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Case Report

Oncology Section

Non-Functional Paraganglioma of Retroperitoneum Mimicking Pancreatic Mass with Concurrent Urinary Bladder Paraganglioma: An Extremely Rare Entity

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ABSTRACT

Paragangliomas are extra-adrenal tumours of the autonomic nervous system, which rarely present as primary retroperitoneal mass mimicking pancreatic malignancy (incidence 2-8 per million populations). Urinary Bladder Paraganglioma are also extremely rare (0.06% of all Bladder Tumour and 6% of Paragangliomas) with most being malignant and high grade tumours. Non-functional varieties of both tumours are usually incidentally diagnosed. The possibility for malignant transformation in them makes surgical excision the treatment of choice. A 45-year-old lady with abdominal pain was investigated to have a complex retroperitoneal mass behind head of pancreas and a urinary bladder mass. Complete excision of retroperitoneal and bladder lesion was done. Histopathological examinations of both specimens were suggestive of Paraganglioma with no abnormal mitotic activity and capsular/vascular invasion. Although concurrent non functional paragangliomas had been reported but the synchronous non-functional paragangliomas of retroperitoneum and urinary bladder reported in this case is extremely rare and is not reported so far in English literature.

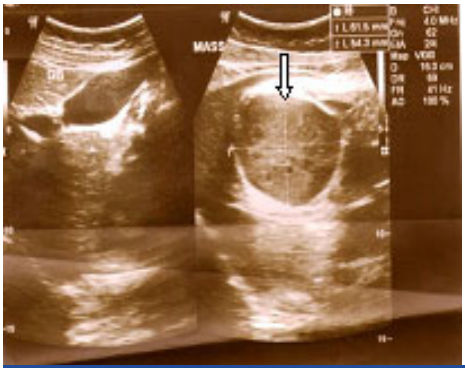
Keywords: Excision, Pancreas, Retroperitoneal mass, Synchronous

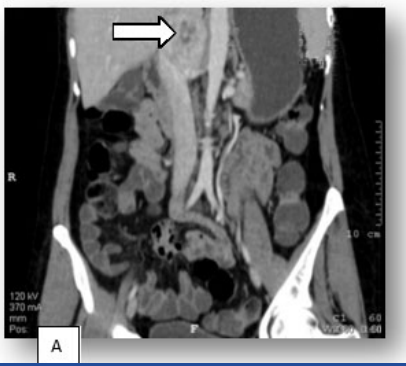
CASE REPORT

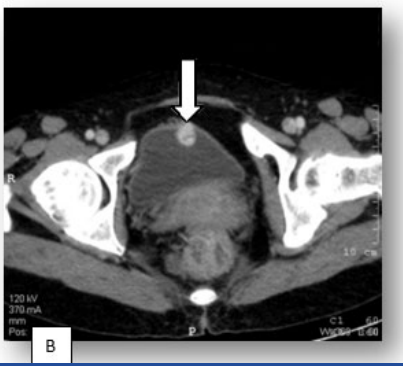
A 45-year-old lady presented with pain in central abdomen and occasional headaches for 2 years. Her family history was unremarkable and she had no previous medical and urinary problem. General and systemic examination was normal. On ultrasound abdomen 6 x 6 cm solid iso-echoic lesion was seen in the retroperitoneum abutting periportal and peripancreatic region displacing hepatic artery and portal vein anteriorly [Table/Fig-1]. CECT abdomen demonstrated 6.5 x 5.4 cm well capsulated lesion in portocaval region with increased enhancement in arterial phase and relative washout in porto-venous phase with central necrotic areas. The portal vein and head of pancreas were displaced anteriorly with no evidence of calcification and metastases [Table/Fig-2a,b]. A 2 x 1.5 cm similar enhancing lesion in anterior bladder wall was present. Urinary metanephrines were normal. Patient underwent cystoscopy and revealed 2 x 2 cm highly vascular lesion in the anterior wall of urinary bladder with normal mucous covering. During exploratory laparotomy a single 6 x 6 cm solid highly vascular mass was seen in the aorto-caval groove without regional lymphadenopathy. Inferior vena cava was compressed with anterior displacement of portal vein and head of pancreas but no invasion into surrounding structures [Table/Fig-3a-c]. Complete excision of retroperitoneal and bladder lesion was done. There was no preoperative or intraoperative fluctuation of blood pressure. Pathological evaluation revealed paraganglioma to be the final diagnosis with no abnormal mitotic activity and capsular/vascular Invasion [Table/Fig-4a,b].

DISCUSSION

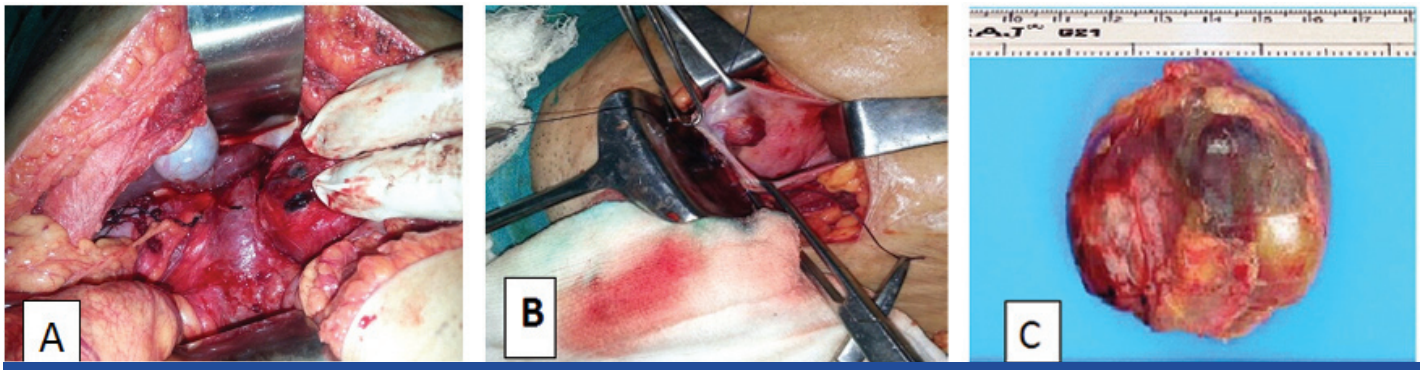
Paragangliomas are extra-adrenal chromaffin [1] tumours that develop from the neuroectodermal cells of the autonomous nervous system. Closely related tumours in extra-adrenal sympathetic and parasympathetic paraganglia are classified as extra-adrenal paragangliomas [2,3]. Middle aged men and women were equally affected in most series [4]. Genetic mutation in genes encoding succinate dehydrogenase enzyme had also been observed [5]. Patients with non-functioning retroperitoneal paraganglioma are usually rare and asymptomatic until the tumour reaches sufficient size to produce symptoms of compression of adjacent organs



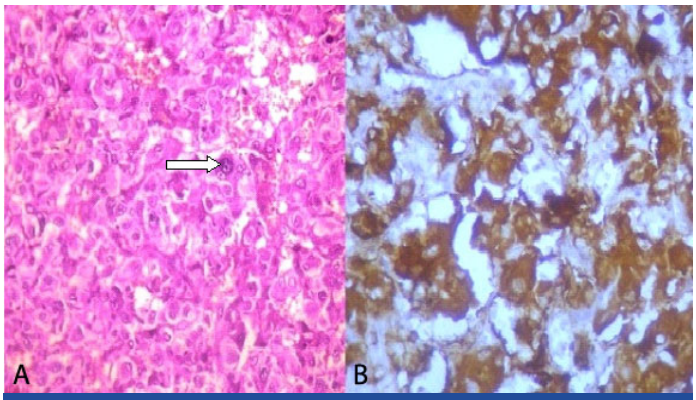




[Table/Fig-1]: Ultrasound abdomen showing retroperitoneal mass abutting peri portal and peri pancreatic region (arrow head), **[Table/Fig-2a,b]:** CECT Abdomen (A) coronal section showing the mass sandwiched between the great vessels, (B) axial view, showing urinary bladder mass (arrow heads)



[Table/Fig-3a-c]: Intraoperative findings (A) Retroperitoneal mass sandwiched between great vessels, (B) Urinary bladder mass, (C) Gross specimen of retroperitoneal mass



[Table/Fig-4a,b]: (A) Large tumour cell with granular eosinophilic cytoplasm, nests of capillary and nuclear pleomorphism or bizarre nuclei (arrow head) (H& E 400), (B) Immuohistochemical study demonstrating strong expression of chromogranin A and synaptophysin, but negative for cytokeratin

[6]. Paragangliomas of the urinary bladder account for 0.06% of all bladder tumours and 6% of extra-adrenal pheochromocytomas with most being high grade and functional in contrast to their retroperitoneal counterparts [7]. Because of low incidence rate, the prognosis of paraganglioma is not well established. Occasionally these tumours are multiple and may be associated with paraganglioma of other sites or with other tumours [3]. Typical symptoms include abdominal pain in 50% of cases (as in our case), nausea, emesis, abdominal distension and weight loss. Functional excretion of metabolites and increased vascularity causes haematuria and other classical symptoms like headache, blurred vision and palpitation especially during micturation. MRI is more sensitive than CT scan in detecting extra-adrenal tumours. Scintigraphy with I-123 labelled MIBG offers superior specificity than CT and MRI [8,9]. Immunohistochemically, neuron-specific enolase (NSE), synaptophysin, neurofilament protein and chromogranin can be demonstrated in the chief cells of paraganglioma. In addition, the delicate sustentacular network can be demonstrated using antibodies to S-100 protein and in a few instances these same cells coexpress glial fibrillary acidic protein. Paragangliomas metastasize approximately in 20% to 42% of the cases. Dissemination can be haematogenous or through the lymphatic system with most common site being the regional lymph nodes, bone, lung and liver. Thus, the possibility for malignant transformation or metastasis of Paragangliomas makes surgical excision the treatment of choice. Surgical therapy includes open or laparoscopic excision for retroperitoneal tumours, transurethral resection and partial or total cystectomy combined with pelvic lymph node dissection, especially in the presence of metastasis for urinary bladder paraganglioma [10]. Radiation therapy has been advocated in patients where surgery is not feasible or for unresectable tumours [11]. Therapy with radionucleotides (I-131 MIBG) or combination chemotherapy (cyclophosphamide, vincristine and darcabazine) may induce partial responses to malignant paraganglioma [12]. Octreotide may also be used for treatment of inoperable paragangliomas [13]. Although multiple functional and non functional Paragangliomas had been reported [2,5,14], synchronous non functional paragangliomas of retroperitoneum and urinary bladder reported in this case is extremely rare and is not found to be reported after extensive journal search. Complete surgical excision of both the lesions was performed and patient recovered well.

CONCLUSION

This case is unique in terms of having two rare synchronous non-functional paraganglioma in retroperitoneum and urinary bladder without any clinical symptoms and to best of our knowledge till date it remains first ever such reported case in literature worldwide. Our case emphasizes the necessity to include extra-adrenal paraganglioma in the differential diagnosis and management of retroperitoneal tumours including pancreatic mass, despite its rarity, with thorough metastatic workup to pickup synchronous or metachronous lesions in other organs. Imaging studies have greatly contributed to a more accurate diagnosis of paraganglioma. Yet, the gold standard for diagnosis remains a pathological one. Lifelong follow-up of patients with retroperitoneal Paragangliomas is essential as silent metastasis and recurrence may occur frequently.

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