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Case Report: A Rare Case of Uterine Spindle Cell Neoplasm

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ABSTRACT

Uterine masses presenting as huge abdominal swelling is a very rare presentation in this current era where advanced imaging modalities and medical care is easily accessible. We discuss a case of a patient who arrived with a massive abdominal enlargement. Myxoid spindle cell neoplasm of the uterus was discovered to be the tumor. Smooth muscle tumors (SMTs) are the most common uterine mesenchymal tumors. On the basis of their gross and microscopic appearances, the majority of uterine SMTs can be classified as benign or malignant. This study outlines a practical method to uterine neoplasm diagnosis based on a systematic evaluation of histologic characteristics, as well as a systematic approach to differential diagnosis based on histologic and immunohistochemical findings.

KEY WORDS: BENIGN UTERINE NEOPLASM, LEIOMYOSARCOMA, MYXOID SPINDLE CELL NEOPLASM.

INTRODUCTION

Huge abdominal tumours have become uncommon in this current era due to the availability of advanced radiological modalities which help to make early diagnosis. Among the abdominal tumours, pelvic tumours are not uncommon in incidence. Tumour characteristics and its nature can be easily identified on imaging modalities such as Computed tomography (CT) or magnetic resonance (MR) imaging. The commonest causes of pelvic lumps are pregnancy, leiomyoma, and functional ovarian cysts. Among all the pelvic masses, those occurring in the reproductive age group are considered benign. Despite advances in oncology, the most common cause of mortality is late diagnosis thus creating a psycho-physio-biological impact on patient who are at an advanced stage of malignancy. Herein, we present an uncommon case of myxoid spindle cell neoplasm in woman with initial suspicion of ovarian malignancy.

Case Report: A 32-year-old Indian housewife, P2L2, BMI 32.86 came to oncosurgery outpatient department, with complaints of gradually increasing abdominal distention for 6 months, accompanied with dull aching pain in abdomen for 4 months which was associated with increased frequency of micturition for 2 months. Her abdomen was distended, and with an abdominal girth of 89 cm at level of umbilicus, a well-defined mass occupying whole abdomen extending till epigastrium with a dull note on percussion. Her Urine pregnancy test was negative. Multiple detectors computed tomography scan of abdomen and pelvis (plain + contrast) (Figure

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1) was suggestive of large solid cystic lesion measuring approximately 14.6 cm (AP)× 22.9 cm (trans) × 25.1 cm (CC) seen arising from the pelvis and occupying most of peritoneal cavity. Solid part of lesion was heterogeneously enhancing.

Figure 1: Shows evidence of large uterine mass on computerised tomography scan

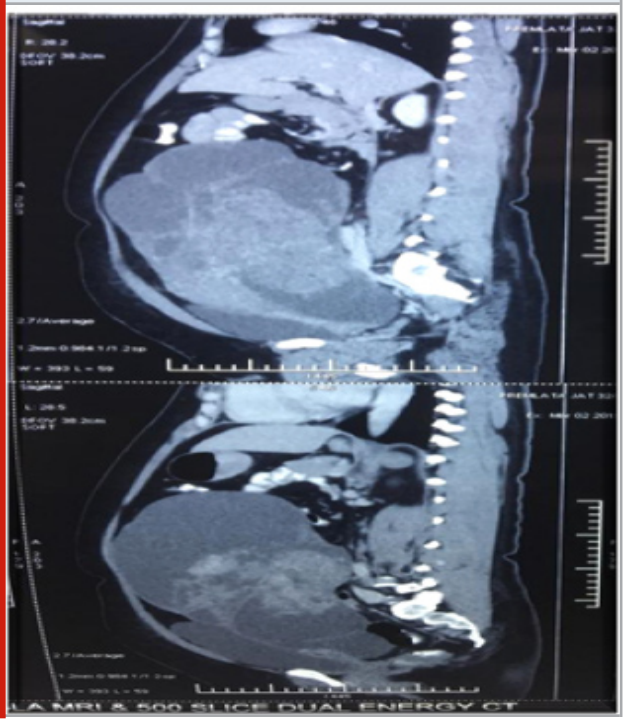
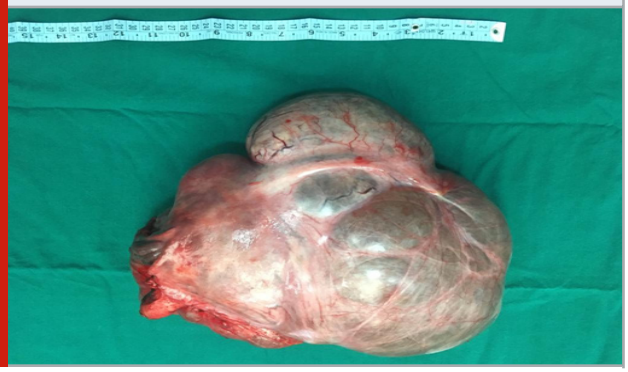


Figure 2: Shows large uterine mass after its removal



Exploratory laparotomy was planned. A huge solid cystic tumour arising from uterus, seen occupying abdomen cavity. Pan hysterectomy with bilateral salphingo-oopherectomy with resection of mass was done. Tumour measuring 35 cm × 30 cm and weighing 5 kg was removed En bloc (see Figure 2). The mass was subjected to frozen section. Frozen section report was suggestive of Myxoid spindle cell neoplasm, possibly of smooth muscle differentiation. Lymph nodes were not involved. An abdominal drain was placed in situ for 5 days. Post-operative period was uneventful. Histopathological examination revealed myxoid epithelioid/spindle cell neoplasm of uterus.

DISCUSSION

Smooth muscle tumours (SMTs) are among the commonest mesenchymal tumours of the uterus (Gemma Toledo et al., 2008). Leiomyosarcoma of uterus is a very uncommon finding in uterus. It accounts for only 8-10% of all the uterine malignancies. Among all the sarcomas, leiomyosarcoma is the commonest seen histological subtype (Santos P et al., 2015). These tumours are most frequently seen in the 4th decade of life. One of the rare morphological variants of uterine leiomyosarcoma is uterine spindle cell leiomyosarcoma. Spindle cell sarcoma is considered as an uncommon variant or subtype of RMS9 which mostly occurs in the para testicular region in children. Thus, it is occurring in uterus is a very rare possibility (Kathpalia SS et al., 2018). We present a rare case of 'Spindle Cell Sarcoma' of uterus which had grown to an exponential huge size. Spindle cell sarcoma of uterus was not considered among the differential diagnosis in our case. The diagnosis was made post operatively after the tumour was subjected to histopathological analysis. The most unusual feature in our case was that it was not invading the surrounding local structures which was in favour of good prognosis.

The Gynaecologic Oncology group have classified uterine sarcomas into non-epithelial and mixed epithelial sarcomas. Furthermore, non-epithelial sarcomas are further divided depending on the type of cancerous cells and its presumed tissue of origin (Kathpalia SS et al., 2018). The non–smooth muscle tumours that originate in uterus have some of the histological and immunohistochemical features like that of uterine smooth muscle tumours (spindle and epithelioid type) which leads to difficulty in classifying these tumours (Bardarov S et al., 2011). Norris and Taylor in 1966 first described the characteristics of these rare tumours. They classified these tumours from benign stromal nodules to malignant sarcomas depending on the degree of mitotic activity. World Health Organisation has defined malignant stromal tumours as high-grade endometrial sarcomas which are poorly differentiated and are completely different in appearance as compared to normal endometrial stroma (Norris HJ et al., 1996).

Leiomyosarcoma, undifferentiated sarcoma, and mixed epithelial and mesenchymal tumors should all be distinguished from uterine RMS. The diagnosis of spindle cell myxoid neoplasm can be aided by extensive tumor sampling, meticulous microscopic examination, and immunohistochemistry labeling (Rubin et al., 1996). published his two case reports of adult spindle cell RMS. Thus, it can be concluded that pure RMS of uterus is a very rare finding and in that spindle cell variant is the rarest (Rubin et al., 1996). Nascimento and Fletcher performed the largest review comparing the most common sites involved by the adult spindle cell RMS. They reported that the head and neck region was the most frequent site of occurrence followed by the retroperitoneum and lower extremity (Nascimento AF et al., 2005).

In our case, on presentation to outpatient department, diagnosis of pelvic tumour was confirmed on imaging study CT scan. Then proceed with exploratory laparotomy and frozen section to avoid tumour seedling & upgrading stage in image guided biopsy. Final histopathological report had been spindle cell myxoid tumour of the uterus. Histopathological diagnosis is important for differentiation of tumour to benign or malignant, low, or high grade. Even HPE diagnosis is difficult because of the numerous variations, minor tumor characteristics, and overlapping morphologic aspects that make distinguishing these entities difficult. 3 Young-Mee-Lim suggests that myxoid ESS of the low-grade variety may follow a different clinical path than typical low-grade ESS. Low-grade ESS is well-known for growing slowly (Lim Y et al., 2017).

Because both have substantial myometrial infiltration and intravascular development, myxoid LMS must be distinguished from endometrial stromal sarcoma with myxoid change (Amant F et al., 2006). It's vital to remember that myxoid LMSs may have lower levels of smooth muscle marker positive than ordinary LMSs, and that U-SMT in general, and LMS in particular, frequently express CD10 (Amant F 2006 & Clement PB 2000). A mitotic index of <2MFs/ 10 HPFs in the absence of tumour cell necrosis or severe cytologic atypia, favoured diagnosis of myxoid leiomyoma (Prat J). In our case, mitotic activity was 0-1/10hpf. Masayuki Shintaku Akiko Fukushima reports a surgical instance of an inflammatory myofibroblastic tumor originating in the uterine corpus with significant myxoid stroma.

Inflammatory myofibroblastic tumors of the uterus can occasionally have a significant myxoid stroma, making distinction from myxoid leiomyosarcoma difficult (Shintaku M et al., 2006). A number of studies on different types of uterine tumours and associated structures were reported (Sharma 2021 & Mounika 2020). Also few key studies related to management of carcinoma are available (Khatib 2018 & Anuraj H 2018). In the management of uterine myxoid spindle cell neoplasm, clinician, radiologist, oncosurgeon and histopathologist all plays vital role for curative treatment. A multimodality approach is required.

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