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Case Report

Surgical Management Paravesical Paraganglioma Using a Robotic Platform

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Abstract

Paraganglioma/ Paragangliomas is/ are the rare tumors arising from peripheral nervous system and are also called as extra- adrenal pheochromocytoma. Pelvic location of these tumors is rare (2%), occurring commonly at the organ of Zuckerkandl. Isolated paravesical paraganglioma in the retro-pubic space has not been reported till date. Herein we report a case of pelvic para-vesical paraganglioma, which was surgically excised using robotic platform.

Keywords: Paraganglioma; Paravesical Mass; Robotic Surgery

Introduction

Paraganglioma/ Paragangliomas is/ are the rare tumors arising from peripheral nervous system and are also called as extra-adrenal pheochromocytoma. Paraganglioma may be functional or non-functional depending upon whether it produces either of adrenaline, nor-adrenaline, both or none of these. Paraganglioma arising from the organ of Zuckerkandl produce adrenaline, rest of the Paragangliomas are either non-functional or produce only noradrenaline. Uncontrolled secretion of noradrenaline can be life threatening and can lead to myocardial infarction, stroke and death. Risk of malignancy is higher with paragangliomas compared to adrenal pheochromocytoma (around 40-50%) [1]. Size of tumor>5cm, infiltration of adjacent tissue, metastases are the features suggestive of malignant Paraganglioma. Most common intra-abdominal location of Paraganglioma include organ of Zukerkandl, which is near bifurcation of the aorta or paravertebral along the aorta [2].

Paraganglioma has been described at various locations in pelvis, like in bladder, prostate and ovary. Paraganglioma can be sporadic (more common) or may occur in association with some inherited diseases such as, von Hippel Lindau syndrome, mutation of succinate dehydrogenase (SDH) gene, multiple endocrine neoplasia 2. Paraganglioma may also occur with simultaneous pheochromocytoma. Herein we report a case of pelvic para-vesical paraganglioma, which was surgically excised using robotic platform .

Case Report

44 years old, hypertensive male, complained of poor urinary flow, hesitancy, burning micturition for 2 months. On Ultrasound (USG) of Kidney urinary bladder, he was found to have pelvic mass in relation to left antero-inferior wall of urinary bladder,

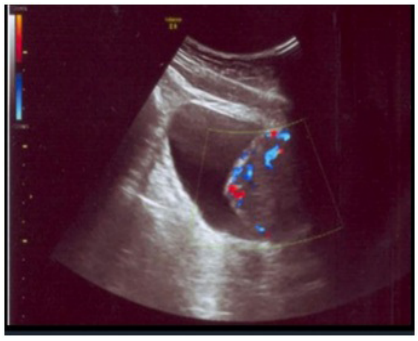

Figure 1: Ultrasound image of the pelvic mass.

A contrast enhanced computed tomography (CT) of abdomen and pelvis revealed well-defined, soft tissue density lesion in paravesical space on left side measuring 57*52*52 mm (Figure 2).

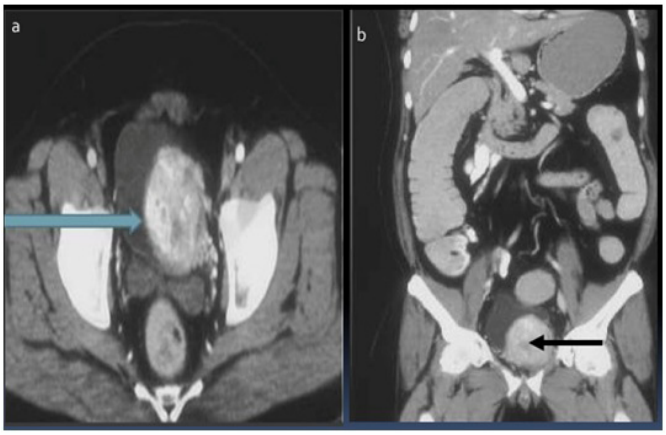

Figure 2 a-b: CT images of the pelvic mass.

Lesion was abutting left antero-lateral wall of urinary bladder with possible area of infiltration of urinary bladder wall, with no definite intraluminal extension. Lesion was seen abutting the left lobe of prostate without any invasion. The lesion was homogenously enhancing from 25 to 45Hounsfield units (HU). Wash out study was not done. On cystoscopy, there was no intra-luminal growth, but an indentation on the left lateral wall could be appreciated (Figure 3).

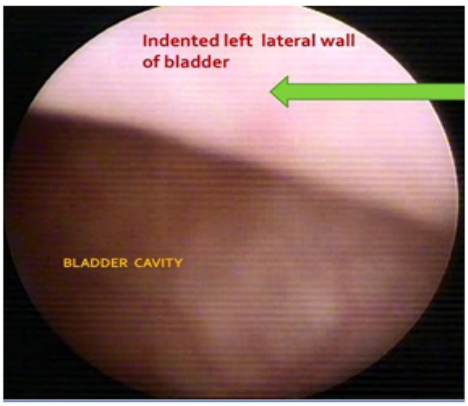

Figure 3: Cystoscopy image showing no vesical infiltration, but indentation.

Trans-rectal ultrasound (TRUS) and Intra cavity ultrasound was done which suggested that the mass was neither infiltrating the rectal wall nor the bladder wall. TRUS revealed round mass separate from bladder, prostate and seminal vesicles. A USG guided transabdominal biopsy of paravesical mass was done, which was suggestive of paraganglioma, immune-histochemical markers chromogranin, synaptophysin and S 100 were positive. Plasma free metanephrins and nor-metanephrins were subsequently measured and found to be within normal range suggesting that, it was a non-functioning paraganglioma.

Patient underwent surgical excision of the mass using a robotic platform, Da Vinci Si TM system (Intuitive Surgical, Sunnyvale, CA, USA). Patient was placed in trendelenburg position with 5 ports placed in fan shaped manner along with an additional 12mm assistant port. The bladder was dropped down and the retro-pubic space dissected, pelvic mass was identified in the retro-pubic space. Dissection was started from left side and continued on right side (Figure 4a,b).

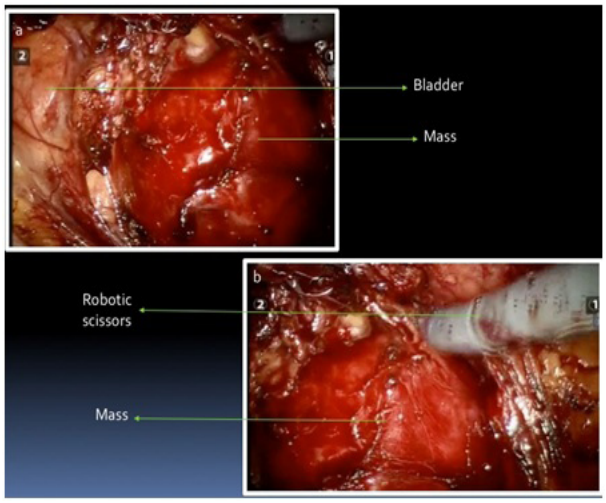

Figure 4a-b: Intraoperative Images.

The mass was separated all around and the surrounding tissue was serially clipped using Hem-o-lok™ (Teleflex, Morrisville, NC) clips or coagulated using Ligasure™ vessel sealing system (Medtronic, Minneapolis, USA). Mass was excised and retrieved through camera port site by extending the incision. There was no fluctuation of blood pressure on handling the mass. On gross pathological examination, weight of specimen was 55 grams and it was 5.0x4.7x4.0cm in dimension. On bisecting the specimen, it showed soft pinkish brown tumor covered by thick capsule (Figure 5a).

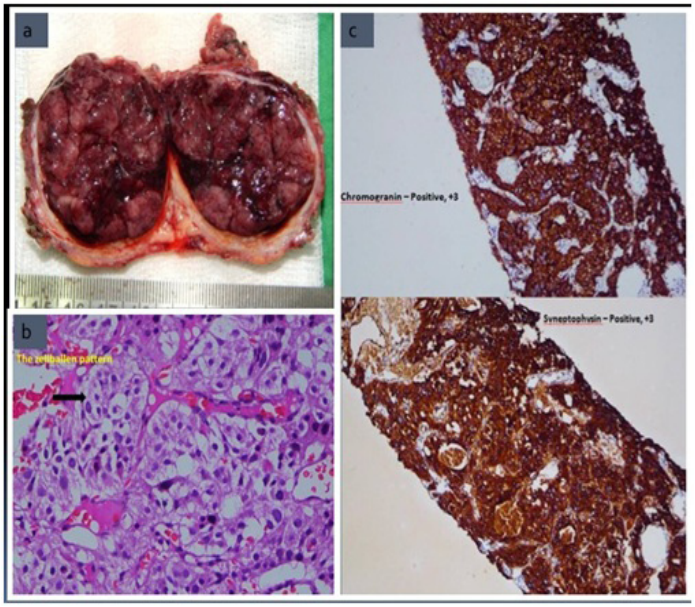

Figure 5a-d: Figure 5a showing gross pathology, Figure 5b showing microscopic pathology and Figure 5c & d showing Immune-histo-chemistry.

On microscopy, the tumor cells were round with acidophilic granular cytoplasm and they had ovoid nuclei with fine granular chromatin, arranged in discrete nests (the zellballen pattern) separated by a prominent vascular network (Figure5b). These tumor cells were positive for chromogranin, S100 and synaptophysin staining (Figure5c). These cells were arranged in zellballen pattern and the nests were separated by silver-positive fibrous septum.

Postoperative course was uneventful; patient was discharged on postoperative day 4 and at 6 months followup there was no recurrence on ultrasound examination and patient was asymptomatic.

Discussion

Primary pelvic retroperitoneal masses have characteristic natural histories, radiological and histopathological features. Contrast enhanced CT scan and high resolution MRI gives important information regarding tumor location, its vascularity, its relation with or invasion of adjacent structures, this helps in surgical planning. Primary pelvic retroperitoneal masses are the rare entities than more commonly occurring lymphadenopathy, abscess, hematoma and extension of sacral, ureteric, rectal diseases. These rare masses pose a diagnostic dilemma, as their anatomy and the pathology are unfamiliar. Extra adrenal pheochromocytomas (paragangliomas) account for 10 % of paragangliomas. Pelvic location of these tumors is rare (2%), occurring commonly at the organ of Zuckerkandl. Paraganglioma of organ of Zuckerkandl can present as pelvic retroperitoneal mass, but these masses are higher up in pelvis. Paragangliomas arising from bladder are also known and account for less than 0.06 percent of bladder tumors and less than 1 percent of all paragangliomas [3]. Isolated paravesical paraganglioma in the retro-pubic space has not been reported till date. Traditionally these masses have been managed by open surgical excision. In this case, we used the advantages of the robotic platform to operate deep in the pelvis and excised the paraganglioma.

Paragangliomas may present with palpitation, headache, diaphoresis, hypertension due to elevated catecholamine production and non-functional paraganglioma may manifest as abdominal pain or symptoms due to mass effect on adjacent structures. Generally adrenal pheochromocytomas are benign but a third of paragangliomas harbor malignancy. Elevated levels of serum and urinary nor-ephinephrine are highly diagnostic of functional paraganglioma [4].Cross sectional imaging like CT; MRIs are helpful for detection, nature and surgical planning of these tumors. They show low intensity on T1, high intensity on T2 image, with enhancement on gadolinium contrast administration [5]. I-123 Meta-iodo-benzylguanidine (MIBG) scan can identify extra-adrenal paraganglioma.

Without adequate biochemical and anatomical evaluation, percutaneous biopsy of such hyper vascular pelvic tumors can lead to hypertensive crisis. In our case, we were not suspecting the pelvic mass to be paraganglioma and hence biopsy was done. Surgeons should keep low threshold for doing serum and urinary nor-metanephrin levels for isolated pelvic masses before proceeding for biopsy or surgical resection as it may have a huge bearing on the management. Preoperative preparation of the patients with metabolically active paragangliomas includes blood pressure control using non-selective alpha-blockers with beta-blockers and salt loading. Malignant nature of this disease cannot be made out even on histology. Malignant nature of this tumor can only be diagnosed once metastasis to an organ other than the one harboring paraganglioma, is established.

Conclusion

Paravesical paraganglioma is rare condition; symptoms may arise due to its biochemical secretions or due to shear mass effect. Excision of these masses using robotic platform is feasible and the advantages of using robotic platform for operating in deep pelvic cavity can be best utilized in such cases.

References

1. Bosch FJ, Goedhals J, De Lange W, Ackermann M, Koning JM (2012) A pelvic paraganglioma presenting as a hypertensive emergency in pregnancy. Journal of Endocrinology, Metabolism and Diabetes of South Africa 17: 145-147.
2. Shanbhogue AK, Fasih N, Macdonald DB, Sheikh AM, Menias CO, et al (2012) Uncommon primary pelvic retroperitoneal masses in adults: a pattern-based imaging approach. Radiographics 32: 795-817.
3. Chang YK, Chiang IN, Chen CH, Wang SM, Lee YJ, et al (2015) Paragangliomas of the urinary bladder: A report of 6 cases and review of the literature. Urological Science 26: 111-114.
4. Geng JH, Huang SP, Shen JT, Chen YT (2014) Paraganglioma of the urinary bladder with pelvic metastasis. Urological Science 25: 91-94.
5. Blake MA, Kalra MK, Maher MM, Sahani DV, Sweeney AT, et al (2004) Pheochromocytoma: An Imaging Chameleon 1. Radiographics 24: 87-99.