Non-Functional Adrenal Gland Ganglioneuroma Masquerading as Chronic Calculus Cholecystitis

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Abstract

Adrenal ganglioneuromas in young adults are rare and ill-understood. We report an incidentally detected adrenal gland tumor diagnosed as ganglioneuroma (mature type) in a 33 years old man who presented with vomiting and epigastric pain for 2 months. Histopathology examination revealed a well-encapsulated benign tumor of mature ganglion cells and Schwann-like cells arranged in fascicles, staining strongly with NSE and s-100 proteins, with adjacent unremarkable adrenal cortex and medulla.

Introduction

Ganglioneuromas (GN) of adrenal gland in adults are very rare and can have serendiptous presentation. We describe such a non-functional adrenal GN in a young man.

Case Report

A 33 years old man presented with complaints of vomiting and epigastric pain of 2 months duration. There was no flank pain, fever, hematuria, pyuria and weight loss. Past history and family history were non-contributory. On examination, he was well-built, afebrile, with pulse 96 /min, blood pressure 160/90 mmHg and body mass index was 31.8 kg/m². Systemic examination was unremarkable. Abdomen was soft without organomegaly. Routine lab investigations were unremarkable with normal blood sugar, hematology, liver, and renal function profile. Hepatitis C/ B antibodies and HIV were non-reactive. Electrocardiogram was unremarkable.

USG revealed unremarkable liver, spleen and pancreas; gall bladder (GB) was distended with 4 mm sized echogenic shadow partially occupying the lumen reported as calculus/sludge. Both kidneys were unremarkable. A hypoechoic area with well-defined margins measuring 8.1 x 8 cm was seen over left supra-renal area displacing left kidney (LK) laterally. CT abdomen revealed well-defined rounded hypodense non-enhancing homogenous mass devoid of necrosis/calcification in the region of left adrenal gland measuring 80 x 78 mm, indistinct from adrenal gland. Other abdominal, pelvic and urogenital organs were normal.

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adjacent well-encapsulated benign
tumor of mature ganglion cells
and Schwann cells arranged in
fascicles

except for contracted GB with few tiny
calculi in lumen. Serum cortisol was
0.52 ng/dL (reference range: 3-16.6 ng/
dL), urinary vanillyl mandelic acid, 8.80
mg/24 hours (reference range: 0-13.6
mg/24 hours) and metanephrine, 110.0
mcg/24 hours (reference range: 25-312
mcg/24 hours). Clinical impression was
benign left adrenal gland tumor.

M R I T 2 w e i g h t e d  i m a g e
showed well-defined encapsulated heterogeneously enhancing left adrenal
mass measuring 77 x 72 mm with
delayed contrast washout, causing
mild compression of underlying LK
upper pole without invading it (Figure
1). Right kidney and adrenal gland were
normal, lymphadenopathy was absent.
Laparoscopic adrenalectomy was
performed to remove non-adenomatous
lesion and sent for histopathology
evaluation.

Gross examination revealed adrenal
gland with tumor weighing 300 grams,
measuring 10 x 9 x 7 cm, nodular,
irregular grey colored, congested and
partially covered with fatty tissue. Cut
section showed a well-
encapsulated tumor with homogenous
grey appearance (Figure 2). Microscopy
examination revealed normal adrenal
cortical and medullary tissue with
adjacent well-encapsulated benign
tumor of mature ganglion cells and
Schwann cells staining strongly
with neuron-specific enolase, X 40

Discussion

Adrenal GN are uncommon tumors.
To our knowledge there are totally less
than 500 adrenal GN reported in the
world literature. Their serendipitous
presentation makes diagnosis difficult
and catches the treating physician by
surprise. It is usually asymptomatic and
hormonally silent. Though GN
synthesizes catecholamines, it rarely
causes hypertension and is incidentally detected on imaging
studies for unrelated symptoms.

However some patients do present with epigastric/ abdominal pain with
diarrhea, vomiting and hypertension as
was noted in our patient. Incidentally
our patient had associated gall stones
and hence calculus cholecystitis was
suspected. Long term studies in Chinese
population showed 180
incidental adrenal tumors resected, out
of which 17 (9.4%) were diagnosed as
adrenal GN. Malignant transformation
is rarely reported. Surgical resection is
the choice of therapy for such tumors.

Tumors arising from neural crest
cells including sympathetic ganglia
and adrenal medulla comprise
pheochromocytoma/paraganglioma,
neuroblastomas, ganglioneuroblastoma
and GN. These are “functional” i.e.
catecholamine-producing or “non-
functional”. The most common location
of GN includes posterior mediastinum
and retroperitoneum; only rarely are
they found in the adrenal gland. Adrenal
GN are more frequent in 4th/5th
decade of life unlike our patient who
was in his 30s. The most common site
of GN are retroperitoneal (32-52%),
mediastinal (39-43%), or cervical region
(8-9%) and only rarely in adrenal
gland.

On 6 months follow-up, the patient
is asymptomatic. GB is status quo.

References

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