Adrenal Ganglioneuroma

Kayastha R, Acharya R, Pradhan S, Tuladhar AS, Shrestha A

Department of Radiology

Nepal Medical College Teaching Hospital,

Kathmandu, Nepal.

Corresponding Author

Rumita Kayastha

Department of Radiology

Nepal Medical College Teaching Hospital,

Kathmandu, Nepal.

E-mail: rumikayastha@gmail.com

Citation

Kayastha R, Acharya R, Pradhan S, Tuladhar AS, Shrestha A. Adrenal Ganglioneuroma. *Kathmandu Univ Med J.* 2020;71(3):316-9.

ABSTRACT

Adrenal ganglioneuromas are rare sympathetic differentiated tumors which originate from neural crest cells. These lesions are usually discovered incidentally on imaging and tend to be hormonally silent. Preoperative diagnosis of adrenal ganglioneuroma remains extremely challenging and the gold standard treatment is adrenalectomy. There is good prognosis after surgery without recurrence. We herein report a case of adrenal ganglioneuroma in a 15 year old female who presented with complaint of abdominal discomfort. Contrast Enhanced Computed Tomography abdomen showed a large septated hypodense right suprarenal mass which was echogenic on Ultrasonography. It showed T1 hypointense and T2 hyperintense signal on Magnetic Resonance Imaging of abdomen and pelvis. Excisional biopsy and histological examination of the mass was suggestive of adrenal ganglioneuroma. This report presents the clinical and radiological data for the rare tumor which would share some experience to facililate the diagnosis of adrenal ganglioneuroma.

KEY WORDS

Adrenal mass, Computed tomography, Ganglioneuroma, Magnetic resonance imaging, Ultrasonography

INTRODUCTION

Ganglioneuromas (GNs) are rare, benign neurogenic tumors that arise from sympathetic ganglia with an excellent prognosis.[1] The tumors are composed of mature Schwann cells, ganglion cells, and nerve fibers.[1] It predominantly affects children and young adults (42%-60%).[2] The median age at diagnosis is 7 years with slight female predominance.[2] The most common locations are the posterior mediastinum (41.5%), retroperitoneum (37.5%), adrenal gland (21%), neck (8%).[3] Unusual sites include the spermatic cord, heart, bone, and intestine.[3]

On Ultrasonography (USG), it appears as a well-circumscribed, homogenously hypo to echogenic suprarenal lesion.[4] It appears as well-defined, solid, lobulated iso to hypo-attenuating suprarenal mass (compared to muscles) on Computed Tomography (CT).[4,5] Fine, punctate calcifications are seen in approximately 20% to 69% of cases.[6] On magnetic resonance imaging (MRI), it has low or intermediate T1 signal, heterogeneously intermediate or high T2 signal with progressive enhancement on post contrast study.[7,8]

CASE REPORT

A 15 year old female presented to Nepal Medical College Teaching Hospital with a chief complaint of abdominal discomfort in right lumbar region for 3 months. General and abdominal examinations were normal. Ultrasonography showed a large well defined echogenic mass in the right upper quadrant on suprarenal region without calcification and minimal vascularity on Color Doppler study. Contrast enhanced CT (CECT) showed a large well defined oval shaped iso to hypodense right suprarenal mass measuring 11.4 x 11 x 10 cm (Volume = 665 cc) without evidence of calcifications. It showed patchy areas of enhancement on delayed phase after administration of intravenous contrast with few enhancing septations within the mass. The mass was extending from T9 to L1 vertebral level. Superiorly, it was abutting the inferior surface of liver and it seemed to be insinuating between the IVC and right crus of diaphragm, abutting and displacing the inferior venacava anteriorly. Posteriorly it was abutting the posterior abdominal wall and posterior portion of 9th, 10th and 11th rib. It was also

abutting the pyloric region of the stomach anteroinferiorly. Medially it was abutting the vertebra. Fat plane was maintained with above mentioned adjacent structures. Right adrenal gland was not separately visualized. It was supplied by a direct branch arising from the left side of abdominal aorta just distal to origin of superior mesenteric artery. Multiple small blood vessels (branches of aberrant artery) were noted within the mass. The inferior adrenal artery arising from right main renal artery was noted along the inferior margin of the mass. There was no evidence of abnormal compression of these vessels. There was no obvious communication of the lesion with neural foramina. There was no abdominal lymphadenopathy. The mass was evaluated to be "non adenoma" and was reported as large adrenal mass with differentials of adrenal lymphangioma, ganglioneuroma and a rare possibility of nerve sheath tumors.

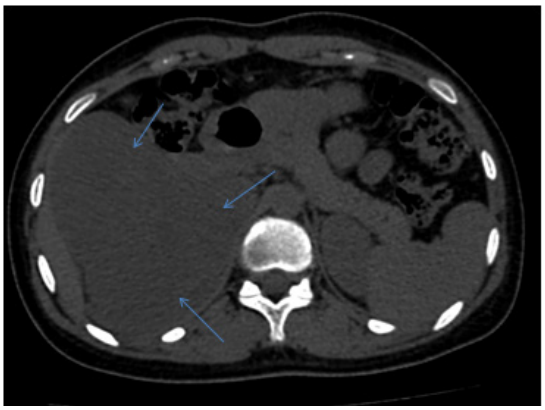

Figure 1. CECT abdomen, axial plain section showing an insinuating right suprarenal iso to hypodense mass without any calcifications.

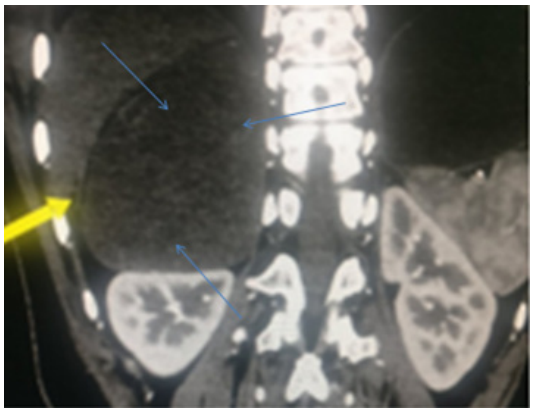

Figure 2. CECT abdomen, coronal section showing right adrenal mass with patchy delayed enhancement and few septa.

Plain MRI of abdomen and pelvis was done which showed well defined oval shaped T1 hypointense right suprarenal mass with mass effect; compressing the right kidney inferiorly. The mass was hyperintense on T2 weighted sequence. Fat suppressed T2 weighted sequence showed hyperintensity within the mass without fat suppression.

Surgery was performed and the mass was excised and send for histopathological examination. The histopathological features were consistent with ganglioneuroma.

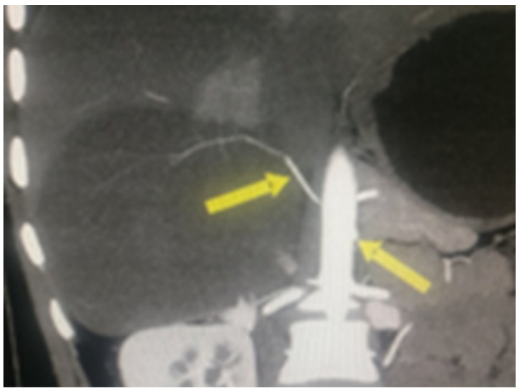

Figure 3. CECT abdomen, coronal section showing aberrant artery supplying the right adrenal mass without any attenuation or filling defect.

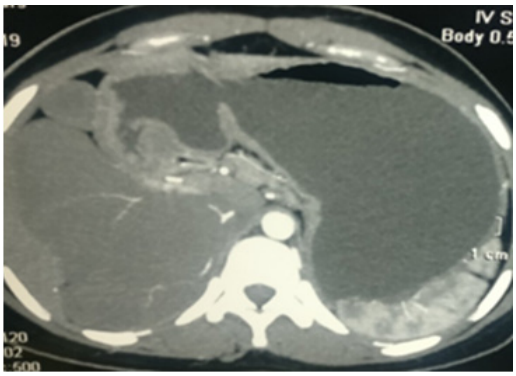

Figure 4. CECT abdomen, axial section showing vessels within the mass without any attenuation and filling defects.

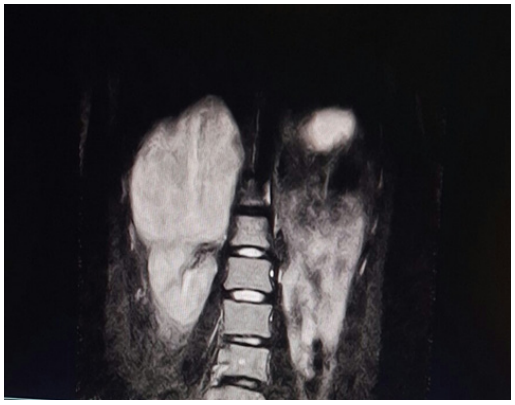

Figure 5. MRI showing T2 weighted coronal image showing hyperintense right suprarenal mass compressing the right kidney.

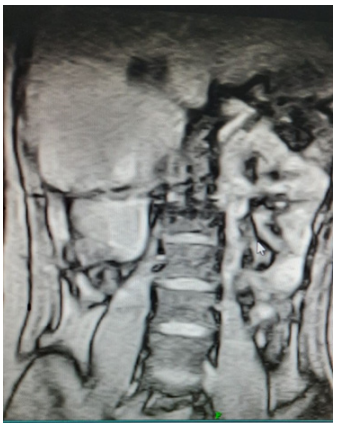

Figure 6. MRI showing Fat suppressed T1 weighted coronal image showing hyperintense right suprarenal mass.

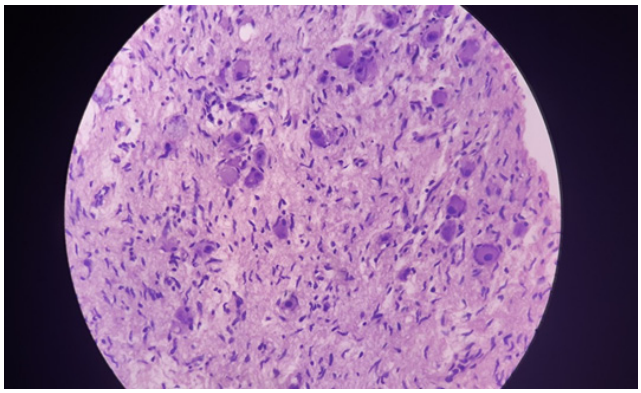

Figure 7. Section showing well circumscribed, encapsulated tumor comprising of nerve cells, revealing wavy nuclei with bilateral pointed ends and fibrillary cytoplasm arranged diffusely.

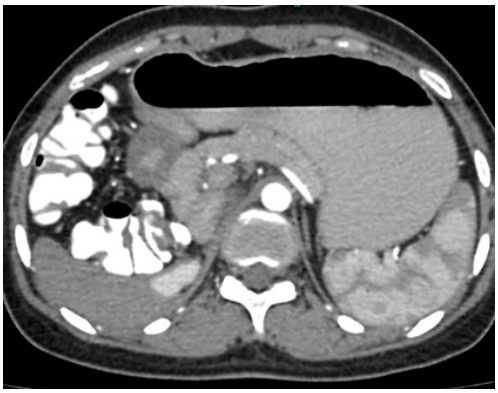

Figure 8. There is no evidence of any adrenal mass in postoperative CECT abdomen axial section.

MICROSCOPIC FINDINGS

Section examined showed a well circumscribed, encapsulated tumor comprising of nerve cells, revealing wavy nuclei with bilateral pointed ends and fibrillary cytoplasm arranged diffusely. At few areas these cells were admixed with plump fibroblastic cells arranged in bundles, interlacing fascicles and occasional storiform pattern. Numerous single scattered as well as clusters of mature ganglion cells characterized by abundant eosinophilic cytoplasm, eccentric nuclei, vesicular nuclear chromatin and prominent nucleoli were also noted. Focal edematous area was seen. There was no evidence of dysplasia or neoplasia. Histomorphological features were consistent with Ganglioneuroma.

Post operative CECT was done after 8 months following the surgery. CECT abdomen and pelvis didn't show any residual adrenal space occupying lesion or recurrences.

DISCUSSION

Most adrenal GNs can be misdiagnosed as other adrenal tumors, since they are rare and they don't have specific imaging properties. Adrenal ganglioglomas have a slow growth pattern and usually asymptomatic. Our case represents a huge adrenal ganglioneuroma in a young female patient with non diagnostic flank discomfort. The largest ganglioneuroma reported to date was 19 cm.[9] Our case had a relatively large tumor as its largest diameter as 11.4 cm.

The adrenal imaging is helpful measure for assessing its differential diagnosis. CECT is an effective radiological examination to support the diagnosis with some typical characteristics: (a) the mass can present in different shape like circular, oval, crescent and lobulated. The majority of the tumor edges are smooth and the boundary is clearly defined.[10-12] (b) The embedded growth pattern: some tumor can grow along the gap of adjacent tissues or vessels and encompass them (including aortaventralis, postcava and renal vessels.[13] This case also had an oval shape with well-defined boundaries with maintained fat planes with adjacent structures and it showed typical embedded growth pattern as it seemed to be insinuating between the IVC and right crus of diaphragm. CT scan often displays low density lesions and delayed enhancement or mild enhancement, while the periphery of GNs show more obvious enhancement.[14] This case also showed hypoattenuating suprarenal mass with patchy delayed enhancement. There are about 25% of the tumors with calcification which present mottling and spotty.[15,16] This rate is 2.4% to 60% by other studies.[15,16] Our case also showed hypoattenuating suprarenal mass with patchy delayed and mild enhancement. However it didn't show any mottling or spotty type of calcifications which is usually common in approximately 25% of the tumors.[15,16] Most of the features of adrenal GNs were present in our case except for the calcifications in the plain CT.

T1 weighted shows a homogenous mass with a signal intensity lower than that of the liver and T2 weighted MRI shows a heterogeneous mass with a predominant signal intensity higher than that of the liver.[7-9] High signal intensity on T2 weighted images is presumed to be caused by a combination of myxoid matrix and relatively low numbers of ganglion cells.[17] These MRI findings were similar to our case.

The other differential diagnosis of adrenal GNs include other adrenal solid masses such as phaechromoctymoma, NB, GNB, adenoma and carcinoma.[17,18] GNB and NB more frequently develop in children.[18] They exhibit a higher degree of cellularity and a smaller extracellular space than does GN.[18] This is the main cause of the higher unenhanced density on CT and a lower hypersignal on T2-weighted MRI. Calcifications are often punctuate in GN and amorphous in NB.[19] Pheochromocytoma secretes catecholamines, and this diagnosis is suspected in younger patients with hypertension. Cross-sectional imaging modalities reveal intense early enhancement in pheochromocytoma and progressive delayed enhancement in GN.[17,18]

Certain biological and radiological features should lead the clinician to consider a diagnosis of GN: no elevated hormonal secretion; presence of punctuate calcifications, no vascular involvement, and nonenhanced attenuation of < 40 Hounsfield units on CT; a homogeneous, hypointense mass on T1-weighted MRI; a heterogeneous, hyperintense adrenal mass on T2-weighted MRI; and poor, delayed enhancement on dynamic MRI.[8]

Adrenal GNs arising from the neural crest cells are usually diagnosed incidentally as they are hormonally silent. The process of radiological pre operative diagnosis is extremely challenging. Histologic examination is necessary for the confirmation of the rare diagnosis. Adrenalectomy is the gold standard for the treatment of adrenal GNs. These patients don't require any adjuvant therapy and the chances of recurrences are minimal. Thus, the overall prognosis of the patients is excellent.

REFERENCES

1. Joshi VV. Peripheral neuroblastic tumors: pathologic classification based on recommendations of international neuroblastoma pathology committee (Modification of shimada classification). *Pediatr Dev Pathol*. 2000; 3: 184-99.
2. Chang CY, Hsieh YL, Hung GY, Pan CC, Hwang B. Ganglioneuroma presenting as an asymptomatic huge posterior mediastinal and retroperitoneal tumor. *J Chin Med Assoc.* 2003; 66:370-4.
3. Geoerger B, Hero B, Harms D, Grebe J, Scheidhauer K, Berthold F. Metabolic activity and clinical features of primary ganglioneuromas. *Cancer.* 2001; 91:1905-13.
4. Qing Y, Bin X, Jian W, Li G, Linhui W, Bing L, et al. Adrenal ganglioneuromas: a 10-year experience in a Chinese population. *Surgery.* 2010; 147: 854-860.
5. Rha SE, Byun JY, Jung SE, Chun HJ, Lee HG, Lee JM. Neurogenic tumors in the abdomen: tumor types and imaging characteristics. *Radiographics.* 2003; 23: 29-43.
6. Bjornsson B, Birgisson G, Oddsdottir M. Laparoscopic adrenalectomies: A nationwide single-surgeon experience. *Surg Endosc*. 2008; 22: 622-6.
7. Lonergan GJ, Schwab CM, Suarez ES, Carlson CL. Neuroblastoma, ganglioneuroblastoma, and ganglioneuroma: radiologic-pathologic correlation. *Radiographics*. 2002; 22: 911-34.
8. Majbar AM, Elmouhadi S, Elaloui M, Raiss M, Sabbah F, Hrora A, Ahallat M. Imaging features of adrenal ganglioneuroma: a case report. *BMC Res Notes*. 2014; 7: 791.
9. MC Lai, CC Wang, WC Lin, KL Liu, KH Huang. Huge adrenal ganglioneuroma. *Urology.* 2011; 78: 58-9.
10. Yang WZ, Wei HJ, Gu DQ. Diagnosis and treatment of adrenal ganglioneuroma in the shape of Wax Drip. *Chinese General Practice.* 2012; 15(12): 1378-9.
11. Fan MH, Wang YJ, Xiong AP. CT and MRI features of adrenal ganglioneuroma: correlated with pathologic findings. *Radiologic Practice.* 2014; 29(1): 85-7.
12. Oderda M, Cattaneo E, Soria F, Barreca A, Chuisa L. Adrenal ganglioneuroma with multifocal retroperitoneal; extension. a challenging diagnosis. *Scientific World Journal.* 2011; 11; 1548-53.
13. Zhang HM. Radiological appearances of ganglioneuromas. *Journal of Clinical Radiology.* 2002; 21(7): 527-30.
14. Park BK, Kim CK, Kim B, Kwon GY. Adrenal tumors with late enhancement on CT and MRI. *Abdom Imaging*. 2007; 32(4): 515-8.
15. Chen XJ, Gao JB. CT findings in ganglioneuromas. *Journal of Clinical Radiology.* 2000; 19(3): 159-61.
16. Allende DS, Hansel DE, Maclennan GT. Ganglioneuroma of the adrenal gland. *J Uro*. 2009; 182: 714-5.
17. Lonergan GJ, Schwab CM, Suarez ES, Carlson CL. Neuroblastoma, ganglioneuroblastoma, and ganglioneuroma: radiologic-pathologic correlation. *Radiographics*. 2002; 22(4):911-34.
18. Dunnick NR, Korobkin M. Imaging of adrenal incidentalomas: current status. *AJR Am J Roentgenol.* 2002; 179(3):559-68.
19. Otal P, Mezghani S, Hassissene S, Maleux G, Colombier D, Rousseau H. et al. Imaging of retroperitoneal ganglioneuroma. *Eur Radiol.* 2001; 11(6):940-5.