoms of vasoactive amines excess. Immature element such as neuroblast is not part of mature GNs explaining lower rate of metaiodobenzylguanidine (MIBG) uptake (57%) compared to neuroblastoma (92%).

Ganglioneuromas are benign, but isolated case reports of ganglioneuromas undergoing malignant transformation are reported. Tumors can grow extremely large and have a propensity to encase vessels without impinging on the vessel lumen (Radin et al, 1997). Metabolically silent adrenal mass with unenhanced attenuation of less than 40 HU and presence of stippled calcifications should raise suspicion for a ganglioneuroma (Maweja et al, 2007).
Diagnosis of ganglioneuroma is nearly always made upon resection, natural history of unresected ganglioneuromas has not been clearly defined. Histopathologic examination is currently the mainstay of diagnosis. Diagnosis of GN should be suggested when an adrenal tumor harbors the following: (1) no hormonal hypersecretion, (2) presence of punctate or discrete calcifications, (3) absence of vessel involvement, and (4) a low non-enhanced T1-weighted signal with a late and gradual enhancement on dynamic magnetic resonance imaging (MRI). The treatment for this condition is complete surgical resection through either an open or laparoscopic approach. The most significant differential diagnosis of ganglioneuroma is neuroblastoma. Increased level of urinary noradrenaline, dopamine, HVA, and VMA are frequently encountered in neuroblastoma, while the level of urinary catecholamine, HVA, and VMA are usually normal in ganglioneuroma. Ganglioneuroma occurs rarely in adrenal gland and preoperative diagnosis is difficult since symptoms vary and often times nonspecific. Due to widespread utilization of abdominal ultrasonography, CT scan and MRI, detection of such tumor has increased, therefore differential diagnosis of an adrenal/retroperitoneal mass should include ganglioneuroma. However, the pre operative diagnosis of ganglioneuroma is difficult. Therefore ganglioneuroma is always a histopathological diagnosis.

REFERENCES: