CASE REPORT

Retroperitoneal Ganglioneuroma (GN): Case report in a 3 year old boy

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ABSTRACT

Background: Neuroblastic tumors are the most common extra-cranial solid tumors in childhood. They arise from the neural crest cells. The most common sites of presentation are the posterior mediastinum, retroperitoneum, head and neck region. GN are usually asymptomatic and are found incidentally on abdominal imaging. The treatment is complete surgical excision. **Case Report:** A 3 year old boy presented to the surgical OPD with pain abdomen. There was a palpable lump present in the left hypochondrium& umbilical region. CT images revealed a soft tissue mass lesion in the left para-spinal region of the retroperitoneum abutting the abdominal aorta, vertebral column, psoas muscle and left kidney. A pre surgical biopsy revealed a benign ganglioneuroma. Total resection of a 6 X 5 X 6 cm sized tumor was obtained via exploratory laparotomy. Histopathological analysis confirmed the diagnosis. **Conclusion:** Ganglioneuroma (GN) is a rare benign tumor, usually asymptomatic. One of the most common site of presentation is retroperitoneum, the main treatment is complete surgical excision.

Key Word: Ganglioneuroma(GN); Abdominal Aorta ; Retroperitoneum.

INTRODUCTION

Ganglioneuromas (GN) and ganglioneuroblastomas (GNB) are tumors of the sympathetic nervous system that originate from neural crest sympathogonia, which are completely undifferentiated cells of the sympathetic nervous system. Along with neuroblastomas, ganglioneuromas and ganglioneuroblastomas are collectively known as neuroblastic or neurogenic tumors. [1, 2]

Ganglioneuromas often present as a solitary, painless, slow-growing mass consisting of ganglion cells, Schwann cells, and fibrous tissue. The most commonly affected sites are the posterior mediastinum (41%), retroperitoneum (37%), adrenal gland (21%), and neck (8%). [3]

Ganglioneuroblastoma is a transitional tumor on the intermediate spectrum of disease between ganglioneuromas and neuroblastomas, containing elements of both malignant neuroblastoma and benign ganglioneuroma. [4] They are most common in children, with a median age at diagnosis of 22 months; most cases are diagnosed before 10 years of age. Although cases of adolescent or adult-onset GNB have been reported, they are extremely rare. [5]

These tumors occur most frequently in the abdomen, but they can grow wherever sympathetic nervous tissue is found. Common locations for ganglioneuromas and ganglioneuroblastomas include the adrenal gland, paraspinal retroperitoneum (sympathetic ganglia), posterior mediastinum, head, and neck; it is uncommon to find them in the urinary bladder, bowel wall, abdominal wall, and gallbladder. GNs are found incidentally in most cases and manifest as asymptomatic masses. The tumor could cause some complications if it becomes large enough to press against the adjacent organs. [6]

Magnetic resonance imaging (MRI) and computed tomography (CT) scanning are the preferred methods for imaging ganglioneuromas and ganglioneuroblastomas. [7, 8, 9] MRI is the modality of choice for evaluating the extension of spinal tumors. [10, 11, 3, 12, 13, 14, 15]

CT scanning is the imaging modality that is most commonly used to evaluate neuroblastic tumors. It has proven to be the superior imaging technique when identifying tumor size, organ of origin, tissue invasion, vascular encasement, adenopathy, and calcifications. Newly diagnosed cases are evaluated with standard chest, abdominal, and pelvic CT scans. [10]

In general, neuroblastic or neurogenic tumors appear radiologically as well-circumscribed, smooth or

lobulated masses that may contain calcifications. The benign (ganglioneuromas) and malignant (ganglioneuroblastomas) forms of these tumors are virtually identical radiologically. The only differentiating factor is the possibility of distant metastases with malignant ganglioneuroblastomas.

PRESENTATION OF CASE

A 3 year old boy not known to have any medical illness, presented to the Surgical out-patient department of Sharda Hospital with complaints of pain in the left side of the upper abdomen for the last 6 months .

Abdominal examination revealed a palpable swelling in the left hypochondrium and umbilical region 5X4cm, ovoid in shape, well defined margins, firm in consistency and non-tender. Systemic examination was unremarkable.

Ultrasound whole abdomen (Fig 1) revealed an ill-defined heterogenous lesion measuring 6X5 cm in the retroperitoneum in the left lumbar region, paravertebral location medial to left kidney.

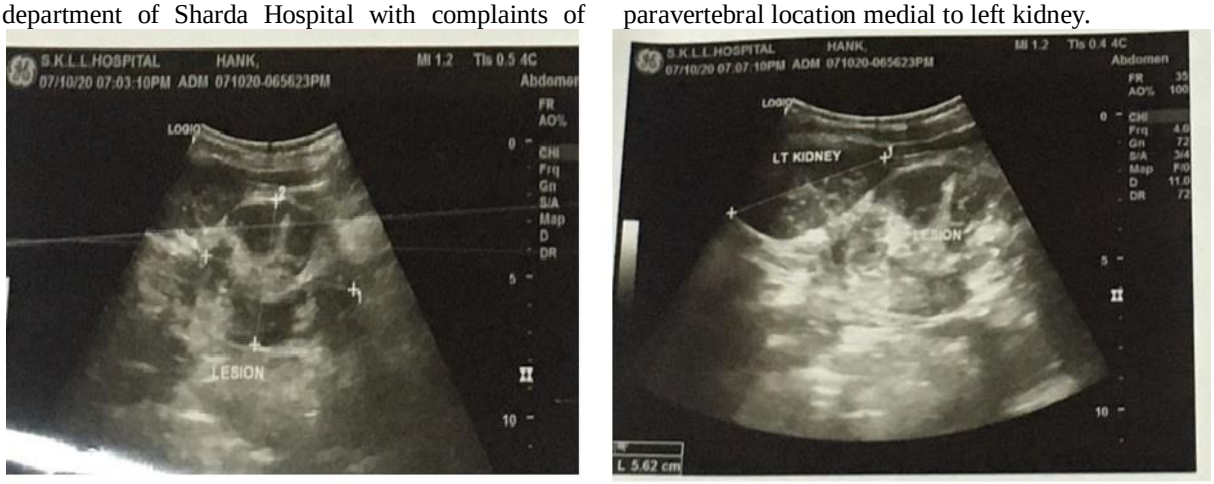


Fig 1: USG whole abdomen - lesion in retroperitoneum.

CECT whole abdomen (Fig 2) revealed a well-defined heterogeneously enhancing oval, soft tissue mass lesion measuring~ 6X 5.1 X 6 cm in the left para-spinal region in the retroperitoneum, which was showing central linear/ branching calcification. Posteromedially the mass was abutting the abdominal aorta, vertebral column, psoas muscle and left kidney. Anterolaterally the mass was displacing bowel loops.

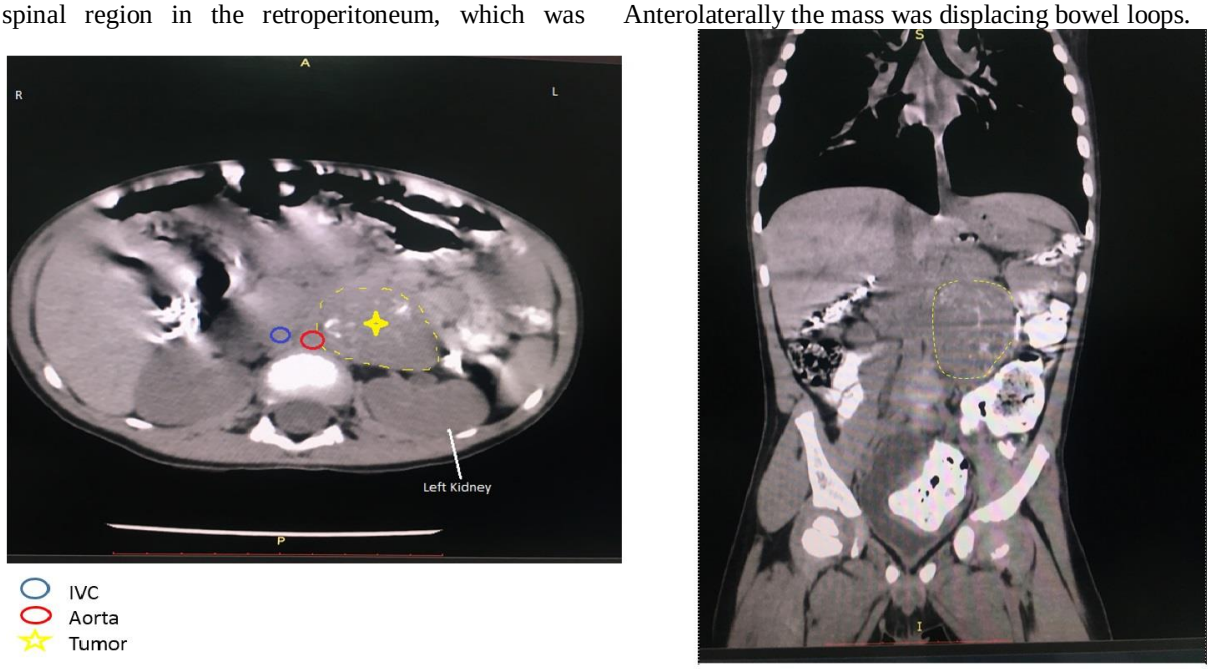


Fig 2: CECT whole abdomen – soft tissue mass in left para-spinal region, abutting abdominal aorta, psoas muscle, vertebral column, left kidney

Pre surgical CT guided biopsy (Fig 3) revealed - ganglioneuroma.

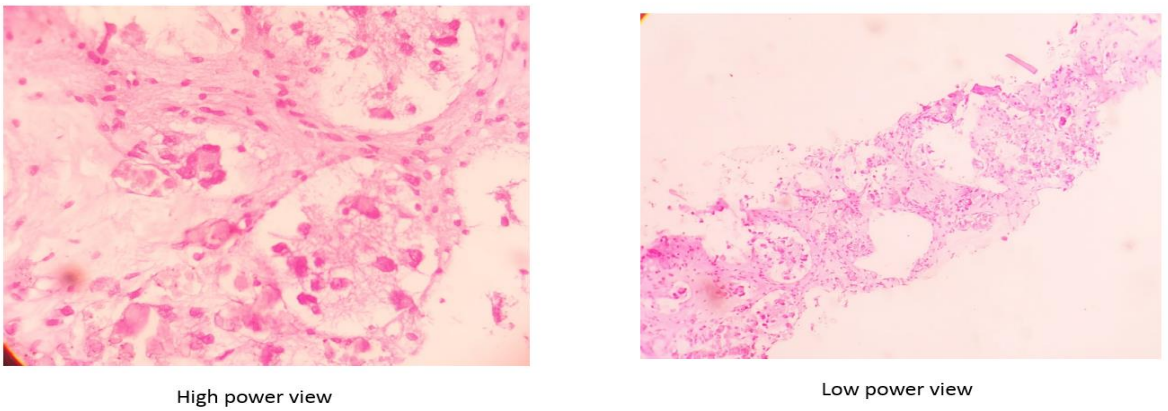


Fig 3: Pre Surgical Biopsy

Surgical management was thus considered and exploratory laparotomy with excision performed. Intra operatively (Fig 4) an encapsulated mass measuring ~ 6 X 5 X 6 cm in size, adhered to the psoas muscle, abutting the abdominal aorta and vertebral column was carefully dissected using blunt dissection and electrocautery.

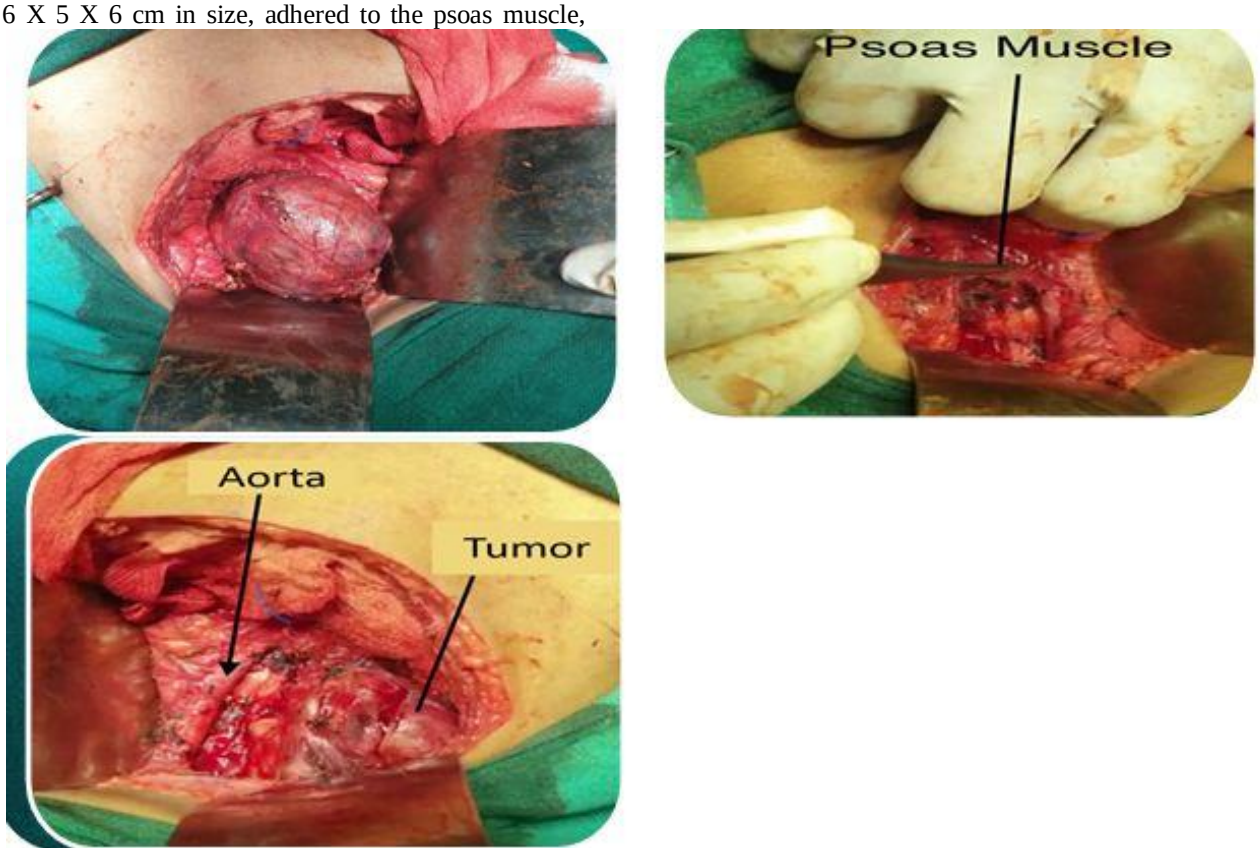
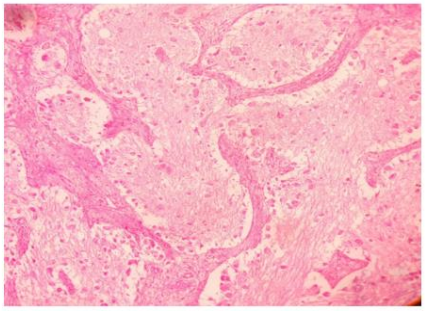
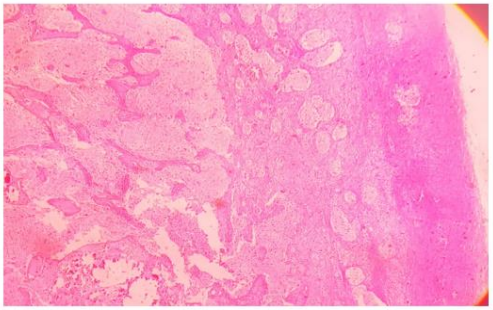


Fig 4: Intra Op Finding – Tumor abutting abdominal aorta, encapsulated mass of 6 X 5 X 6 cm excised

Histopathology (Fig 5) – Ganglioneuroblastoma intermixed (Favourable histology), margins uninvolved by tumor, no lymphovascular invasion seen.



High Power view showing tumor cells in lobules with abundant granular eosinophilic cytoplasm, distinct cell borders, nuclear enlargement, eccentric nuclei and prominent nucleoli. (H&e, 40x)



Low Power view showing circumscribed tumor in lobules with tumor cells showing abundant granular eosinophilic cytoplasm, distinct cell borders, nuclear enlargement, eccentric nuclei and prominent nucleoli. (H&e, 10x)

Fig 5: Post Op Histology

DISCUSSION

Retroperitoneal GNs account for 37.5% of all GNs and about 0.72%-1.6% of primary retroperitoneal tumors [16,17,18]. GNs can also occur in the vertebra, neck, and cerebellopontine angle region (trigeminal); all three of which are relatively rare [19-22]. GNs are found incidentally in most cases and manifest as asymptomatic masses [17,24,25]. The tumor could cause some complications if it becomes large enough to press against the adjacent organs. Occasionally, GNs occurring in the adrenal gland can secrete vasoactive intestinal peptides, dopamine, and cortisol, which lead to diarrhea, hypertensive crisis, and male-like metabolic disorders in women [26-28].

GNs are mainly composed of ganglion cells, mucus matrix, nerve fibers, and mature Schwann cells, and the first two of them are characteristic components in histopathology [29,30]. The pathological features of GN are closely related to its CT findings. The presence of mucus matrix in tumors determines the hypodensity on plain CT scans. The mucus matrix has been found to delay the absorption of contrast agents, which leads to the delayed enhancement of GNs[31,32]. Calcifications have been noted in 20%-60% of GNs, and most of them are punctate, which is one of the differences between GN and neuroblastoma [34,35]. Ko et al[29] and Duffy et al[36] suggested that the presence of fat components in GN may be one of the characteristics of GN, but their sample size was too small to verify this. Some scholars have proposed that the blood vessels are often surrounded or compressed by GNs instead of being invaded, although most of them are small vessels[31,37], and this finding further suggested that GNs are benign. However, there are some reports suggesting that GNs could behave aggressively, and recurrence or malignant transformation [17,38,39] and complete surgical excision are the most optimal choice for the treatment[23,40]. Therefore, patients with GN still need long-term radiological follow-up. In addition, some scholars have put forward a different view, arguing that incomplete resection of GN does not increase the risk of progression if residual tumors are less than 2 cm in diameter[40].

In conclusion, GNs appear as hypodense masses on plain scans and present delayed and mild enhancement on contrast enhancement. Pathology is the gold standard for the diagnosis of GN, andganglion cells are their important features. Surgical excision is the best treatment for GN, and postoperative radiotherapy and chemotherapy are unnecessary. However, in cases where it is difficult to completely dissect the tumors and blood vessels, partial resection could still relieve the pressure symptoms created by the tumors. Long-term radiological follow-up after the operation is necessary, even though the biological behavior of GN is benign, because there is still a tendency to be malignant, especially in patients with GN who have undergone only local resection.

CONCLUSION

Ganglioneuroma (GN) is a rare benign tumor, usually asymptomatic. One of the most common site of presentation is retroperitoneum, the main treatment for that is complete surgical excision. Regular follow-up is needed due to late recurrence.

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