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Retroperitoneal mucinous adenocarcinoma in a young male-diagnostic dilemma: A case report

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Abstract
The retroperitoneal tumors are commonly secondaries from ovaries, pancreas or colon. Among the primary retroperitoneal tumors, mucinous adenomas are more common than mucinous adenocarcinomas. Clinically, they may present as abdominal masses, weakness of limbs and weight loss. Radiologically, they present as cystic masses, the exact origin of which cannot be ascertained. Here, we present a case of 30 years male with symptoms of difficulty in passing urine, pain and weakness of left lower limb with thinning of the limb since 6 to 7 months and accompanied by history of erectile dysfunction. PET CT showed left mesorectal fascial thickening with loss of fat plane, indentation of left posterior bladder wall and no metabolically active Lymph node being detected.

Keywords: Retroperitoneal mucinous, male-diagnostic dilemma, abdominal masses

Introduction
Retroperitoneal primary mucinous adenocarcinoma is extremely rare and the histogenesis of this tumor remains unknown. It causes clinical symptoms only when the mass grows to a sufficiently large size. Laboratory studies lack the appropriate levels of specialization for this tumor and the imaging methods merely reveal cystic lesions, neither of which result in accurate diagnosis. Surgical resection is standard for the treatment. Chemotherapy has not been rendered an efficacious treatment modality.

Case Report
A 30 year old male with no known comorbidities, presented with complaints of difficulty in passing urine for 8 months. There was weakness and thinning of left lower limb for 6 to 7 months along with history of weight loss and erectile dysfunction. Clinically and radiologically, the patient was diagnosed as a case of presacral mass. USG Abdomen and Pelvis showed moderate hydroureteronephrosis due to narrowing of UV junction and no other abnormality detected. CT Scan of Abdomen and Pelvis showed mild left hydroureteronephrosis due to narrowing in distal ureter caused by heterogeneous enhancing mass in left iliac region measuring 2cm in diameter; findings suspicious of neoplastic/infective etiology. MRI of Lumbar spine showed mild disc bulge at L4-5 and L5-S1, indenting anterior thecal sac. PET CT showed FDG avid left presacral soft tissue lesion measuring 32 x 25 mm (SUV=1.45) and left mesorectal fascial thickening measuring 16x11 mm (SUV=1.93) with loss of fat plane; indentation of left posterior bladder wall; No metabolically active lymph node detected; Suspicious uptake in posterior sacral ala; Features suggestive of neoplastic mass-primary urothelial malignancy/ metastatic pelvic node.
Considering the above findings patient underwent exploratory laparotomy. Presacral mass with left VUJ and left seminal vesicle were sent for Histopathological examination. Grossly, the specimen measured 5 x 4 x 1 cm and on cut section, multiple yellowish white nodules with the largest measuring 0.4 cm were noticed. Microscopy showed fibrocollagenous tissue infiltrated by atypical epithelial cells arranged in glandular pattern. Pools of extracellular mucin were seen. Tumor cells were seen exhibiting perineural and lymphovascular invasion. Seminal vesicle, vas deferens and ureter showed normal histomorphology.
Immunohistochemistry (IHC) was carried out on the tissue sections and revealed that the atypical epithelial cells were positive for CK7, CK20, CK 19, AMACR, CDX2, CA125 and CEA and negative for PSA, WT1, PAX 8.
Tumor markers were within normal limit. Pan colonoscopy showed no evidence of bowel malignancy.

Based on the above histopathology, IHC and colonoscopy findings, a diagnosis of primary retroperitoneal mucinous adenocarcinoma was made. Patient was started on treatment with Capecitabine.

Discussion

The Primary retroperitoneal mucinous adenocarcinomas (PRMCs) are extremely rare entities. They have a female predominance with a female to male ratio of 9.4:1 and more commonly seen in young adults. PRMC is most of the time detected in the lateral retroperitoneal spaces. Pre-operative diagnosis remains elusive as there is no pathognomonic clinical, laboratory or imaging finding to facilitate the diagnosis of PRMC.

The pathogenesis of PRMC remains unclear with the four proposed main hypotheses. First hypothesis, the heterotopic supernumerary ovarian tissue plays a role. Second hypothesis states that mucinous epithelium has overgrown all other components from the monodermal variant of teratomas. Third hypothesis is about intestinal duplication. And the fourth being Coelomic metaplasia.

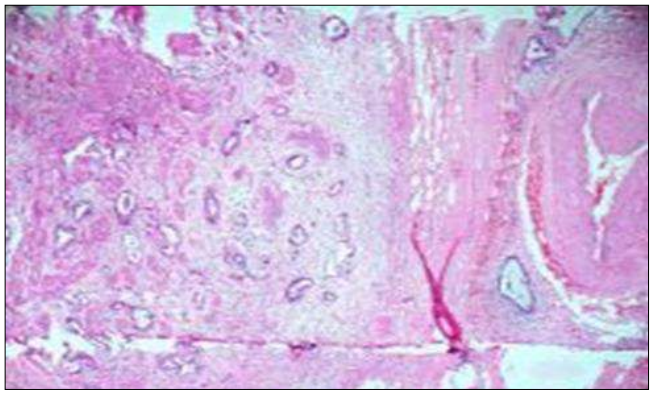

Fig 1: Atypical epithelial cells lying in glandular pattern

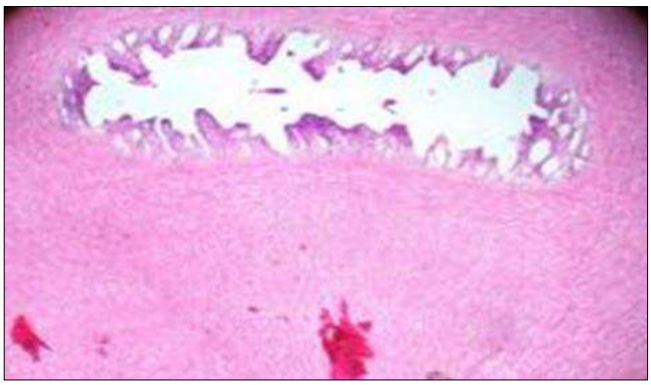

Fig 2: Non-involvement of seminal vesicle (10X)

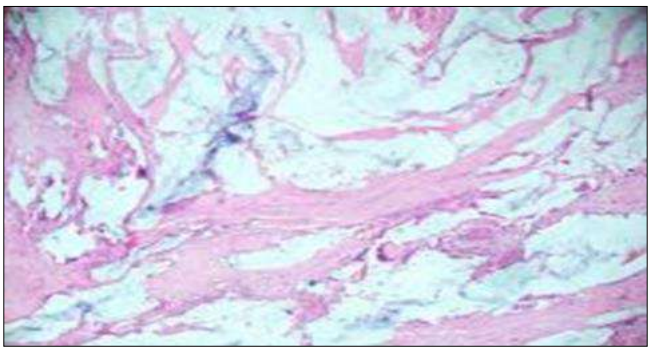

Fig 3: Extracellular pools of mucin

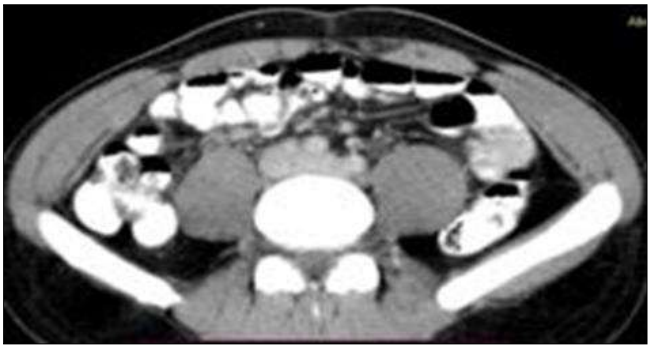

Fig 4: CT image shows presacral mass

Conclusion

PRMC constitutes one of the differentials of a retroperitoneal mass and clinicians must note when encountered with such lesions. Any delay in the diagnosis and treatment of PRMCs may lead to complications. There are no clearly defined treatment guidelines being established for the management of PRMC. Exploratory laparotomy with radical resection with no spillage or ruptures is the treatment of choice and also the most important prognostic tool. The role of chemotherapy for the treatment of PRMC is yet to be determined. With FDG PET, the lesion may exhibit low SUV so retroperitoneal carcinomas are easily confused as benign entity on imaging.

Ethical Approval

Informed consent of the patient in the study was obtained for publication of this case report.

References

1. De León DC, Pérez-Montiel D, Chanona-Vilchis J, Dueñas-González A, Villavicencio-Valencia V, Zavala-Casas G. Primary retroperitoneal mucinous cystadenocarcinoma: report of two cases. World Journal of Surgical Oncology. 2007 Dec;5(1):1-6.
2. Jiang H, Jin K, You Q, Fang W, Xu N. Retroperitoneal primary mucinous adenocarcinoma: A case report. Oncology Letters. 2011 Jul 1;2(4):633-6.
3. Joldoshova A, Belousova T, Cai Z, Billah S. Primary retroperitoneal mucinous cystic neoplasm leading to multifocal invasive adenocarcinoma of the small and large intestine. Human Pathology: Case Reports. 2020 Nov 1;22:200419.