An Unusual Cause of Urinary Retention: A Case Report

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Pelvic paraganglioma is a rare neuroendocrine tumor with variable presentation depending upon whether it is functional or not. We report such a case of young male who presented to us with intriguing cause of urinary retention and diagnostic challenges that we faced during management of this patient, later found out to have non-functional pelvic paraganglioma.
Keywords: extra-adrenal paraganglioma, pelvic sarcoma, pheochromocytoma.

Paraganglioma is a rare tumor. Since the clinical patterns of paragangliomas are commonly described together with those of pheochromocytoma, the specific incidence of paraganglioma is largely unknown. The combined estimated annual incidence of pheochromocytoma/paraganglioma is approximately 0.8 per 100,000 person years.[1] We present a case report of extra-adrenal paraganglioma in a young man who presented initially with acute retention of urine highlighting the challenges we faced during the management of this patient.

Case summary

30 years old, thinly built, pale, normotensive, male presented to emergency of other center with acute retention of urine following which foley catheterization was done and 1 liter of urine drained. A huge mass of around 14 cm was palpable after emptying of the bladder, in the supra pubic region. Contrast enhanced computed tomography (CECT) **(Figure 1)** of abdomen, pelvis and chest was done which revealed very large heterogenous tumor in the pelvis measuring 14.4x10.7x16.6cm demonstrating peripheral serpiginous enhancement and central non-enhancement with regions of

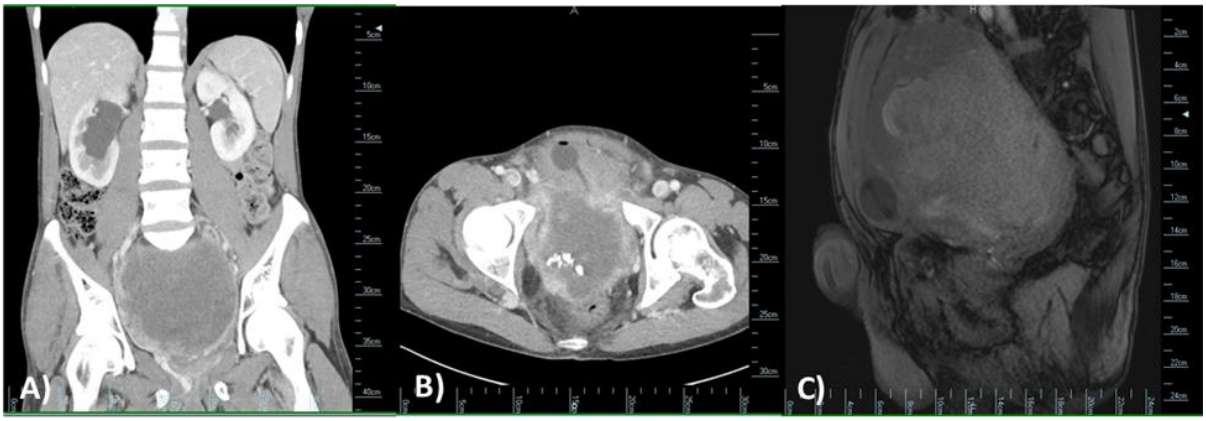

Figure 1: Contrast enhanced CT (CECT) of abdomen showing A) Large heterogenous tumor in pelvis with peripheral serpenginious enhancemen, B) Lesion does not clearly invade the surrounding adjacent structure, C) MRI of pelvis showing mass contacting posteriorly lumbosacral junction but without direct invasion

coarse calcifications, markedly compressing bladder and sigmoid colon **(Figure 1B)**. To clearly delineate the mass and the surrounding soft tissues, magnetic resonance imaging (MRI) of the pelvis was done **(Figure 1C)** which showed similar findings with added information regarding the contact of the mass to the lumbosacral vertebral body. Rectum, sigmoid colon, prostate and seminal vesicles were separate from the tumor.

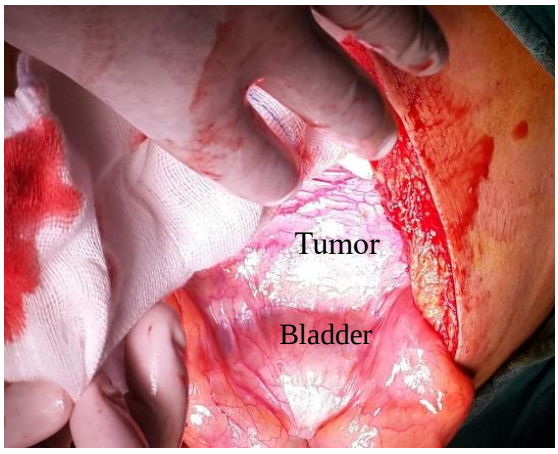

Figure 2: Intraoperative picture of the tumor posterior to the bladder

CT guided biopsy was done which was in favor of pelvic sarcoma. Patient then presented after 2 weeks to our center with Foley catheter in situ for further management.

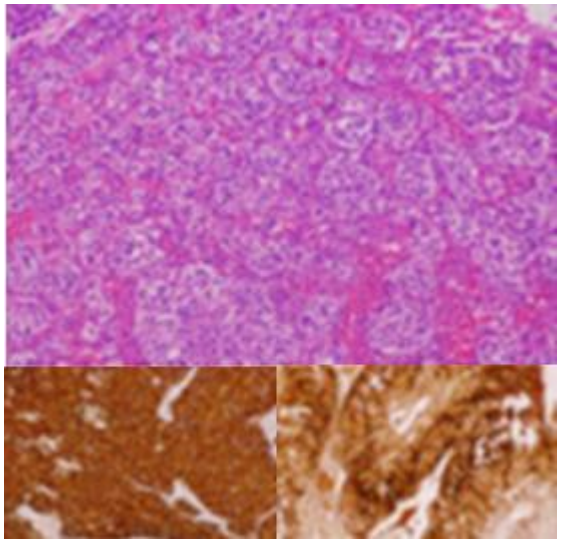

Figure 3: Histopathological picture showing Tumor arranged in nesting pattern separated by fibrovascular septae. IHC: Positive for Synaptophysin, Chromogranin, NSE, CD56

As the mass was growing in size due to internal hemorrhage evidenced by patient's sharp drop in hemoglobin, we opted for

upfront surgery. Patient was planned for preoperative angio-embolization because of highly vascular nature of the tumor followed by surgery and adjuvant chemotherapy. Main feeder artery was successfully embolised and patient taken for surgery. Preoperatively cystoscopy was done for assessment of the urinary bladder, however cystoscope could not be negotiated beyond prostatic urethra and hence skipped. Intraoperatively, we found a large well capsulated and highly vascular retroperitoneal tumor around 15cm in diameter compressing the urinary bladder anteriorly and rectum posteriorly **(Figure 2)**. Distal right ureter was involved and adherent to the tumor but plane maintained with the left ureter and rectum. Dense adhesions were present at the base of the bladder posteriorly and near prostatic urethra which led to some spillage of tumour and opening of the bladder as well. The patient finally had excision of pelvic mass (R2) with right distal ureterectomy with neoureterovesicostomy and double J stenting. Later, histopathological examination and immunohistochemistry (IHC) revealed the tumor to be extra-adrenal paraganglioma **(Figure 3)**. We could not retrieve the previous histopathological slide that had been reported as sarcoma in order to compare and confirm their findings.

Postoperatively, 24-hour urine metanephrines were sent which was normal. MRI was repeated on 3 months follow up which showed no recurrence of the tumor. Patient is currently on regular follow-up with medical oncology team.

Discussion

Paraganglia are a group of non-neuronal cells derived from the neural crest which lie in close proximity to the sympathetic ganglia. Tumor of the paraganglionic tissue are known as paragangliomas and they are made up of neuroendocrine cells. 2004 WHO classification used the term "extra-adrenal paraganglioma" to denote an extra-adrenal tumor of sympathetic or parasympathetic paraganglia origin, regardless of secretory status.[2] Presentation depend upon tumor location, catecholamine secretion, and other factors. Mostly sympathetic paragangliomas are functional with features of hypertension, episodic headache, sweating, palpitation and micturation syncope (hallmark of catecholamine secreting bladder paraganglioma).[3] Our patient had no symptoms related to catecholamine secretion however he presented with acute retention of urine due to mass effect. Preoperative 24-hour urinary metanephrines was not sent because CT guided biopsy which was done in other center was suggestive of pelvic sarcoma and also due to lack of clinical suspicion. Radiological as well as radio–isotope imaging are not diagnostic. These are highly vascular tumors and it is difficult to predict, on the basis of histologic findings, whether a paraganglioma is benign or malignant.[4] IHC typically confirms the diagnosis with strong positivity for neuron-specific enolase (NSE), synaptophysin and chromogranin. Many authors believe that, biopsy or even cystoscopy may provoke hypertensive crisis and should not be done

in pelvic or urinary bladder paraganglioma.[5] In our case, both of those procedures were done as the pre-operative suspicion of paraganglioma was very low and fortunately, did not result in any adverse events.

Due to lack of robust prognostic indices of recurrence, lifelong close follow up is necessary with history and physical examination, along with radiological surveillance. Secreting tumors can be followed in addition with hormonal markers.[6]

Conclusion

Extraadrenal paragangliomas presenting as pelvic mass is very rare. Non-secreting tumors pose diagnostic challenges, whereas secreting tumors demand careful preoperative metabolic optimization.

Conflict of Interest

None

References

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