DOI: 10.7860/JCDR/2015/14306.6420

Case Report

Surgery Section

Paraganglioma of Urinary Bladder Presenting as An Early Preeclampsia with Successful Perinatal Outcome After Surgery: A Case Report and Review of Literature

KAJE YOGESH DATTATRYA[1], TANWAR HARSHAWARDHAN VEDPALSINGH[2], SHELKE UMESH RAVIKANT[3], SINGH ABHISHEK GAJENDRA[4], PATWARDHAN SUJATA KIRAN[5]

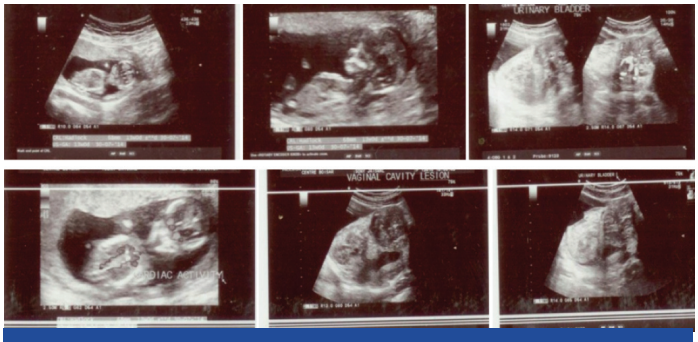
ABSTRACT

Paraganglioma in urinary bladder associated with pregnancy is extremely rare with a prevalence of less than 1%. We report a case of 25-year-old pregnant female who presented with gross haematuria and clot retention during first trimester. MRI pelvis showed a mass antero- inferior to bladder. Transurethral resection biopsy revealed paraganglioma of the urinary bladder. Her workup showed neither raised plasma free normetanephrine levels. Patient underwent partial cystectomy during second trimester. Postoperatively, she is normotensive with normal serum free normetanephrine levels. At term, she delivered a healthy female child. This case highlights a successful perinatal outcome with timely intervention, adequate preoperative control of hypertension and counselling. We report a case of paraganglioma of urinary bladder presenting as an early preeclampsia with successful perinatal outcome after surgery.

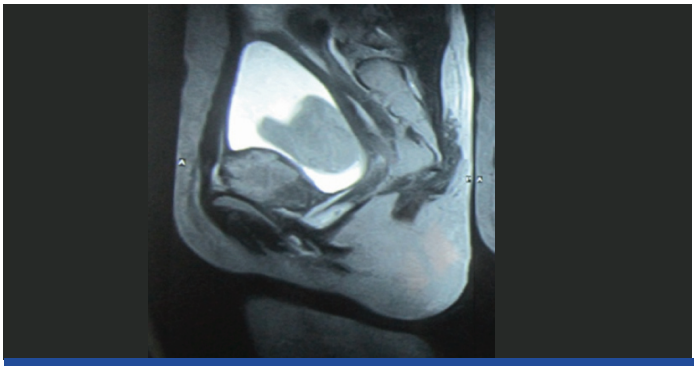
Keywords: Transurethral resection, Free normetanephrine, Partial cystectomy

CASE REPORT

A 25-year-old woman, presented with a history of painless haematuria. She was G2P1L0A1 with 9 weeks 2 days of gestation at presentation. Haematuria was gross, continuous and was associated with passage of blood clots. There was no history of any preceding trauma, surgical procedure done or addiction to smoking or tobacco chewing. Family history was not contributing. On examination, she had pallor. Her BP was 160/100 mm of Hg. Per abdominal examination was unremarkable except the palpable gravid uterus. Haemoglobin was 7.5 mg% with other parameters were normal. Her renal function test, liver function test and coagulation assay was normal. Ultrasonography (USG) Pelvis showed heterogeneously hypo-echoic, well defined mass lesion anterior to vaginal cavity involving inferior wall of urinary bladder and protruding into the lumen of it. It measured 6.5 × 5.5 cm in size, had significant vascularity on Doppler. A 10.8 × 5.6 cm hyper-echoic lesion probably a blood clot in bladder was also seen [Table/Fig-1]. Magnetic Resonance Imaging (MRI) of pelvis showed a large soft tissue mass of about 5.3 × 4.8 × 5.4 cm on left side pelvis, anterior inferior to bladder with infiltration of its wall. A blood clot 10 × 5 cm was seen in bladder lumen [Table/Fig-2]. On cystoscopy, there were clots in bladder, which were evacuated. There was 6x5 cm mass arising from anterior-superior wall whose transurethral resection (TUR) biopsy was taken. Procedure was uneventful. On histopathological analysis lesion turned out to be paraganglioma of the urinary bladder infiltrating the lamina propria. On immunohistochemistry, it showed synaptophysin and Chromogranin A positivity. Patient's history was reviewed again. It was found out that she had an abortion during last pregnancy during first trimester because of preeclampsia six months back. In addition, there was history of palpitations, headache, sweating and micturition syncope. Her endocrine work-up revealed neither raised plasma free nor- metanephrine levels (1404 pg/ml). Patient's blood pressure was controlled with tablet Prazocin 10 mg/day, tablet Amlodepine 5mg/day and tablet, Atenolol 50 mg/day and strict monitoring was done for same. After proper counselling, decision of watchful waiting was taken to allow foetal maturity. Patient underwent partial cystectomy during second trimester. Foetal viability was confirmed postoperatively. Postoperative period was uneventful. Her blood pressure was normal post resection of bladder tumour. At term, she delivered a healthy female child by caesarean section because of prolonged first stage. Patient had regular follow-up and on follow-up, she was normotensive and her post partum serum free normetanephrine levels were normal. She had mild urinary frequency, urgency with urge incontinence. She is presently on anticholinergics. She voids around 150-200 ml at each void as suggested by her voiding diary.



[Table/Fig-1]: Ultrasound Pelvis showing a mass lesion anterior to vaginal cavity involving inferior wall of urinary bladder and protruding into the lumen with significant vascularity. Note the viable fetus and a blood clot in bladder



[Table/Fig-2]: MRI pelvis showing a large soft tissue mass anterior inferior to bladder infiltrating its wall. A blood clot 10 x 5 cm was seen in bladder lumen

DISCUSSION

Paraganglioma is a neuroendocrine neoplasm, occurring at various sites in the body. Ninety seven percent of paraganglioma are benign while remainder are malignant. Extra-adrenal phaeochromocytomas are known as paragangliomas. The majority of extra-adrenal tumours occur intra abdominally (85% occur below the diaphragm) along the sympathetic chain or from the organ of Zuckerkand. Prevalence of paraganglioma in urinary bladder (UB) is less than 1% and those associated with pregnancy are extremely rare [1]. In the genitourinary tract, the UB is the most common site (79.2%), followed by the urethra (12.7%), pelvis (4.9%), and ureters (3.2%) [1]. It arises from the chromaffin tissues of the sympathetic nervous system present in the wall of the UB. Primary paraganglioma of the urinary bladder is rare, making up less than 0.05% of all bladder neoplasms [2]. They are most commonly situated at the dome or the trigone of the bladder [3]. The spectrum of presentation varies from painless haematuria, micturition syncope, hypertensive crisis, palpitations, anxiety, sweating and hot flushes. Preoperative diagnosis is usually suspected on the grounds of the typical clinical symptoms and sign and further confirmed by measurement of free normetanephrine levels in both urine and plasma [4]. Paraganglioma associated with pregnancy may present with a spectrum of presentation such as pre-eclampsia, palpitations, sweating, pallor, orthostatic hypotension and glycosuria and hypertension that may be episodic. Paragangliomas and phaeochromocytomas are rare causes of hypertension and only 0.1-1% of all cases of hypertension are due to these tumours. The prevalence of paragangliomas and phaeochromocytomas during pregnancy is five per million [5]. The diagnosis is often missed during pregnancy, as the clinical picture mimics that of pre-eclampsia, leading to potentially lethal complications for both the mother and the foetus [6]. Malignancy is difficult to diagnose in paraganlioma as there are no reliable definitive histological characteristics to distinguish between bening and malignant tumours [7]. Surgery is the definitive treatment of choice for these tumours, with appropriate alpha-adrenergic and if needed, subsequent beta-adrenergic blockade to prevent a hypertensive crisis. In both pregnant and non-pregnant individuals with catecholamine secreting tumours Phenoxybenzamine is the alpha blocker of choice [8]. Radical cystectomy, partial cystectomy and transurethral resection are the reported treatment options for localized or locally advanced disease. The above therapies have shown good survival rates, approximately only 3 % of patients with reported follow up died due to their cancer, however, over 20% of patients had recurrence or metastases at the last known follow-up [9]. Surgical treatment is rarely curative in the face of metastatic paraganglioma. However, by reducing co morbid conditions (i.e. hypertension) and reducing tumour burden, it may adequately prolong survival but adjunct therapies are usually indicated. Thus, counselling of the patients should be done according to their individual presentation and disease status. On review of the literature it was found that many centres believe in terminating the pregnancy electively before proceeding with partial cystectomy in such circumstances. A good outcome with vaginal delivery has been described in only few selected cases [8].

CONCLUSION

In our case, we had taken patient for clot evacuation and TUR biopsy was done in view of USG findings, keeping carcinoma of bladder in mind. Despite of its functional nature we had no hypertensive crisis intraoperatively. Thus, we diagnosed paraganglioma retrospectively. Although rare, it should be included in the differential diagnosis of early preeclampsia. Contrary to the routine, we excised the paraganglioma safely with continuation of pregnancy. Adequate preoperative control of hypertension, detailed counselling of patient's family and Endocrinologist's expertise proved crucial in our success.

Early diagnosis can be made by characteristic cystoscopic appearance though this has not been specifically investigated in the literature till date. Due to the rarity of these tumours, the clinical features and the cystoscopic appearance need to be assessed and such case reports documented in the future.

REFERENCES

[1] Demirkesen O, Cetinel B, Yaycioglu O, Uygun N, Solok V. Unusual cause of early preeclampsia: bladder paraganglioma. *Urology*. 2000;56(1):154.
[2] Dahm P, Gschwend JE. Malignant non-urothelial neoplasms of the urinary bladder: a review. *European Urology.* 2003;44:672-81.
[3] Ansari MS, Goel A, Goel S, Durairajan LN, Seth A. Malignant paraganglioma of the urinary bladder. A case report. *Int Urology and Nephrology.* 2001;33:343-45.
[4] Jurascheck F, Egloff H, Ruemi A, Laedlein-Greilsammer D. Paraganglioma of urinary bladder. *Urology.* 1983;22(6):659-63.
[5] Kalra JK, Jain V, Bagga R, Gopalan S, Bhansali AK, Behera A, et al. Pheochromocytoma associated with pregnancy. *J Obstet Gynaecol Res*. 2003;29(5):305–08.
[6] Pearson GAH, Eckford SD, Levy A. A reason to panic in pregnancy. *Lancet.* 2009;374(9691):756.
[7] Cheng L, Leibovich BC, Cheville JC, Ramnani DM, Sebo TJ, Neumann RM, et al. Paraganglioma of the urinary bladder: Can biologic potential be predicted? *Cancer.* 2000;88(4):844-52.
[8] Wald O, Shapira OM, Murar A, Izhar U. Paraganglioma of the mediastinum: challenges in diagnosis and surgical management. *J Cardiothorac Surg*. 2010;5:19.
[9] Kempasiddaiah GM Thimmaiah R, Desai MG. A rare case of early pregnancy with paraganglioma of urinary bladder. *IJCRR*. 2014;6(4):15-20.

PARTICULARS OF CONTRIBUTORS:
1. Registrar, Department of Urology, Seth G.S. Medical College and KEM Hospital, Mumbai, Maharashtra, India.
2. Registrar, Department of Urology, Seth G.S. Medical College and KEM Hospital, Mumbai, Maharashtra, India.
3. Registrar, Department of Urology, Seth G.S. Medical College and KEM Hospital, Mumbai, Maharashtra, India.
4. Ex-Registrar, Department of Urology, Seth G.S. Medical College and KEM Hospital, Mumbai, Maharashtra, India.
5. Head of the Department, Department of Urology, Seth G.S Medical College and KEM Hospital, Mumbai, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Tanwar Harshawardhan Vedpalsingh,
Registrar, Department of Urology, New Building, Seth G.S. Medical College and KEM Hospital,
E Boreges Marg, Parel, Mumbai, Maharashtra-400012, India.
E-mail: harsh_bj1@rediffmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Apr 03, 2015**
Date of Peer Review: **May 21, 2015**
Date of Acceptance: **May 28, 2015**
Date of Publishing: **Sep 01, 2015**