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MALIGNANCIES OF PELVIC ORGANS IN FEMALE – MYRIAD OF MIMICS

Oncology

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KEYWORDS

INTRODUCTION

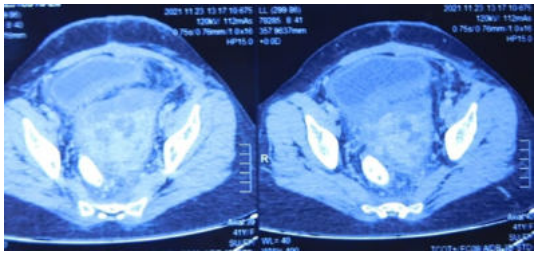
Malignancies of Pelvic Organs in females represent complex diagnostic and treatment challenges. Hidden by bony pelvis with non-specific symptoms and signs, they are often detected late in their course. Further dilemma arises while assessing the organ of origin, particularly in advanced disease. Majority arise from gastrointestinal and genitourinary tract, with less common sites including connective tissues, nerves and lymphovascular structures. Work up for these patients include imaging, bio chemical markers and biopsy. Imaging is also needed to determine whether tumor operability or neoadjuvant treatment is needed. The goal of this retrospective case analysis was to report on a series of patients who presented with a presumptive diagnosis of ovarian cancer in surgical oncology department and all were finally diagnosed to have a different malignancy. We describe the clinical presentation, surgical management, adjuvant treatment, and overall outcome in these patients.

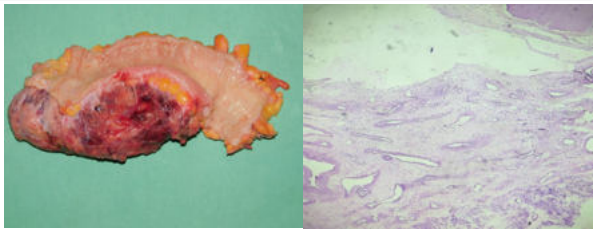
MATERIALS AND METHODS

Retrospective data was collected from hospital data base of the patients who presented with an abdominal or pelvic mass/pain to the department of surgical oncology, Government Royapettah hospital from June 2021 to December 2021. The departmental review board approval was obtained prior to study. Data were collected from the patient's hospital files on age, presenting symptoms, tumor size on physical examination, findings on ultrasonography/CT abdomen and pelvis, serum CA-125 levels, intraoperative findings, primary site of disease, pathologic diagnosis, adjuvant therapy and disease status at last follow-up. The patients' inclusion criteria in this study were those, who presented with abdominopelvic mass/pelvic pain, radiologic imaging showing a pelvic or abdominopelvic mass, and intraoperative/final histopathology revealing another malignant tumor.

CASE 1

41 year old premenopausal lady, was operated outside for a suspected uterine or ovarian neoplasm. Imaging with MRI showed a T1 hypointense and T2 mixed intense 18*13 cm lesion in the posterior cul de sac probably arising from left ovary. Intra operatively the lesion was seeing arising separately from uterus or adnexa, hysterectomy with oophorectomy was proceeded with in order to gain access to the lesion. The lesion was biopsied and any further procedure was deferred due to unavailability of technical expertise. Biopsy from the lesion revealed a spindle cell tumor probably arising from rectum. IHC revealed that to be a CD 117 negative, DOG -1 positive GIST. Exon 12 mutation of PDGFRA was also detected. Patient was put on a course of T.Imatinib 400 mg for one month. She presented to us with a vague lower abdomen mass which was felt on per rectal examination. We did wide excision with low anterior resection. There was an 8*8 cm globular mass seen arising from meso sigmoid abutting sigmoid colon, rectum and bladder. Post-operative histopathology showed a low grade GIST arising from rectum with adequate proximal, distal and circumferential clearance. Patient is now on adjuvant sunitinib therapy.



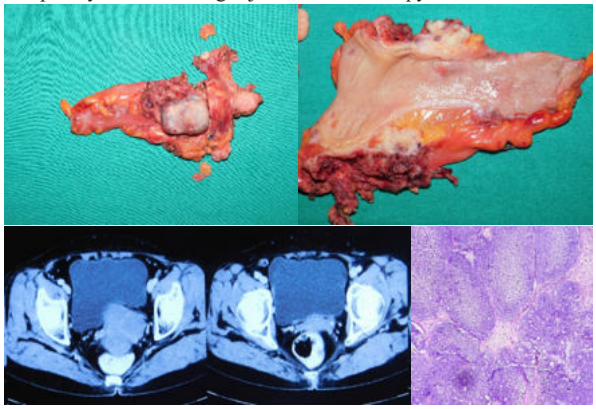


CASE 2

58 year old postmenopausal lady, evaluated for lower abdomen pain and a vague hypogastric mass. CECT showed a 12.5*10*16 cm well defined heterogeneously enhancing soft tissue dense lesion in the pelvis in the right adnexa extending to right lower quadrant with patchy haemorrhage. Multiple retroperitoneal nodes were seen with the largest in the left para aortic region measuring 1.9 cm. CA 125 was 74.9 U/ml. patient underwent a laparotomy and intraoperatively the lesion was seen arising from the mesentry of small bowel for which a wide resection was done. Post-operative HPE was suggestive of Gastrointestinal stromal tumor with clear margins all around. Mitotic rate was <5/5 sq mm and focal areas of necrosis. However IHC revealed a Desmin and SMA positive, DOG-1 negative tumor favouring leiomyosarcoma.

CASE 3

63 year old lady was evaluated for lower abdomen pain and distension. Contrast imaging showed a 9.3*7.9*7.7 cm sized soft tissue density lesion with necrotic degeneration arising from left adnexa and occupying the pelvic cul de sac. Fat plane with rectum was obscured with focal rectal wall thickening. CA 125 was 517 and CEA was 2.3. Patient underwent diagnostic laparoscopy which showed no metastatic disease elsewhere in the abdomen. There was an 8*6 cm tumor of varied consistency that seemed to be arising from the rectal wall. Low anterior resection was completed. Post-operative HPE revealed a 5.5*6 cm friable tumor arising from the bowel wall with features of extensive necrosis and sclerosis. IHC study showed it to be a poorly differentiated carcinoma probably of neuroendocrine origin because of a standalone Neuron specific enolase positivity. Patient recovered completely and is awaiting adjuvant chemotherapy



S.No	Age	Month/ year	Symptoms	Imaging	CA 125
1	41	09/21	DUB, abdomen mass	18*13 cm lesion in the posterior cul de sac which was T1 hypo and T2 mixed intense	110

2	58	11/21	Abdomen pain	12.5*10*16 cm heterogeneously enhancing soft tissue lesion	74.9
3	70	10/21	Abdomen mass	Stage 4 carcinoma sigmoid colon with ovarian metastasis	122
4	62	10/21	Abdomen mass	Carcinoma sigmoid with ovarian mass	344
5	63	12/21	Lower abdomen pain, distension	9.3*7.9*7.7 cm heterogenic lesion possibly from left adnexa	517

S. No	Intra op finding	Post op HPE	IHC	Adjuvant
1	8*8 cm mass arising extramurally from the rectum	Low grade GIST	DOG -1 (+), CD 117 (-), PDGFRA (+)	T.Sunitinib 50mg 1 OD
2	15*10 cm lesion from the uterus	leiomyosarcoma	Desmin, SMA (+)	observation
3	12*8cm left ovarian cyst – twisted	Benign fibroma with microinvasive carcinoma of colon	Not done	observation
4	Rt ovarian mass involving sigmoid, ileum and urinary bladder	Poorly differentiated carcinoma	Transitional cell carcinoma	chemotherapy
5	8*6 cm lesion from the anterior wall of rectum	Porrly differentiated carcinoma	NSE (+), synaptophysin, CD 34, S100 (-)	chemotherapy

DISCUSSION

Malignant tumors arising in the female pelvis could harbour a variety of diagnoses. While a presumptive ovarian malignancy is the most common working diagnosis, the dilemma doesn't end there. Studies and independent case reports from institutes across the world have addressed this conflict by employing various diagnostic modalities and algorithms. The following are the most common entities posing a diagnostic challenge. With adequate background knowledge and clinical suspicion, early referral to oncologist and appropriate lines of management become possible.

Primary ovarian tumors could be epithelial, germ cell or sex cord–stromal tumors. Epithelial ovarian tumors are the most common accounting for more than 90%, occurring in post-menopausal age group. Whereas, germ cell tumors and epithelial borderline tumors are frequent in premenopausal women. An adnexal mass is usually first detected with pelvic Ultrasonogram. MRI is the preferred imaging for further characterization of a sonographically indeterminate mass. Once the organ of origin is established to be the ovary , further management is based on patient age, elevated levels of tumor markers, SI of the lesion on T1-weighted images without and with fat saturation and on T2-weighted images, and lesion morphology. Diagnostic laparoscopy when employed appropriately is a useful tool.

Gastrointestinal stromal tumors (GISTs) are the most common non epithelial tumors of the gastrointestinal tract predominantly located in the stomach and small bowel. Approximately 10% of GISTs occur in the colon and rectum which may be located within the true pelvis. MRI features of GISTs are variable, with most appearing as large well-defined exophytic masses with heterogeneous enhancement due to central necrosis and haemorrhage. Small GISTs (5 cm) present as lobulated tumors with intratumoral cystic change, and mucosal ulceration. While hepatic and peritoneal metastases are common, lymph node involvement is unusual. The MRI appearance of rectal GIST often overlaps with that of other pelvic malignancies; however, invasion of adjacent organs, bowel obstruction, and regional lymphadenopathy are uncommon with GIST and a complete resection is almost always possible.

Leiomyosarcomas are soft-tissue sarcomas with tumor cells resembling differentiated smooth muscle cells. The retroperitoneum is among the most common locations, and many of these tumors arise from the inferior vena cava (IVC) or other veins, such as renal, genital, or iliac veins. These tumors show a substantial female predominance. The tumor is most often iso- to hypointense relative to skeletal muscle and heterogeneously enhancing after intravenous contrast. In the series of cases reported by Cooley et al, the most frequent sites of haematogenous metastasis were the lungs, peritoneum, and liver, followed by muscle, bones, and lymph nodes. Multidisciplinary team management is required for this rare tumor entity Poorly differentiated Neuro endocrine carcinomas (NEC) and well differentiated neuro endocrine tumors (NET) are rare tumors in female genital tract most of which are cervical small cell carcinoma and ovarian carcinoids. However data from the Surveillance, Epidemiology and End Results (SEER) suggests that the incidence of rectal NETs have been increasing over the last decades partly due to improved imaging techniques and they represent 29% of all GEP-NETs in the latest report, establishing the rectum as the most common location, slightly above the small intestine. Rectum as the primary site has a good prognosis with the stage at presentation being the most important determining factor. Studies over the years have shown the rate of lymph nodal spread and distant metastasis to be about 8% and 4% respectively. Full length colonoscopy needs to be done and a complete resection can offer cure. Systemic therapy is reserved for grade 3 tumors. Peptide receptor radiotherapy and ablative techniques are helpful in salvaging metastatic diseases

CONCLUSION

These rare non metastatic tumors arising from the female pelvic organs can pose a diagnostic dilemma even in the most experienced hands. Appropriate imaging and prompt referral to specialist centres can help in complete resection since this can be the only chance to offer a cure.

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