Case Report

Ganglioneuroma of the adrenal gland and retroperitoneum: A case report

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Abstract

Context: Ganglioneuromas are benign tumors of the sympathetic nervous system that rarely arise in the adrenal gland. Majority of cases are detected incidentally since they are usually asymptomatic. Up to the current era of laparoscopic adrenal mass excision, this unusual entity has not been adequately reported in the surgical literature. **Case Report:** A 51 year old male with history of hypertension was found to have abdominal bruit during a regular physical examination. A 4 cm right adrenal mass with upper pole calcification and a 6 cm retro-pancreatic mass were subsequently found on a computed tomography scan. Endoscopic ultrasound-guided needle biopsy was indeterminate. Preoperative endocrine evaluation showed mildly elevated vanillyl mandelic acid with normal 24-hour cathecolamine, metanephrine and cortisol levels. Histopathologic examination after an uneventful laparoscopic excision was consistent with ganglioneuroma. **Conclusions:** Ganglioneuroma occurs rarely in adrenal gland and preoperative diagnosis is difficult since symptoms are usually nonspecific. Due to widespread utilization of abdominal imaging, however, it should be included in differential diagnosis of adrenal or retroperitoneal mass. Histopathologic examination is currently the mainstay of diagnosis.

Keywords: Ganglioneuroma, retroperitoneum, adrenal gland, excision.

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Introduction

Ganglioneuromas, benign tumors of the sympathetic nervous system, are rare to arise in the adrenal gland. They are usually asymptomatic and hormonally silent. Majority of cases are detected incidentally during work-up for unrelated conditions. They usually develop from the great sympathetic chains, extending from the base of the skull to the neck, posterior mediastinum, retroperitoneum, and adrenal glands.

Currently, histopathologic examination is the only tool to diagnose ganglioneuroma and to differentiate it from other neural crest tumors. Up to the current era of laparoscopic adrenal mass excision, this unusual entity has not been adequately reported in the surgical literature.

Case Report

A 51 year old man was referred to Geisinger clinic by his primary care physician for an abdominal bruit that was found during a regular physical examination. Past medical and surgical histories were only significant for mild hypertension. Abdominal computed tomography (CT) scan revealed a 4 cm right adrenal mass with upper pole calcifications and a 6 cm retropancreatic mass (Figures 1 and 2). Both masses are mainly hypodense on arterial imaging and heterogenous on delayed phase images. Endoscopic ultrasound (EUS) guided needle biopsy was indeterminate. Preoperative endocrine evaluation only

showed mildly elevated vanillyl mandelic acid (VMA) with a normal 24-hour cathecolamine, metanephrine and cortisol levels. Intraoperatively, the tumors were able to be separated off the surrounding structures without major difficulty. Histopathologic examination after a complete laparoscopic excision was consistent with the diagnosis of ganglioneuroma.

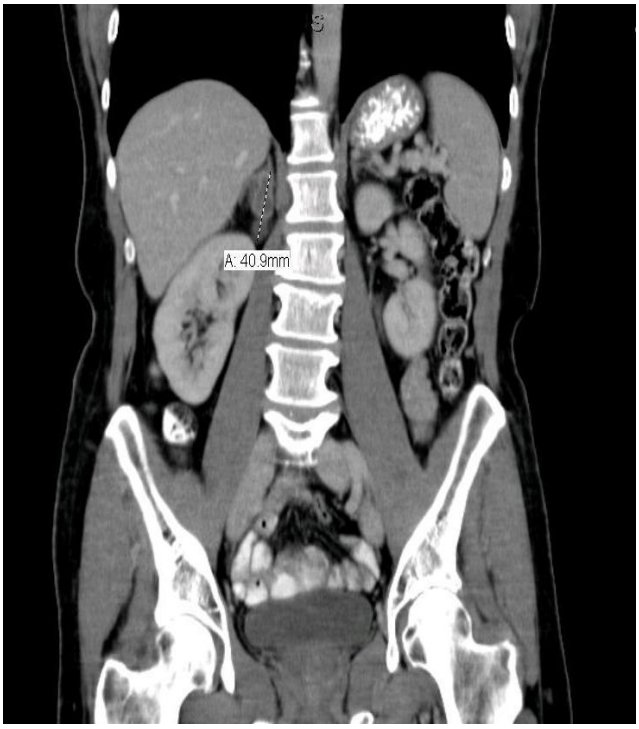

Fig. 1 Coronal section of the right-sided adrenal ganglioneuroma.

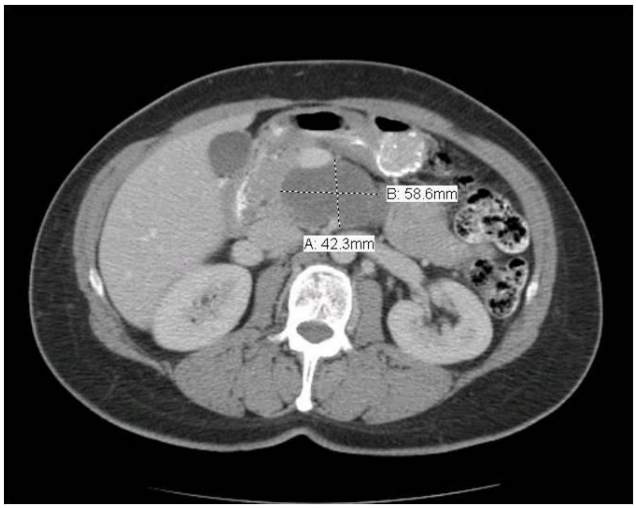

Fig. 2 Axial section of the retropancreatic mass (5.8 x 4.3 cm).

Discussion

Ganglioneuroma (GN) originates from cells of the neural crest that include the sympathetic ganglias and the adrenal glands. It is composed of ganglion cells, neurites, schwann cells, and fibrous tissues. This tumor affects children and young people preferentially with three-fifth developed before age 20. Females are more prone to be affected than males. Majority of GNs are thoracic and retroperitoneal in location while adrenal GNs are rare.

Clinical symptoms of GNs are non-spesific, mosty hormonally silent and related to their size/locations. Despite their generally benign nature, GNs may come to attention due to compression of their neighboring structures [1]. Approximately up to 30% of patients were found to have elevated plasma and urinary catecholamine but they rarely develop symptoms of vasoactive amines excess. Immature element such as neuroblast is not part of mature GN explaining lower rate of metaiodobenzylguanidine (MIBG) uptake (57%) compared to neuroblastoma (92%) [2].

Diagnosis of GN should be suggested when an adrenal tumor harbors the following: (1) no hormonal hypersecretion, (2) presence of punctate or discrete calcifications, (3) absence of vessel involvement, and (4) a low non-enhanced T1-weighted signal with a late and gradual enhancement on dynamic magnetic resonance imaging (MRI). The treatment for this condition is complete surgical resection through either an open or laparoscopic approach.

The most significant differential diagnosis of ganglioneuroma is neuroblastoma. Increased level of urinary noradrenaline, dopamine, HVA, and VMA are frequently encountered in neuroblastoma, while the level of urinary cathecolamine, HVA, and VMA are usually normal in ganglioneuroma [3]. In the present review of 14 cases displayed in Table 1, an increased urinary catecholamine level and sex hormone were noted in several patients, therefore pharmacological diagnostic tests for such hormonal activities may not necessarily be discriminative in the differential diagnosis of these two related tumors.

Conclusions

Ganglioneuroma occurs rarely in adrenal gland and preoperative diagnosis is difficult since symptoms vary and oftentimes nonspecific. Due to widespread utilization of abdominal ultrasonography, CT scan and MRI, detection of such tumor has increased, therefore differential diagnosis of an adrenal/retroperitoneal mass should include ganglioneuroma. Histopathologic examination is currently the mainstay of diagnosis.[17]

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Table 1 Previously published case reports of adrenal and retroperitonal ganglioneuromas

Authors	Age	Sex	Abdominal Symptoms	Systemic Symptoms	Largest Dimension(cm)	Hormones Secreted	Treatment
Sylvie Maweja et al [4]	20	M	None	None	5	None	Adrenalectomy
Erem C et al [5]	46	F	R flank pain	Paroxysmal Hypertension	4	Dopamine, Homovanilic acid	Adrenalectomy
Arredondo M et al [6]	50	M	None	None	6.5	None	Adrenalectomy
Gupta R et al [7]	40	F	R upper flank pain	None	8.5	None	Adrenalectomy
Ito H et al [8]	33	F	Epigastric pain	None	4	Dopamine	Adrenalectomy
Erem C et al [9]	68	F	None	None	6	None	Adrenalectomy
CA Koch at al [10]	63	F	None	Chest pain, Hypertension	5	Metanephrine, Normetanephrine	Adrenalectomy
Diab DL et al [11]	56	F	None	Hirsutism, Clitoromegaly	5	Testosterone	Laparoscopic Adrenalectomy
Aygen Ozbay et al [12]	77	F	Watery Diarrhea	Sweating, Palpitation	12	Vanyllylmandelic acid, Adrenaline	Adrenalectomy
Mukai M et al [13]	37	M	None	None	10	Aldosterone	Adrenalectomy
Lora MS et al [14]	6	F	None	MEN 2B	4	None	Laparoscopic Adrenalectomy
Lora MS et al [14]	11	M	None	MEN 2A	7	None	Laparoscopic Adrenalectomy
Moriwaki Y et al [15]	26	F	None	None	8	None	Adrenalectomy
Hinyokika Kiyo at al [16]	38	M	Upper abdominal pain	None	11	None	Adrenalectomy