Adrenal ganglioneuroma: A rare case presentation

P. N. Tungenwar, A. H. Bhandarwar, Saurabh Gandhi, Rachana Binayke, Samarth Agarwal, Ajay Pai

ABSTRACT

Introduction: Ganglioneuromas are benign, well differentiated tumors arising from the sympathetic nervous system. However, they arise rarely in the adrenal glands. Ganglioneuroma (GN) is a very rare (0–6% of incidentalomas) tumor that arises from sympathetic ganglion/chain and is made of mature ganglion cells, Schwann cells, neurites and nerve fibers. Most cases of adrenal ganglioneuromas are incidentally diagnosed since they are mostly asymptomatic and produce symptoms rarely due to compression of neighboring structures.

Case Report: We hereby present a rare case of an adult female patient presented with pain in abdomen diagnosed with a non-secretory adrenal mass who underwent laparoscopic excision of the mass. She was finally diagnosed on pathological examination to be an adrenal ganglioneuroma, one of the very rare tumors reported in literature.

Conclusion: Adrenal ganglioneuromas occur rarely and difficult in diagnosing preoperatively since symptoms vary and are mostly nonspecific. Ganglioneuroma should not be missed as a differential diagnosis of an adrenal mass. Histopathologic examination plays a crucial role in diagnosis.
Adrenal ganglioneuroma: A rare case presentation

P. N. Tungenwar, A. H. Bhandarwar, Saurabh Gandhi, Rachana Binayke, Samarth Agarwal, Ajay Pai

ABSTRACT

Introduction: Ganglioneuromas are benign, well differentiated tumors arising from the sympathetic nervous system. However, they arise rarely in the adrenal glands. Ganglioneuroma (GN) is a very rare (0–6% of incidentalomas) tumor that arises from sympathetic ganglion/chain and is made of mature ganglion cells, Schwann cells, neurites and nerve fibers. Most cases of adrenal ganglioneuromas are incidentally diagnosed since they are mostly asymptomatic and produce symptoms rarely due to compression of neighboring structures. Case Report: We hereby present a rare case of an adult female patient presented with pain in abdomen diagnosed with a non-secretory adrenal mass who underwent laparoscopic excision of the mass. She was finally diagnosed on pathological examination to be an adrenal ganglioneuroma, one of the very rare tumors reported in literature. Conclusion: Adrenal ganglioneuromas occur rarely and difficult in diagnosing preoperatively since symptoms vary and are mostly nonspecific. Ganglioneuroma should not be missed as a differential diagnosis of an adrenal mass. Histopathologic examination plays a crucial role in diagnosis.

Keywords: Ganglioneuroma, Adrenal, Adult, Laparoscopic excision

INTRODUCTION

Ganglioneuromas are benign, well differentiated tumors arising from the sympathetic nervous system. However, they arise rarely in the adrenal glands. Ganglioneuroma is a very rare (0–6% of incidentalomas) tumor that arises from sympathetic ganglion/chain and is made of mature ganglion cells, Schwann cells, neurites and nerve fibers. The most common sites are the posterior mediastinum and the retroperitoneal space [1]. Most cases of adrenal ganglioneuromas are incidentally diagnosed since they are mostly asymptomatic and produce symptoms rarely due to compression of neighboring structures. Retroperitoneal ganglioneuromas are usually non-secreting and asymptomatic but when they reach large sizes they cause pressure symptoms locally [2]. We hereby present a rare case of an adult female patient presenting with pain in abdomen diagnosed with a non-secretory adrenal mass who underwent laparoscopic excision of the mass. She
was finally diagnosed on pathological examination to be an adrenal ganglioneuroma, one of the very rare tumors reported in the medical world literature [3, 4].

Adrenal ganglioneuromas (GN) are rare tumors gaining origin from the neural crest cells of the sympathetic nervous system. This family of ganglion cell tumors includes GN which is benign, ganglioneuroblastoma which are of intermediate differentiation, and neuroblastoma which is a highly malignant lesion. Neuroblastoma and ganglioneuroblastoma mostly occur in infants and children, whereas GN has more prevalence in adolescents and young adults.

Laparoscopic adrenalectomy is now the gold standard in management of small adrenal masses. The standard treatment for benign adrenal tumors is laparoscopic or/ minimal access surgery. Large tumors (> 6 cm) are relative contraindications to laparoscopic adrenalectomy as they are more in favor of a diagnosis of adrenal carcinoma. We present in the following report a peculiar presentation of this condition and its laparoscopic excision followed by histopathology proving a diagnosis of ganglioneuroma.

**CASE REPORT**

A 40-year-old female presented with a history of vague pain in abdomen since 15 days and not associated with local flank swelling, constipation, or anorexia. There was no significant past surgical or medical history. No history of recent travel and family history of colitis or inflammatory bowel disease. On physical examination, the abdomen was soft, there was no distension or tenderness.

Computed tomography scan of the abdomen and pelvis with contrast study was suggestive of a right suprarenal mass lesion measuring approximately 4x2 cm with internal necrotic and calcified areas and moderate relatively homogenous with post contrast enhancement suspicious of neurogenic tumor (Figure 1A). Routine laboratory investigations including thyroid function tests were within normal range (Table 1).

Laparoscopic excision of the suprarenal mass was performed and sent for histopathological examination (Figure 1B).

On gross examination of specimen was a solid firm well delineated mass of size 4x3x2 cm brown in colour on the outside and cut section revealed pale greyish white appearance with peripheral golden yellow tissue with focal calcification (Figure 1C). Microscopic sections of the histopathological specimen showed ganglion cells and Schwann cells arranged in interlacing bundles suggestive of ganglioneuroma (Schwannian stroma dominant neuroblastic tumors) with peripheral normal adrenal parenchyma and no evidence of malignancy. On immunohistochemistry ganglion cells showed positivity for chromogranin and S-100 Protein was positive in fibres and focally in ganglion cells (Figure 2).

<table>
<thead>
<tr>
<th>Table 1: Endocrine investigations showing non-secreting nature of tumor</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Unit</strong></td>
</tr>
<tr>
<td>Triiodothyronine (T3) ng/mL</td>
</tr>
<tr>
<td>Free thyroxine (FT4) pmol/L</td>
</tr>
<tr>
<td>Thyroid-stimulating hormone (TSH) IU/mL</td>
</tr>
<tr>
<td>Aldosterone (pmol/L) decubitus</td>
</tr>
<tr>
<td>Standing</td>
</tr>
<tr>
<td>Plasma Renin activity (ng/ml/hr) Decubitus</td>
</tr>
<tr>
<td>Standing</td>
</tr>
<tr>
<td>Plasma basal cortisol µg/dl</td>
</tr>
<tr>
<td>Plasma Free Metanephrine pg/dl</td>
</tr>
<tr>
<td>Plasma Free Nor-Metanephrine pg/dl</td>
</tr>
<tr>
<td>Urine VMA mg/24 h</td>
</tr>
</tbody>
</table>

* Not detectable

Figure 1: (A) Computed tomography scan of abdomen and pelvis showing right adrenal mass (arrow) with calcifications, (B) Intraoperative picture showing the adrenal mass in relation to the kidney and the liver, (C) Cut section revealing Greyish white surface of tumor (arrow) with small arrowhead showing adrenal parenchyma.
DISCUSSION

Ganglioneuromas can occur in the central nervous system or in the peripheral nervous system (sympathetic) [1]. It usually affects children and young patients below the age of 20. Females are affected more than males [2]. The most common site of GN are retroperitoneum (40–50%) followed by mediastinal location (30–40%), or cervical region (up to 10%) and only rarely in adrenal gland [3–5].

The incidence of adrenal ganglioneuroma is 1/ million population being most commonly sporadic in occurrence but could be associated with other syndromes like multiple endocrine neoplasia type 2/neurofibromatosis type 2.

Ganglioneuromas are clinically asymptomatic and mostly hormonally inactive. Despite being benign and hormonally inactive in nature, GNs may cause pressure symptoms due to compression of their adjacent structures [5, 6]. Up to one-third of patients have elevated catecholamine levels but they rarely develop symptoms due to their excess.

Ganglioneuroma (GN) arises from the neural crest cells—the sympathetic ganglia and the adrenals. It is composed of Schwann cells, ganglion cells and fibrous tissues. The differential diagnosis of a tumor with high resemblance to ganglioneuroma is a neuroblastoma. In neuroblastoma, levels of urinary homovanillic acid (HVA) and vanillylmandelic acid (VMA) are usually raised, while the levels of urinary HVA and VMA in ganglioneuroma are within normal ranges [7]. Neuroblasts are not a part of mature GN. Thus the meta iodo benzyl guanidine uptake of a GN is lower than that of a neuroblastoma.

The GN should be diagnosed when the following findings are noted in a case of an adrenal mass: (1) non-secreting, (2) absent vessel involvement, (3) calcifications in the gland, and (4) magnetic resonance imaging (MRI) findings of a non-enhanced T1-weighted signal with late and gradual enhancement. The final treatment for adrenal ganglioneuromas is resection by either open or laparoscopic method [8–10].

CONCLUSION

Adrenal ganglioneuromas occur rarely and difficult in diagnosing preoperatively since symptoms vary and are mostly nonspecific. Due to widespread utilization of imaging modalities like abdominal ultrasonography, computed tomography and magnetic resonance imaging, detection of such tumor has increased. Thus, ganglioneuroma should not be missed as a differential diagnosis of an adrenal mass. Histopathologic examination plays a crucial role in diagnosis.

**********

Author Contributions

P. N. Tungenwar – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

A. H. Bhandarwar – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Saurabh Gandhi – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Rachana Binayke – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Samarth Agarwal – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Ajay Pai – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

© 2016 P. N. Tungenwar et al. This article is distributed under the terms of Creative Commons Attribution License which permits unrestricted use, distribution and reproduction in any medium provided the original author(s) and original publisher are properly credited. Please see the copyright policy on the journal website for more information.
REFERENCES


ABOUT THE AUTHORS


Ajay H. Bhandarwar is Professor at Department of General Surgery, Grant Government Medical College & Sir JJ Group of Hospitals, Mumbai, India. His area of interest are upper gastrointestinal, diaphragmatic and metabolic surgeries. His research interests include hiatus hernia repairs and its lifestyle consequences. He has published several papers in International and National Journals.
E-mail: abhandarwar@gmail.com

Pravin N. Tundenwar is Associate Professor at Department of General Surgery, Grant Government Medical College & Sir JJ Group of Hospitals, Mumbai, India. His area of interest are solid organs and hernia repair surgery. His research interests include metabolic surgeries with keen interest in newer trends.
E-mail: pravint30.pt@gmail.com

Saurabh S. Gandhi is Assistant Professor at Department of General Surgery, Grant Government Medical College & Sir JJ Group of Hospitals, Mumbai, India. His area of interest are gastrointestinal and metabolic repair surgery. His research interests include endoscopic thyroid, hernia surgery with keen interest in newer trends.
E-mail: saur1234@gmail.com
Rachana Binayke is Assistant Professor at Department of Pathology, Grant Government Medical College & Sir JJ Group of Hospitals, Mumbai, India. She earned undergraduate degree (M.B.B.S) from Mumbai University and postgraduate (MD) Pathology Degree from Seth G.S. Medical College and K.E.M Hospital, Mumbai, India. Her research interests include neuropathology, pulmonary pathology and immunopathology and intend to pursue fellowships in neuropathology, immunopathology and oncopathology.

Samarth Agarwal is Lecturer in Surgery at Department of General Surgery, Grant Government Medical College, Mumbai, India. He earned undergraduate degree (MBBS) from TN Medical College, Mumbai, India and postgraduate degree (M.S. General Surgery) from Grant Governement Medical College, Mumbai, India. He has published 10 research papers in National and International Academic journals. His research interests include minimally invasive laparoscopic, urological and bariartric surgeries. He intends to pursue MCH urology in future.

Ajay Pai is Lecturer at Department of General Surgery, Grant Government Medical College & Sir JJ Group of Hospitals, Mumbai, India. His area of interest are upper gastrointestinal and metabolic repair surgery. His research interests include endoscopic thyroid, hernia surgery with keen interest in newer trends. He wants to pursue mch gastrointestinal surgery in future.
About Edorium Journals
Edorium Journals is a publisher of high-quality, open access, international scholarly journals covering subjects in basic sciences and clinical specialties and subspecialties.

But why should you publish with Edorium Journals?
In less than 10 words - we give you what no one does.

Vision of being the best
We have the vision of making our journals the best and the most authoritative journals in their respective specialties. We are working towards this goal every day of every week of every month of every year.

Exceptional services
We care for you, your work and your time. Our efficient, personalized and courteous services are a testimony to this.

Editorial Review
All manuscripts submitted to Edorium Journals undergo pre-processing review, first editorial review, peer review, second editorial review and finally third editorial review.

Peer Review
All manuscripts submitted to Edorium Journals undergo anonymous, double-blind, external peer review.

Early View version
Early View version of your manuscript will be published in the journal within 72 hours of final acceptance.

Manuscript status
From submission to publication of your article you will get regular updates (minimum six times) about status of your manuscripts directly in your email.

Our Commitment

- Six weeks
  You will get first decision on your manuscript within six weeks (42 days) of submission. If we fail to honor this by even one day, we will publish your manuscript free of charge.*

- Four weeks
  After we receive page proofs, your manuscript will be published in the journal within four weeks (31 days). If we fail to honor this by even one day, we will publish your manuscript free of charge and refund you the full article publication charges you paid for your manuscript.*

Favored Author program
One email is all it takes to become our favored author. You will not only get fee waivers but also get information and insights about scholarly publishing.

Institutional Membership program
Join our Institutional Memberships program and help scholars from your institute make their research accessible to all and save thousands of dollars in fees make their research accessible to all.

Our presence
We have some of the best designed publication formats. Our websites are very user friendly and enable you to do your work very easily with no hassle.

Something more...
We request you to have a look at our website to know more about us and our services.

We welcome you to interact with us, share with us, join us and of course publish with us.

* Terms and condition apply. Please see Edorium Journals website for more information.