

Bladder Paraganglioma- Always be ready for crisis even if non functional

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Abstract

A 62 years old hypertensive lady was incidentally detected to have a paravesical mass during evaluation of upper abdominal pain. Imaging and cystoscopy revealed extramucosal mass lesion in the dome of the bladder. She underwent partial cystectomy and intraoperatively had hypertensive crisis with features of cardiac ischemia. She was resuscitated and mass was completely excised. Her postoperative period was uneventful. The histopathology revealed paraganglioma of the bladder. This case report is to emphasize the need for active intraoperative monitoring during paravesical mass excision, especially in hypertensives even if they are nonfunctional.

Keywords: Paraganglioma, Extra-adrenal, Hypertension, Urinary bladder, Pheochromocytoma.

Case Report

Extra-adrenal pheochromocytomas are called as paragangliomas. They are rare and constitutes approximately 10% of total pheochromocytomas.¹ Bladder paragangliomas are still rare but usually they are functional and symptomatic.² Few cases were reported to be silent and nonfunctional.³ We report a case of bladder paraganglioma which was non functional on evaluation but showed functional activity during intra-operative manipulation.

A 62 year old multiparous postmenopausal woman has presented with upper abdominal pain of three months duration. She had no history of hematuria or lower urinary tract symptoms. She was hypertensive for past 10 years and well controlled with amlodipine 5 mg once a day. Her complete hemogram and renal function tests were within normal limits. Ultrasound examination showed a well circumscribed mass in the dome of urinary bladder with normal upper tracts. The mass was further categorized using magnetic resonance imaging (Image 1- panel A). She underwent diagnostic cystoscopy which showed extrinsic compression at the dome of the bladder otherwise mucosa and ureteric orifices were normal. Her 24 hour urine catecholamines were within normal limits. She was taken up for exploratory laparotomy.

Well circumscribed tumor of size 6 x 5 x 3 cm was noted to arise from the dome of the bladder (Image 1- panel B). Partial cystectomy was done in view of suspected benign pathology. Intraoperatively during handling of the tumor, blood pressure levels rose to 210/110 mmHg. Myocardial ischemia was noticed with ECG changes and raised troponin I levels (1116) pg/ml. Blood pressure was controlled using intravenous sodium nitroprusside and nitroglycerine infusion. Postoperatively patient had persistent hypotension which needed inotropic support for one day. Her

troponin I levels reduced to 4.7 pg/ml on the postoperative day 2. Otherwise her postoperative period was uneventful. Cut surface of the tumor showed grayish white and yellow solid tissue with microcysts filled with clear fluid. Microscopic examination revealed large polyhedral cells arranged in cellular lobules with tumor origin from muscular layer while mucosa was unremarkable (Image 2- panel A,B). Immunohistochemistry revealed paraganglioma of the bladder with stains positive for neuron specific enolase and negative for cytokeratin (Image 2- panel C, D). Additional stains were positive for S 100, chromogranin, synaptophysin and vimentin confirming the origin of tumor from neural crest cells (Image 3- panel A, B, C, D). At three months follow up she was normotensive with normal urinary catecholamines.

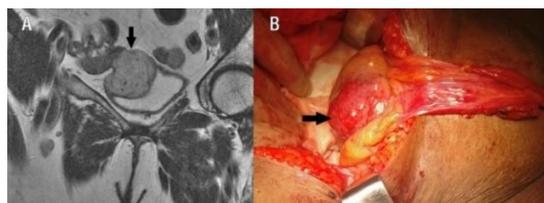


Fig. 1

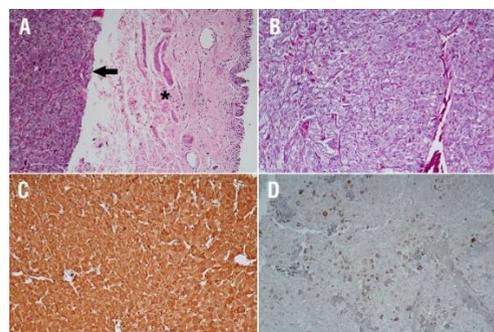


Fig. 2

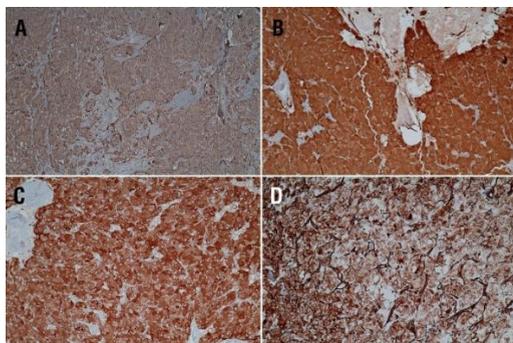


Fig. 3

Discussion

Bladder is the most common site of genitourinary tract paragangliomas (80%) followed by urethra and upper tracts.⁴ In urinary bladder, dome followed by trigone are the most common sites of involvement.² About 15% of bladder paragangliomas are non-functional and asymptomatic.² On cystoscopy, most of the tumors show submucosal bulge without mucosal involvement.² Though computerized tomography and magnetic resonance imaging are useful in tumor localization and finding metastasis,¹³¹ Iodine metaiodinebenzylguanidine (MIBG) is helpful in functional localization of pheochromocytoma.⁴ Symptomatic and suspected lesions should have metabolic evaluation by plasma or urine metanephrines preferably by plasma levels as they are more sensitive and specific.⁵ In functional tumors, preoperative evaluation is the key to successful perioperative management. Roizen et al. proposed a set of criteria to objectively assess the efficacy adequate preoperative alpha blockade.⁶ Tumor manipulation of unprepared patient can lead to hypertensive crisis intraoperatively

which can be difficult to manage because of volume contracted status of the patient and unpreparedness of the anesthesia team. In our case fortunately hypertensive crises was managed by nitroglycerine infusion and fluid boluses but sometimes it can be lethal. Fluctuating glycemic status, electrolyte imbalances and postoperative hypotension mandates intensive care monitoring in all suspected pheochromocytomas.⁷ We stress the need for preparedness of surgeon and anesthesia team for hemodynamic crisis even in pre-operatively non functional paraganglioma. Hence in hypertensive patients with submucosal bladder lesions, even if it is non functional, preoperative optimization, intraoperative vigilance and postoperative monitoring is necessary for better outcome.

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