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Case Report

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GANGLIONEUROMA PRESENTING AS AN ADRENAL INCIDENTALOMA IN A CHILD: A CASE REPORT

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ABSTRACT

An adrenal ganglioneuroma is a rare entity, hormonally Non functioning tumor.it can be presented as adrenal incidentaloma and it can mimic other adrenal malignancies like neuroblastoma. Careful assessment is necessary for an exact diagnosis. A definitive diagnosis made by histological examination. The prognosis is excellent. Here we present a case report of 8 years old girl who came with adrenal incidentaloma and diagnosed as ganglioneuroma based on histology and treated by laparoscopic adrenalectomy with no recurrence of tumor after 5 years of surgery, the case report emphasize the importance of exact diagnosis in adrenal incidentaloma in children to rule out other adrenal malignancy like neuroblastooma and pheochromocytoma

KEYWORD: Ganglioneuroma, neuroblastoma, incidentaloma, Adrenalectomy.

INTRODUCTION

An adrenal ganglioneuroma is a rare entity, hormonally Non functioning tumor.it can be presented as adrenal incidentaloma and it can mimic other adrenal malignancies like neuroblastoma. Careful assessment is necessary for an exact diagnosis. A definitive diagnosis made by histological examination. The prognosis is excellent. Here we present a case report of 8 years old girl who came with adrenal incidentaloma and diagnosed as ganglioneuroma based on histology.

CASE REPORT

8 years old Saudi girl presented with hematuria and headache and history of preceding streptococcal throat infection. she was admitted and diagnosed as poststreptoccoal glumerunephritis. she was managed conservatively. abdomen ultrasound and CT scan were requested and showed left adrenal mass. There was No history of weight gain. No history of headache. No sweating. No history of pubic hair or axilary hair. No breast tissue development. No clitromegally or voice change. Unremarkable family history of malignancy.

On examination she Looks well,no dysmorphic feature. Her weight was 17.8 Kg on 10th centile. Her height 119 cm on 25th centle. Vital sign were within normal rang. Chest examination revealed bilateral good air entry with no added sound. Cardiovascular and abdominal examination were within normal. she was prepubetal stage for breast and pubic hair. Investigation reveled normal complete blood count, renal and liver profile. Urine analysis showed microscopic hematuria. ASOT: 1490 (Normal up to 199).

Endocrine work up: Dexamethasone suppression test was normal Urinary catecholamines and metanephrines were unremarkable ACTH,Cortisol, aldosterone and renin levels were normal. Abdominal ultrasound showed left suprarenal mass (figure 1). Abdominal CT showed left adrenal mass measures 3.5 x 5.3 x 6 cm. no calcification, cysts or necrosis most likely Ganglioneuroma. (figure 2a,2b) Patient underwent laparoscopic left Adrenalectomy Histology report revealed classical Ganglioneuroma Patient was discharged within few days with good clinical condition. No recurrence of tumor was detected in next 5 year of diagnosis. Informed written consent from the parents of the patient was obtained.

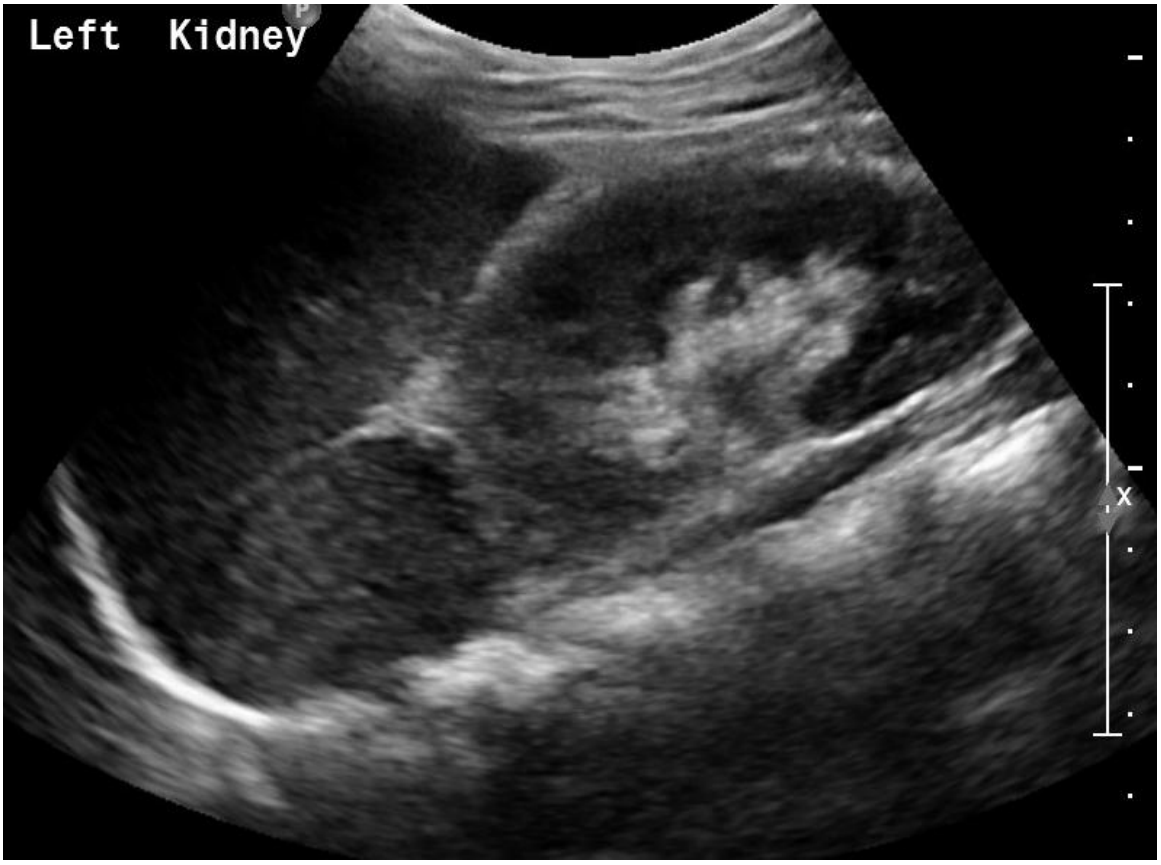

Figure 1: abdominal US: There is a soft tissue mass seen in the left suprarenal region, measures about to 5.6 x 3 cm. No calcifications.

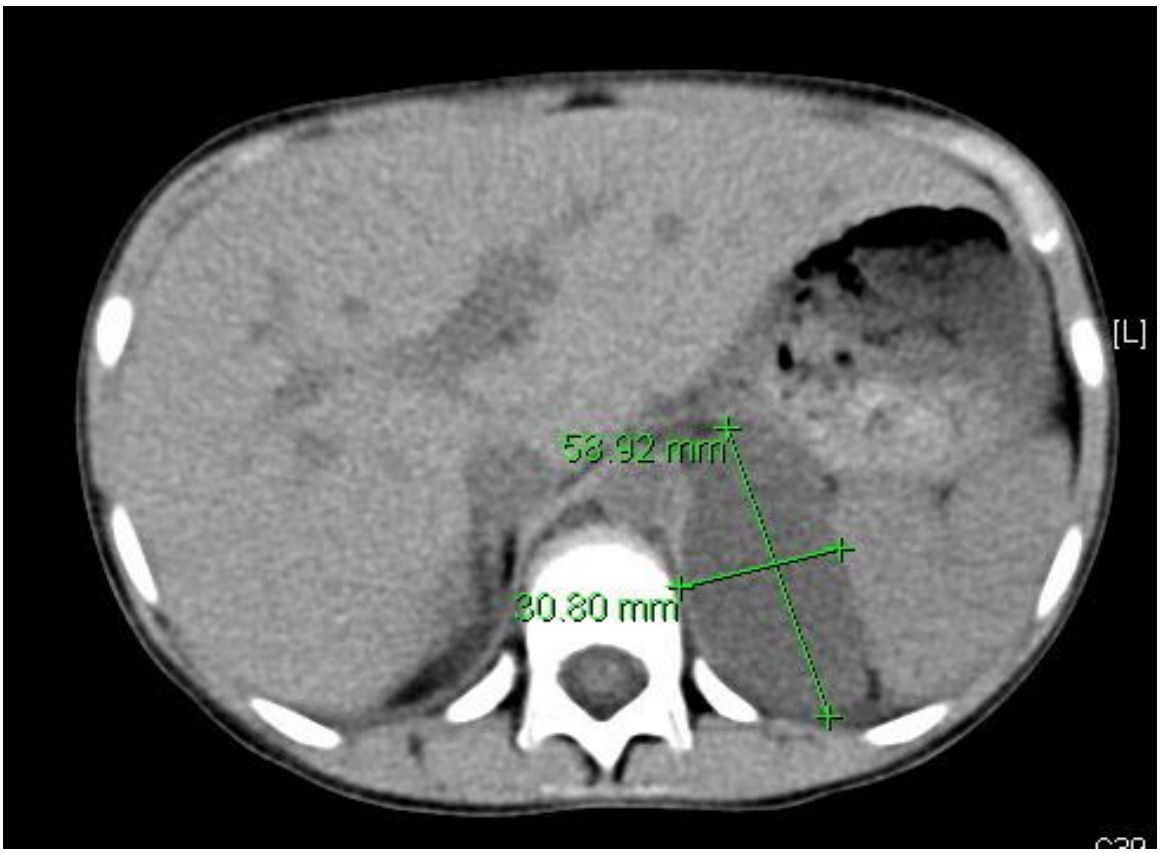

Figure: 2 (a) Abdominal CT showed left adrenal mass measures 3.5 x 5.3 x 6 cm. no calcification, cysts or necrosis.

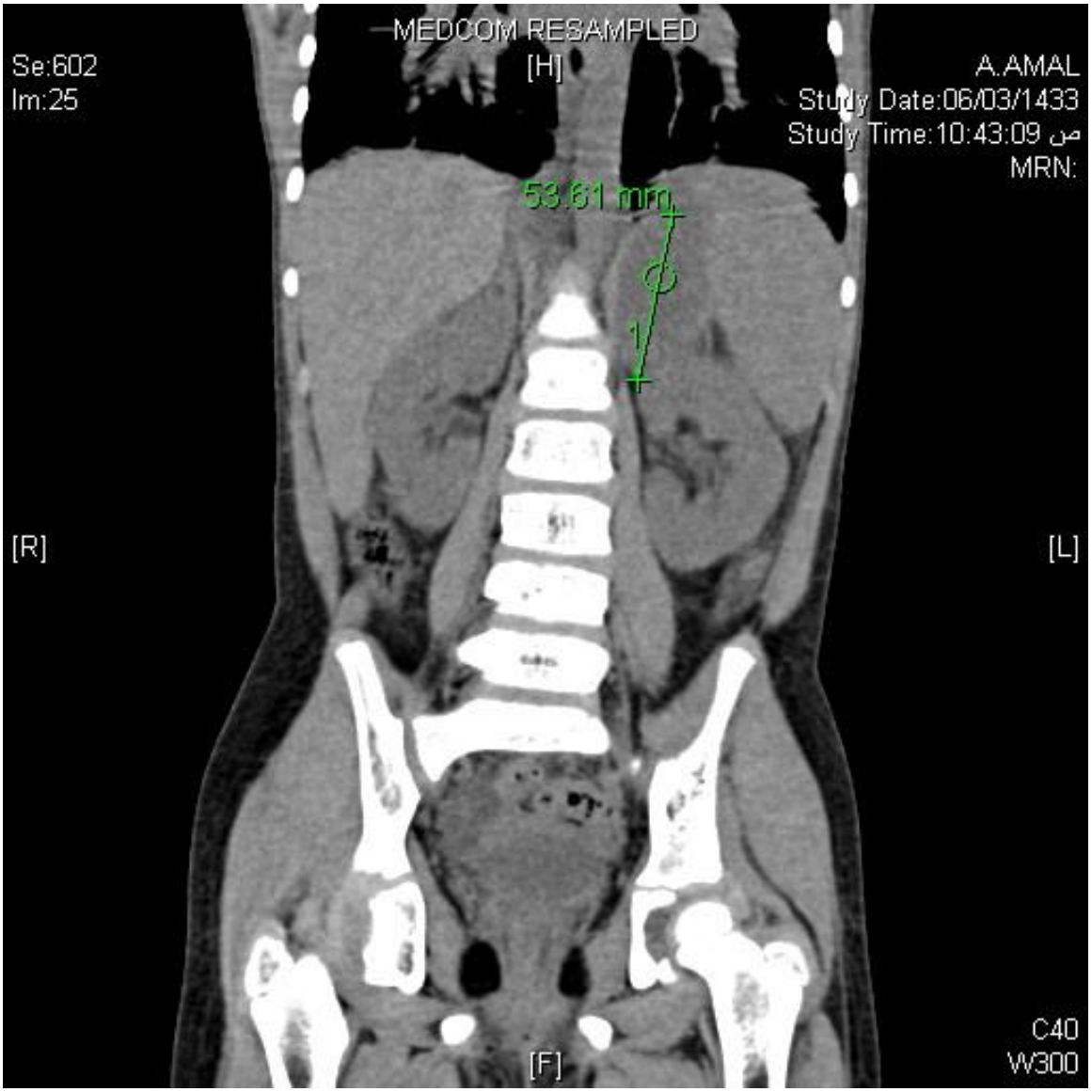

Figure 2 (b): Abdominal CT showed left adrenal mass measures 3.5 x 5.3 x 6 cm. no calcification, cysts or necrosis.

DISCUSSION

Ganglioneuromas (GN) are rare benign tumors originated from neural crest tissue. most commonly found in posterior mediastinum and retroperitoneum. However it is rare to a raised from adrenal gland.

It is begin tumor. most patients are asymptomatic and most cases diagnosed incidentally (1). The prevalence of this entity is 0.2% in young patients, however it can reach to 7% in patient after age of 70 (2). histologically, it is formed of mature Schwann cells and ganglion cells in fibrousstroma (3). GN classified as neurogenic tumors group which includes ganglioblastoma and neuroblastoma. However it is benign condition unlike other form of neurogenic tumor (4). Although the classical imaging features on CT and MRI have been well described, the exact Radiological diagnosis difficult to be made prior to surgery (1). Qing et al. reported that the misdiagnosis rate of GN based on radiology prior to surgery reach up to 64.7% (5). Patients treated by laparoscopic adrenalectomy for a benign neurogenic tumor have an excellent prognosis. Long-term follow-up is needed to avoid recurrence which was reported in few cases. (1)

CONCLUSION

An adrenal GN is a rare entity,hormonally Non functioning tumor.

GN can mimic other adrenal malignancies like neuroblastoma.

Careful assessment by endocrinology workup and radiology is necessary for an exact diagnosis. A definitive diagnosis made by histological examination.
The prognosis is excellent with widely spread laparoscopic adrenalectomy.

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