Symptomatic pelvic paraganglioma in an adult

Paragangliomas are relatively rare in adults, with most arising from para-aortic sympathetic chain. Paraganglioma localized at the extravesical pelvic cavity is extremely rare, only few case reports are published. We report a case of paravesical paraganglioma in a 45-year-old man who initially presented with irritable bladder and palpitations after voiding for 6 years and was being treated symptomatically for urinary tract infection and anxiety. Cross-sectional imaging showed a paravesical mass and was treated by robotic excision of the mass.

**Key words:** Catecholamine, cross-sectional imaging, extra-adrenal pheochromocytoma, paraganglioma, paravesical

**INTRODUCTION**

Paragangliomas are neuroendocrine tumors which have a roughly symmetric distribution with extension from the skull base down to the pelvic floor.\(^1\) The most conspicuous member of this system is known as the adrenal medulla. Extra-adrenal paraganglia, defined as paraganglia located outside the adrenal gland, can be divided into two broad groups: Paraganglia associated with the parasympathetic system and those related to the sympathetic system.\(^2\) The former are located in the head, neck, and anterior mediastinum and are believed to have a chemoreceptor function. The latter are predominantly found in the posterior mediastinum and retroperitoneum along with the thoracolumbar paravertebral region, and they are thought to have a similar function to that of the adrenal medulla. In addition, small paraganglia are found within viscera such as the urinary bladder and gallbladder.

**CASE REPORT**

A 45-year-old male with average built resident of high altitude area in India. He had a long standing complaint of irritable bladder and palpitations specially after voiding for about 6 years. He was treated by the local physicians symptomatically with antibiotics. His earlier ultrasounds (USG) were normal with no residual urinary volume. About a year back USG abdomen revealed a well-defined soft tissue mass in left paravesical area indenting the bladder wall. After some time contrast-enhanced computed tomography abdomen followed by magnetic resonance imaging lower abdomen were done which showed left paravesical enhancing soft tissue mass measuring 4.5 cm × 3.0 cm in size [Figures 1 and 2], indenting the wall of urinary bladder with no evidence of surrounding infiltration, doubt of paravesical paraganglioma was raised. There were no enlarged loco regional nodes. Furthermore, lab investigations were performed directed towards paraganglioma work up including serum cortisol, urinary vanillylmandelic acid, metanephrine (serum and urinary) with the results and reference ranges shown in Table 1.

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How to cite this article: Syed Z, Chaturvedi AK, Bashir W, Sunaina D. Symptomatic pelvic paraganglioma in an adult. Onc Gas Hep Rep 2016;5:26-8.
All the tests were within normal limits but the patient was apparently symptomatic with irritable bladder and palpitations during and after voiding.

Patient underwent robotic excision of the mass. Intra-operative and postoperative periods were uneventful. Patient is doing well for 1 month. He is on regular follow-up. Histopathology examination revealed encapsulated tumor with tumor cells having round nuclei with vesicular chromatin. Tumor cells express synaptophysin, S100 highlights the sustentacular cells. Immunostain for CK is negative [Figures 3 and 4]. Findings are consistent with paraganglioma.

### Table 1: Laboratory results

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
<th>Reference range</th>
<th>Method</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum cortisol (µg/dL)</td>
<td>30.22</td>
<td>5.2-35.0</td>
<td>CLIA</td>
</tr>
<tr>
<td>Urinary vanillylmandelic acid (mg/day)</td>
<td>4.8</td>
<td>&lt;13.6</td>
<td>Ion exchange resin</td>
</tr>
<tr>
<td>Serum metanephrine (pg/mL)</td>
<td>27.6</td>
<td>&lt;90</td>
<td>EIA</td>
</tr>
<tr>
<td>Urinary metanephrine (µg/day)</td>
<td>137.36</td>
<td>&lt;350</td>
<td></td>
</tr>
</tbody>
</table>

**DISCUSSION**

In this case report we presented a middle aged man belonging to high altitude area who came to our hospital (a cancer hospital) for the evaluation of paravesical pelvic soft tissue mass. He was mildly symptomatic for about 6 years, the symptoms appeared after voiding urine. Patient was operated and postoperative histocytogy was in favor of a paraganglioma. Preop laboratory investigations were within normal limits. The emphasis is on suspecting the disease. Patient symptoms should be taken seriously. Probably the best time to take blood was when the patient is symptomatic that is, after voiding, though more study is needed in this regard.

In 2004 the World Health Organization defined a pheochromocytoma as an intra-adrenal paraganglioma, whereas closely related tumors of extra-adrenal sympathetic or parasympathetic paraganglia are classified as extra-adrenal paragangliomas. In general, about 80% of pheochromocytomas are located in the adrenal medulla.¹

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**Figure 1:** Noncontrast T1-weighted magnetic resonance imaging showing soft tissue mass in left parvesical region

**Figure 2:** Contrast-enhanced computed tomography axial section showing enhancing left parvesical soft tissue mass

**Figure 3:** Tumor cells expressing chromogranin (DAB; ×200)

**Figure 4:** Symptomatic pelvic paraganglioma in an adult (a), tumor cells expressing chromogranin (DAB; ×200) (b), tumor cells expressing synaptophysin (DAB; ×200) (c), sustentacular cells highlighted by S-100 [arrow] (DAB; ×200)
Extra-adrenal sympathetic paragangliomas in the abdomen most commonly arise from chromaffin tissue around the inferior mesenteric artery (the organ of Zuckerkandl) and aortic bifurcation, less commonly from any other chromaffin tissue in the abdomen, pelvis, and thorax. Extra-adrenal parasympathetic paragangliomas are most commonly found in the neck and head.

Main signs and symptoms of catecholamine excess include hypertension, palpitations, headache, and sweating.

**CONCLUSION**

Only few cases of paragangliomas have been reported in an adults in pelvic region. It is extremely rare. This case was clinically symptomatic but with apparent normal serum and urinary metanephrine (metabolic products of catecholamines). Samples should be taken with great care and especially when the person is symptomatic like during/after voiding in this case. Release of catecholamines is paroxysmal and may be triggered by some physical strain like contraction of the detrusor muscle in this case.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**


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