

Incidentally Detected Adrenal Ganglioneuroma in an adult Female with left Adrenal Mass

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SUMMARY

Adrenal masses can be divided into functional and non-functional masses. Functional adrenal masses usually cause syndromes such as Cushing's syndrome, pheochromocytoma, and primary aldosteronism. On the contrary, non-functional adrenal masses are often asymptomatic and discovered incidentally during radiologic investigations (Incidentaloma). Ganglioneuroma is a benign tumor arising from sympathetic nervous system and made of mature ganglion cells, Schwann cells, neurites and nerve fibers. Ganglioneuroma of adrenal is a quite rare entity and is often asymptomatic.

This article presents a 32-year-old woman with asymptomatic left adrenal mass. After resection of the mass, pathologic examination revealed ganglioneuroma of the adrenal.

Keywords: Ganglioneuroma, Adrenal mass, Incidentaloma

INTRODUCTION

Ganglioneuromas (GNs) are benign tumors arising from sympathetic nervous system. The most common sites of GNs are the retroperitoneum and the posterior mediastinum^{1,2}.

Adrenal GN is a very rare entity. They are usually asymptomatic and detected incidentally. Adrenal incidentalomas are masses that are discovered unexpectedly during a radiologic investigation performed for indications unrelated to adrenal disease. With widespread use of imaging studies such as ultrasonography, computerized tomography (CT) and magnetic resonance imaging (MRI), frequency of adrenal incidentalomas have increased. On the other hand, with increasing age, the prevalence of adrenal incidentalomas increase too. The prevalence is 0.2% in young patients, 3% in those over 50 years of age and 7% in people over 70 years of age². GNs account for 0-6% of the adrenal incidentalomas^{1,3}.

Here in, we report a case of the adrenal incidentaloma in young female that pathologic examination revealed GN.

CASE REPORT

A 32 – year – old female presented with a left adrenal mass detected as an incidental finding. The patient had a negative history of sweating, palpitation, headache, weight gain, muscle weakness, nocturia, and polyuria. There was no significant past medical history. On physical

examination she was normotensive and without abdominal organomegaly. Other physical findings were unremarkable. The laboratory findings were as follows:

Hb= 13.2 g/dl, WBC= $8 \times 10^3/\text{mm}^3$ (neutrophil=65.8%, Lymphocyte=32.4%, mixed=1.8%), platelet= $328 \times 10^3/\text{mm}^3$, blood sugar=87 mg/dl, creatinine= 0.8 mg/dl, Na= 140 meq/L, K= 4.6 meq/L.

Liver function test and urinalysis were normal. Urinary level of vanillyl mandelic acid (VMA) and metanephrine were within normal ranges.

Abdominopelvic ultrasonography showed a large well defined homogenous hypoechoic to mixed echo solid mass in left suprarenal region. Abdominopelvic CT scan demonstrated a 90 × 70 × 95 mm thick wall cystic mass in left adrenal gland (Fig. 1). Because, it was not possible to rule out an adrenal malignancy, she underwent exploratory laparotomy and left adrenalectomy was performed without complications. Histopathological examination of the specimen indicated that it was well- defined, rubbery and white in color. The microscopic sections from adrenal mass, showed proliferation of benign looking spindle cells in fascicles with clusters of mature ganglion cells. No necrosis or cellular atypia was present. No neuroblastic component was identified in multiple sectioning. The diagnosis of ganglioneuroma was made for the mass (Fig. 2).

Post- operative course was uneventful. On 6 months follow- up, the patient is well and asymptomatic.

CASE REPORT

Fig. 1: Abdominopelvic spiral CT scan showed a large homogenous thick wall cystic mass in left supra renal region.

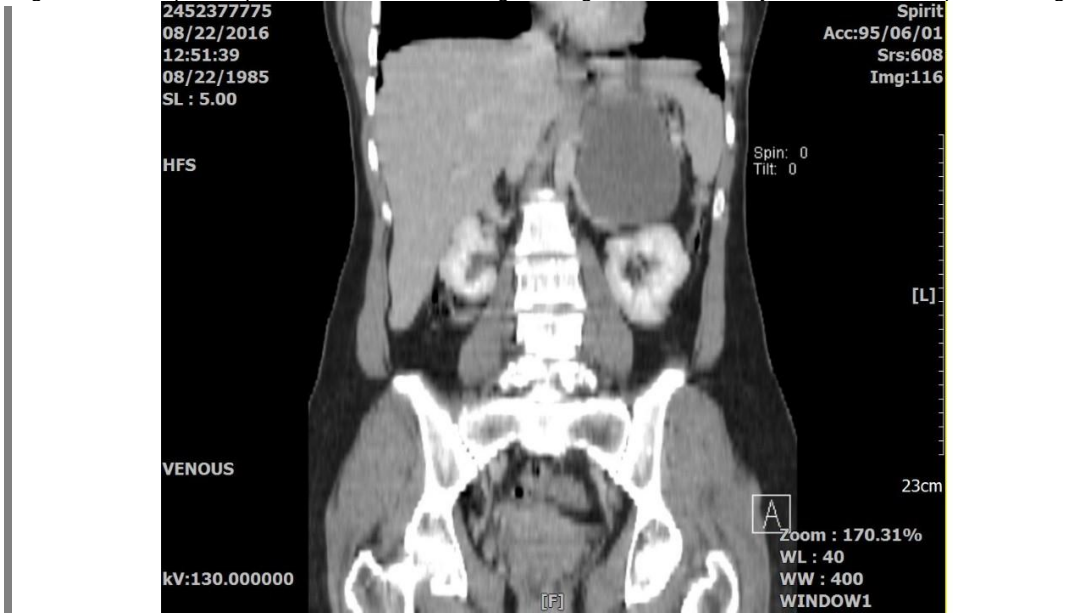
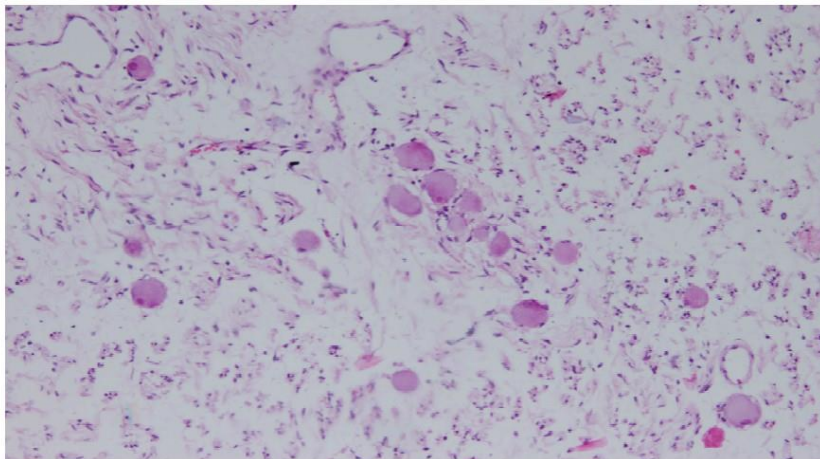


Fig.2: Microscopic section of adrenal mass showing clusters of mature ganglion cells surrounded by fascicles of Schwann-like cells. (Hematoxyline and Eosin stain, × 100)



DISCUSSION

GNs are benign, well- differentiated tumors arising from primordial neural crest cells. GNs are the benign end of the spectrum of ganglion cells tumors.

Neuroblastomas are at the malignant end and ganglioneuroblastomas are intermediate. GNs are most commonly found in the retroperitoneum (40-50%) and the posterior mediastinum (30-40%) and adrenal gland is very rare involved by GN [1, 3]. Although GNs are generally considered as hormonally inactive silent tumors, but they can secrete several hormones such as cortisol, cate

cholamine, vasoactive intestinal polypeptide (VIP) and androgenic hormone, and so, they may produce hypertension, diarrhea, flushing, and virilization [4-5]. Therefore, the vast majority of adrenal GNs have been diagnosed by radiologic investigations incidentally (incidentalomas). Adrenal GNs should be suspected radiologically in an adrenal mass containing discrete calcifications, non – functioning, non enhanced attenuation of less than 40 Hounsfield units on CT scan [6]. On MRI, findings of a non – enhanced T1- weighted signal, slightly high T2 – weighted signal with late and gradual

enhancement are in favor of adrenal GNs [1, 6]. Although the radiologic appearance of adrenal GN on CT scan and MRI have been well described, the misdiagnosis rate of adrenal GN on CT scan and MRI have been reported as high as 64.7%⁷.

Calcification within adrenal mass is characteristic of GN on CT scan; however, only 29% to 60% of GNs show calcification^{3,4,6}. Therefore, the definite diagnosis of adrenal GN is confirmed only by histopathologic examination. A relationship exists between the size of an adrenal lesion and its malignant potential with larger masses more likely to exhibit malignant features. So, most experts recommend 4^{cm} as the cut off diameter that warrant surgical resection^{4,8}. Complete surgical resection is highly recommended for treatment of adrenal GN and to differentiate it from other neural crest tumors and adrenal malignancy. The prognosis of adrenal GN following surgical resection is excellent, whereas, regular and long term follow-up is recommended in these patients.

CONCLUSION

Adrenal GN is a rare entity and is usually asymptomatic. So, it should be considered in differential diagnosis of any adrenal incidentalomas. Surgical excision is treatment of choice for adrenal GNs. Close follow-up and periodic radiologic evaluation is recommended often surgery.

Conflict of interest: The authors declare that they have no conflict of interest.

REFERENCES

1. Tungenwar P, Bhandarwar A, Gandhi S, Binayke R, Agarwal S, Pai A. Adrenal ganglioneuroma: A rare case presentation Int J Case Rep Images (IJCRI). 2016;7(8):519-23.
2. Grumbach MM, Biller BM, Braunstein GD, Campbell KK, Carney JA, Godley PA, et al. Management of the clinically inapparent adrenal mass ("incidentaloma"). Ann Intern Med. 2003;138(5):424-9.
3. Ozturk H. Adrenal Ganglioneuroma Presenting As Left Renal Mass. Worldly oncol. 2014;5(2):90-2.
4. Adas M, Koc B, Adas G, Ozulker F, Aydin T. Ganglioneuroma presenting as an adrenal incidentaloma: a case report. J Med Case Rep. 2014;8(1):131.
5. Georger B, Hero B, Harms D, Grebe J, Scheidhauer K, Berthold F. Metabolic activity and clinical features of primary ganglioneuromas. Cancer. 2001;91(10):1905-13.
6. Maweja S, Materne R, Detrembleur N, de Leval L, Defechereux T, Meurisse M, et al. Adrenal ganglioneuroma. Am J Surg. 2007;194(5):683-4.
7. 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(6):854-60.
8. McDougal WS, Wein AJ, Kavoussi LR, Partin AW, Peters CA. Campbell-Walsh Urology 11th Edition Review: Elsevier Health Sciences; 2015.