



PELVIC MESENCHYMAL TUMOURS – PRESURGERY VS POSTSURGERY: SURGEONS' DIAGNOSTIC DILEMMA

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KEYWORDS :

INTRODUCTION

Mesenchymal tumors of uterus are fairly common in gynaecologic practice. They are a very heterogeneous group, comprising of smooth muscle tumors and stromal tumors, which in turn shows marked variability in histological appearances, modes of treatment and clinical behaviours. Most of these tumors are benign, and are often managed by conservative surgeries like enucleation, partial excision or by hysterectomy in some cases. Accurate prediction of potential malignancy in preoperative imaging is often confounded by many factors, leading to inappropriate surgical treatment in many cases. Incomplete excision and suboptimal surgeries result in outcomes like local recurrences and metastases. Accurate postoperative characterisation of pathologic subtypes is also equally important as this has a crucial role in determining adjuvant treatment if any, follow up visits and predicting treatment outcomes. Although routine histopathological examination is sufficient, on several occasions, even it becomes a great diagnostic challenge. We hereby report a series of cases which posed a diagnostic challenge to us.

Case 1 – Broad Ligament Fibroid Vs Leiomyosarcoma

A 48 yr old perimenopausal lady presented with abdominal pain on and off since 1 year. History of abdominal hysterectomy with right salpingo-oophorectomy from elsewhere for AUB-L (PALMCOEIN Classification) in 2013. Computed Tomography(CT) - Abdomen and Pelvis with contrast (Fig.1), showed large iso-dense Space Occupying Lesion (SOL) with well-defined margins showing small calcific specks, noted in the lower abdomen superior to the urinary bladder with normal intra-lesional vessels. Previous histopathological report (HPR) was leiomyoma. Posted for laparotomy with pre operative diagnosis of large solid mass - ovarian fibroma/ broad ligament fibroid. Intraoperative - irregular nodular mass of size 18 x 13 x 8 cm, of unknown origin, left ovary normal, capsule intact, mass weighing 1kg excised. Histopathology report came as, neoplasm composed of spindle cells arranged in bundles and fascicles. A few areas show marked atypia, nuclear pleomorphism with coarse clumped chromatin, few with a prominent nucleoli. Mitoses noted 14/10hpf. Areas of coagulative and fibrinoid necrosis present – leiomyosarcoma, Federation Nationale des Centers de Lutte Contre le Cancer (FNCLCC) grade 2.



Figure 1. Case 1. – Pre-operative Diagnosis Of Broadligament Fibroid.

Case 2 – Leiomyosarcoma Vs Cellular Leiomyoma

A 54 yr old lady presented with history of heavy menstrual bleeding 3 months duration, who had previous regular cycles with normal flow. CT Abdomen and Pelvis with contrast showed left adnexal well defined lobulated mass measuring 7x 6x 4cm lesion showing broad base to the lateral uterine wall. Cystic area noted along its superior margin. Multiple prominent intra-lesional vessels noted – possibly broad ligament fibroid/ subserosal fibroid with malignant transformation to be ruled out. She underwent TAH + BSO with enbloc removal of left adnexal mass by opening broad ligament. Uterus normal size, both ovaries and tubes normal, false broad ligament fibroid. Histopathology report: ill-circumscribed cellular neoplasm composed of scanty cytoplasm, round to oval nuclei. These cells are intervened by fascicles of elongated smooth muscle cells. Many small arteriole like vessels, thick walled vessels and dilated lymphatic spaces are noted within the tumour. Occasional mitotic figures noted [$<5/10$ hpf] - cellular leiomyoma / Endodermal Stromal tumours. IHC done. Desmin, SMA, CD 10 positive, Ki67 $< 1\%$ cells are positive - favour of cellular leiomyoma than low grade endometrial stromal tumors (LGESS). However, immune- histochemistry (IHC) marker, h-caldesmon is required for confirmation which was not available for us.

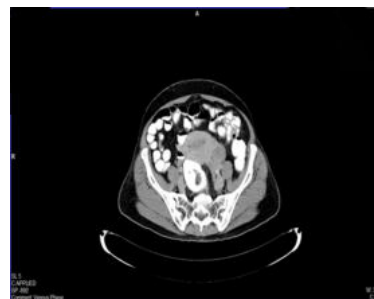


Figure 2. Case 2. Pre Operative Diagnosis Of Leiomyosarcoma.

Case 3 – Sarcoma Vs Vaginal Cellular Leiomyoma

A 44 year old perimenopausal lady presented with heavy menstrual bleeding since 1 year and vulval mass near the introitus since 2 months. She was diagnosed with AUB -L and enlarged B/L Bartholins cyst, severe anaemia corrected by PRBC transfusion. Underwent dilatation and curettage with marsupialisation in June 2018. Histopathology came as leiomyoma [marsupialised specimen]. MRI done – bilateral perineal mass of size 9x9x7cm on the right side and 3x2cm on the left side, with T2 hypointense signals with moderate post contrast enhancement, involving distal vagina, labia and urethral wall – mesenchymal/fibrous tumour/ sarcomatous lesion. She underwent TAH + BSO and bilateral perineal swelling excision in October 2018. HPR: uterine – leiomyomatosis/ ?intravenous leiomyomatosis. Paravaginal tumour: cellular leiomyoma with degeneration. Due to high chance of recurrence, patient was on follow up. She again reported with labial swelling. MRI done – 6x6x5cm irregular lobular lesion – right lateral aspect of distal third of vagina -? recurrence / residual disease. But clinically highly suspicious of sarcoma. Same excised through perineal approach without any residual disease macroscopically. HPR came as cellular

leiomyoma.

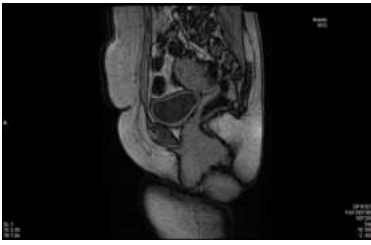


Figure 3. Case 1. – Pre-operative Diagnosis Of Sarcoma.

DISCUSSION

Uterine leiomyomas and leiomyosarcomas[ULMS] are at opposite ends of the pathologic spectrum of uterine smooth muscle tumors. In between, there are several leiomyoma variants, such as mitotically active, cellular [CMs], and atypical leiomyomas, as well as smooth muscle tumors of uncertain malignant potential (STUMP). Leiomyomas are the most common mesenchymal tumours of uterus, comprising about 70% of all uterine masses. They are usually treated by enucleation, or simple hysterectomy. Sarcomas are relatively rare among uterine tumors, but with high recurrence rate. Of all uterine tumors about 3-7% are leiomyosarcomas,[1] and endodermal stromal tumors[EST] constitute 0.2 to 1.5%.

Histopathological examination is sufficient to distinguish between endometrial stromal sarcoma [ESS] and smooth muscle tumors in most of the cases, but the distinction between ESTs and highly CMs, on some occasions poses a great diagnostic challenge to the pathologist. The differentiation between EST and CM is necessary on account of several reasons. Firstly, highly CM follows a benign course with recurrence usually, while ESTs, especially ESS results in metastases. Secondly, the therapy for the two varies. ESSs are usually ER positive, so respond to anti-estrogen therapy. This would be useful, especially in metastatic ESS or extra-uterine ESS; while for CM, myomectomy will suffice. Moreover, IHC also does not provide a solution to this problem because a completely endometrial stroma-specific IHC profile is not available. This could be due to the fact that both endometrium and myometrium arise from Mullerian duct, hence often express similar antigens with a considerable overlap between some markers. Additionally, endometrial stromal cells have the capacity to differentiate into well-developed smooth muscle cells. So, smooth muscle actin (SMA), muscle-specific actin (MSA), and desmin may be expressed by both the tumors. Role of desmin as a discriminatory marker is also controversial, however the consensus is that if desmin is diffusely and strongly positive, it favours a CM over an EST.[2] The management of early stage high grade uterine sarcoma[HGUS] usually includes total abdominal hysterectomy and bilateral salpingo-oophorectomy. The value of lymphadenectomy and debulking of gross extra-uterine disease remain unclear. HGUS represents an aggressive subtype with poor outcomes regardless of the stage at presentation. The patterns of relapse into the abdominal cavity and distant metastases highlight the limited role of adjuvant RT to the pelvis and the greater unmet need evaluating the role of systemic therapy. Total abdominal hysterectomy and bilateral salpingo-oophorectomy are the standards of care for LMS. Adjuvant pelvic RT does not have a survival benefit in LMS but may reduce local pelvic recurrences, possibly for disease that has spread beyond the uterus (FIGO stages II–IV). The risk of disease recurrence following Stage I ULMS has been reported as 50%–70% at 2 years.

Though at-times preoperative imaging are inconclusive, mostly atypical features at preoperative imaging may suggest potential malignancy, and one should be wary of

enucleation/morcellation. Principles of oncologic resection should be employed in all cases of suspected malignancy. But can we ?. Because STUMP, when found in young females - fertility is a greater concern !. Also in cellular leiomyoma due to its aggressive growth and recurrence, pre operative imaging points towards sarcoma, even-though the patient is young. Now the decision is in surgeons' hand !!.

CONCLUSION

Much remains to be learned clinically, particularly regarding those histologic entities that lie between, called leiomyoma variants, LMS and EST. Nonetheless, the bottom line is that, at the present time, the diagnosis of all mesenchymal tumors of uterus cannot be made with certainty especially leiomyoma variants until a pathologist, guided by recently updated World Health Organization criteria (3), thoroughly examines the specimen. Histopathology together with IHC, though it proves to be of great value in arriving at definitive diagnosis, had grey areas. In our peripheral settings, a tumour board with surgeon, radiologist and pathologist will result in proper diagnosis, selection and plan of management, and also prompt referral to oncology centres in case of diagnostic dilemmas.

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