Abstract:
We report a rare case of ganglioneuroma causing dorsal compressive myelopathy. A 6 years old male child presented with progressive paraparesis of 2 months duration. Child had a swelling in the lower dorsal region of the back for past 2 years. Child had dull midback pain for the past one year which was not disturbing the child's routine activities. Investigation with magnetic resonance imaging (MRI) dorsal spine and whole spine screening showed posterior mediastinal and retroperitoneal paravertebral mass in the left lower dorsal level, extending into the spinal canal with compression of the cord. Patient underwent surgical excision of the mass and histopathological examination (HPE) revealed ganglioneuroma.

Keyword:
Compressive myelopathy, ganglioneuroma

Introduction:
Ganglioneuroma (GN) is the uncommon, benign representative of the peripheral neuroblastic tumours (PNTs) that occur prevalently in childhood.[1] The International Neuroblastoma Pathology Committee (INPC) criteria[2] defines GN as a schwannian stroma-dominant tumor, predominantly composed of ganglioneuromatous stroma with a minor component of scattered collections of differentiating neuroblasts and/or maturing or mature ganglion cells. De novo GN differs from malignant PNTs in that it affects older children, produces normal or slightly elevated amounts of catecholamines[3,4] and is often asymptomatic.[3,5,6] Symptoms result from the compressive effect of the tumor on neighbouring tissues.[3,5]

Abdominal neurogenic tumours are derived from immature cells of the sympathetic nervous system, most commonly located in the retroperitoneum, especially in the paraspinal areas and adrenal glands.[7]

Case report:
A six year old male child was admitted with complaints of weakness
of both lower limbs for the past 2 months duration. Child had dull midback pain for the past one year which was not disturbing the child’s routine activities. Since 2 months child had difficulty in holding the chappals, buckling of knees and difficulty in getting up from squatting position. There was no weakness of neck, upper limbs and trunk. History did not suggest bladder & bowel involvement. No history suggestive of trauma, fever, meningeal irritation or tuberculosis.

On examination he was well built. No neurocutaneous markers. A non tender, not warm, diffuse, firm swelling of 8 x 6 cms was seen in the back at the left thoracic region. Child had minimal tenderness over the swelling. His higher mental functions and cranial nerve examination were normal. Examination of the spinomotor system showed normal bulk, tone and power of the upper limbs. Lower limb hypertonic bilaterally. Proximal upper limb deep tendon reflexes were normal. Knee jerk and ankle jerk were exaggerated bilaterally. Abdominal reflex was absent in the lower quadrants on either side with bilateral plantar extensor. Sensory system was normal. Rest of the examination was not contributory. The power was 3/5 and distally 0/5.

Figure 1: X-ray dorsal spine oblique view showing erosion of vertebral bodies (arrows)

Figure 2a: T1WI sagittal

Figure 2b: T1WI post contrast coronal

Figure 2c: T1WI post contrast axial
Figure 2a – 2f: MRI Dorsal spine with whole spine screening shows large enhancing solid heterogeneously intense lesion involving the left paravertebral region, extending from D6 – D12 level, with extension into spinal canal from D8 – D12 level, with indentation over cord and displacement of cord to right. MR myelogram shows block at these level. His complete haemogram, chest x-ray, renal parameters, electrocardiogram and catecholamine levels were normal. Imaging of brain, chest and pelvis was normal. Patient was assessed and excision of the lesion was carried out. Under ETGA, prone position, left paravertebral curvilinear incision extending over D6 – D12 was marked. Skin and posterior abdominal muscles opened. D8 – D12 laminectomy was performed. The mass was seen encroaching into spinal canal through vertebral foramen from left side causing compression of the cord at the level of D8 – D12. 9,10,11,12 ribs were excised and the mass was well delineated in the retroperitoneum. It was neither adherent to major vessels nor renals. The mass was firm, nonsuckable, moderately vascular, greyish in colour. Total excision of the mass was carried out and sent for histopathological examination which revealed ganglioneuroma.
Post operatively the patient showed improvement of the motor weakness over a period of one month with lower limb power of 3/5 proximally and 4/5 distally.

Figure 3: Lesion visible after opening the posterior abdominal wall muscles.

Figure 4a: Macroscopic appearance - Multiple grey white tissues 6 (maximum) x 2 x 1cm with evidence of encapsulation. Cut section shows glistening, fleshy whitish tissues with yellowish areas. Some foci were hard to cut.

Figure 4b: Microscopic appearance - Section shows a neoplasm composed of nests of mature ganglion cells and neurons separated by schwannian rich stroma. Occasional focus shows primitive neuroblasts and calcification. Impression: Ganglioneuroma-maturing type.

Figure 5: Post operative CT showing complete excise on of tumour.

Discussion:
Ganglioneuromas are tumours of the sympathetic nervous system that originate from neural crest. This benign tumour usually occurs in childhood, grows slowly and there are no known risk factors. When tumour is in the retroperitoneum or posterior mediastinal region, may cause compression of the spinal cord or spinal deformity. Common locations for ganglioneuromas include the adrenal gland, paraspinal retroperitoneum, posterior mediastinum, head and neck. Preoperative diagnosis of retroperitoneal ganglioneuromas is often difficult and the diagnosis is usually based on histopathological findings after surgical excision of the tumour.

Magnetic resonance imaging and computed tomography scanning are the preferred methods for imaging ganglioneuromas. MRI is the modality of choice for evaluating the extension of spinal tumours. They appear as well-circumscribed, smooth or lobulated masses that may contain calcifications. Rib and vertebral foraminal erosions, increased intercostal spaces and vertebral body pedicle erosions may be seen with both retroperitoneal and posterior mediastinal tumours. Since GN can tightly adhere to, or encase major vascular structures, attempting
resection may lead to severe, even life-threatening complications. Although the potential risks of operating on a GN are well known, reports on surgery-related complications, including blindness and neurological dysfunctions, are limited to few single case reports. [1]

Tumor resection remains an option for these patients, considering that it may lead to serious complications when performed in presence of imaging-defined risk factors. We believe that the benign nature of GN requires a more cautious surgical approach when surgical risk factors are identified. In such instances a watchful waiting policy should be established, with debulking surgery becoming justified when excessive growth has occurred or significant disturbances have arisen.

Surgery should be performed if the symptoms are due to tumour encroachment of vertebral foramina. [8] Most ganglioneuromas are benign and are compatible with long-term, disease-free survival with surgical excision. [9]

References: