

Retroperitoneal Ganglioneuroma Masquerading As Renal Mass – A Case Report

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Abstract: Ganglioneuroma (GN) is a rare tumor of neuro ectodermal origin, commonly occurring at the retroperitoneum in childhood. Due to lack of radiological specificity most of the cases are diagnosed post operatively on histopathological study only. So it's important to keep it in differential diagnosis. Overall survival is good in these group of tumors.

Keywords : ganglioneuroma, neuro ectoderm, retroperitoneum

I. Introduction

Ganglioneuroma are fully differentiated benign tumors of the sympathetic nervous system arising from the neuroectodermal cells¹. These are benign tumors found in older individuals; the median age at diagnosis ranges from 5.5 to 10 years; with a slight female predominance (1.5:1). The most common locations are the posterior mediastinum (41%), retroperitoneum (37%), adrenal gland (21%), and neck (8%). Although catecholamine synthesis is an almost constant feature of all the neurogenic tumours, GN rarely led to symptoms¹.

II. Case report

A six year old male child presented with history of dull aching pain abdomen since one month with acute onset nausea and non-projectile vomiting for 8 days along with decreased appetite. He is a full term baby by birth with no significant past history and a healthy younger female sibling. The general and systemic examination of the patient revealed no abnormality except a non-tender mass in left flank. His complete blood count, serum electrolytes, urinary VMA and routine urine examination were normal. On ultrasonography and CT scan revealed a large well defined lobulated retroperitoneal lesion at the left renal hilar region. Patient underwent radical nephrectomy and specimen was submitted for histopathological examination.

Grossly left kidney showed a growth involving the middle pole and part of upper pole of the kidney measuring 6.6 x 5.5 cm which was encapsulated, greyish white, solid & homogenous with adjacent normal renal parenchyma. Microscopy revealed interlacing fascicles of Schwann cells and mature ganglion cells of variable size with fine granular eosinophilic to brown cytoplasm, prominent eccentrically placed nucleoli. There was no evidence of mitosis, necrosis and atypia. Immunohistochemistry was done which showed positivity with chromogranin, S100, NSE & desmin negativity. Based on histopathological features diagnosis of ganglioneuroma was rendered.

III. Discussion

GN represent the fully differentiated members of the group of tumors arising from sympathetic nervous system and are invariably benign². Reported incidence is one in a million³. They are most commonly asymptomatic & common in childhood. Rarely, they are associated with hypertension, watery diarrhea, and hypokalemia or masculinization^{4,5,6}. Metabolically active tumors secrete VMA, homovanillic acid (HVA), vasoactive intestinal peptide (VIP) or serotonin which may cause symptoms related to it. They are sometimes incidentally found when they compress the adjacent structure and causes symptoms. They are only rarely found in the adrenal gland, their most common location being the posterior mediastinum and retroperitoneum². As most of the tumors are clinically silent and radiological investigation has limited role in diagnosis, it causes pre-operative diagnostic dilemma for most of the clinicians. Review of literature also suggest that most diagnose it as a retroperitoneal tumor presenting as renal mass. Still CT is the most commonly used imaging modality for assessment because it reveals the extent of the tumor, organ of origin, regional invasion, vascular encasement, adenopathy, and calcification. Interestingly, GNs tend to partially or completely surround major blood vessels, with little or no compromise of the lumen⁷. Grossly, they are large, encapsulated masses of firm consistency with a homogeneous, solid, greyish white cut surface having a focally edematous appearance. Microscopically,

mature subtype contains varying numbers of mature ganglion cells and Schwann cells together with variable amounts of collagen⁸. They are immunohistochemically positive for chromgranin, S100, NSE and synaptophysin. In the International Neuroblastoma Pathology Committee (INPC) classification, GN is subdivided into GN, maturing and GN, mature subtypes^{9,10}. The GN, maturing subtype was previously classified as “stroma-rich, well-differentiated NT” in the original Shimada classification¹¹. Surgical excision of GN, mature subtype is usually curative. Very rarely, GN, mature subtype may transform into malignant peripheral nerve sheath tumor either spontaneously or after irradiation for neuroblastoma or ganglioneuroblastoma¹². Usually prognosis is good. But local recurrence has been reported, so periodic radiologic surveillance is performed after resection.

Figures and Tables

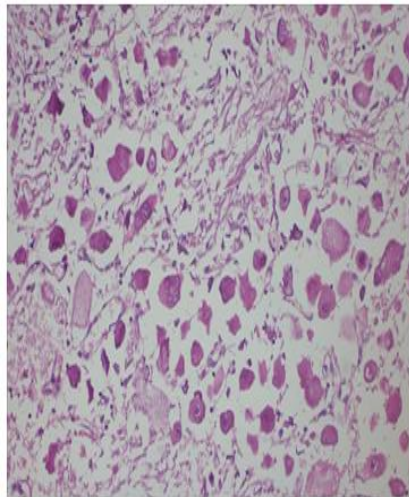


Fig: H&E stain 100X

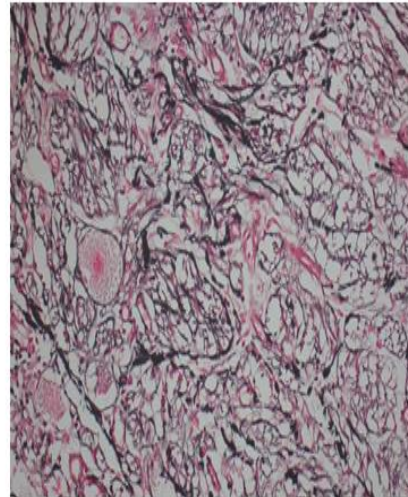


Fig: JMS stain 200 X

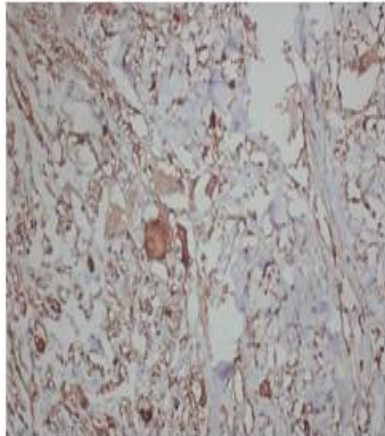


Fig: NSE IHC stain, 100x

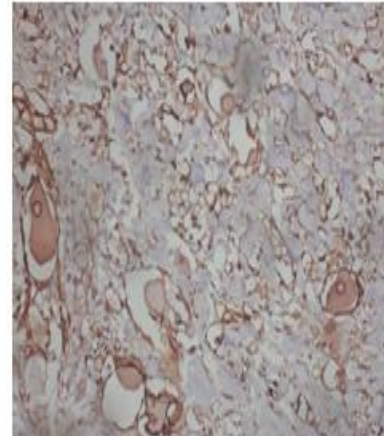


Fig: S100 IHC stain, 200 x

III. Conclusion

Though it is a rare entity, one must consider it is under the differential diagnosis. Clinician should be aware of the fact that most of these tumors may be symptomless and difficult to diagnose under radiological investigation. Histopathology is still the gold standard in diagnosis and overall prognosis is good.

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