Case Report

UTERINE SMOOTH MUSCLE TUMOUR OF UNCERTAIN MALIGNANT POTENTIAL PRESENTING WITH AN UNUSUAL GROSS APPEARANCE A CASE REPORT

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Abstract

Smooth Muscle Tumors of Undetermined Malignant Potential or STUMPs are rare and interesting tumors from both the standpoint of histological diagnosis and classification, as well as clinical management mainly because, as a group, its natural history is poorly understood.We report a case of STUMP presenting as a cystic uterine mass in a 48 year old peri-menopausal female . Grossly received uterus revealed a large thick wall cystic cavity filled with mucinous fluid . Microscopy revealed features suggestive of a low grade leiomyosarcoma ,however immunohistochemistry confirmed the diagnosis of STUMP. The patient was kept under follow up. She showed no evidence of local recurrence and continues to be under close surveillance 18 months post diagnosis. To our knowledge,uterine smooth muscle tumors with uncertain malignant potential with such an unusual gross appearance has not been reported in the literature.

Key Words: Smooth muscle tumor with uncertain malignant potential (STUMP),unusual gross appearance.

Introduction

Smooth muscle tumors of uncertain malignant potential were first described by Kempson et al in the year 1973(1). STUMPs represent a histologically heterogeneous group of uterine smooth muscle tumors that do not clearly fall into the category of either leiomyomas or leiomyosarcomas(2).

Smooth muscle tumors are histologically categorized into leiomyomas and leiomyosarcomas, based on the combination of histological parameters such as mitotic activity, cytological atypia and coagulativetumor cell necrosis(3). These are interesting tumors from both the standpoint of histological diagnosis and classification as well as clinical management mainly because, as a group, its natural history is poorly understood.

Herein we report a case of uterine STUMP with an unusual gross appearance.

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Case Presentation

A 48 year old perimenopausal female presented to the gynaecological out patient department with increased frequency of micturation, incomplete evacuation of bladder and pain in lower abdomen of short duration. She had history of amenorrhoea followed by continous bleeding for 15 days. On per vaginal examination a hard, irregular mass of 24 weeks was felt on the right side of pelvis with restricted mobility.

A CT Scan with an oral and intravenous contrast was initially performed that revealed a large, homogenous, well defined , encapsulated lesion of size 17 ×15×12 cm probably arising from the anterior aspect of uterus. There was no calcification or necrosis appreciated in the lesion, neither was there any evidence of lymphadenopathy, free fluid in the abdomen or any mural nodule noted. The ovaries were not identified on the CT Scan and hence a Pelvic USG was performed. This re-demonstrated the mass lesion arising from the uterus containing a fluid filled thick walled cavity.Serum CEA -125 was within normal range.Considering the above findings a provisional clinical diagnosis of uterine fibroid

with cystic degeneration was kept and the patient was subjected to Subtotal abdominal Hysterectomy and bilateral salpingo-oopherectomy.

Pathological Findings:

Grossly the specimen of uterus revealed a large,thick walled cavity of size 8.5 × 2 cm arising from the lower uterine segment, the inner surface of which was nodular with haemorrhagic areas.

Microscopy showed a fibro-muscular wall covered by sheets and clusters of spindle cells showing diffuse moderate pleomorphism with large dense irregular nuclei with 0-1 prominent nucleoli. 2-3 mitosis/10 HPF was present and scanty necrosis was also seen.Immunohistochemistry staining showed positivity forvimentin,desmin,smooth muscle actin. Negative staining for Pan CK and EMA was noted. Ki-67 index of<0.5% was present.

Considering the above features a diagnosis of STUMP was made.

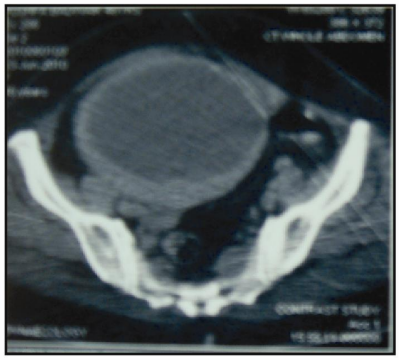

Fig 1 CT Scan showing a homogenous, hypodense lesion arising from the uterus.

Fig-2 Large thick wall cystic cavity arising from the lower uterine wall.

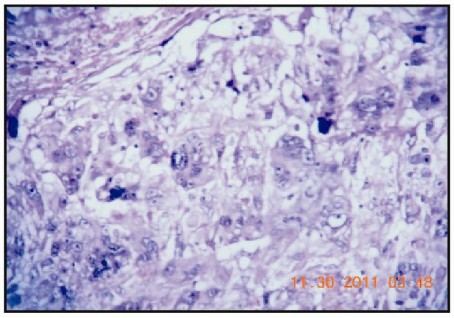

Fig 3 -Microscopy showing sheets & clusters of moderately pleomorphic / multinucleated cells with large bizzare nuclei. (100x)

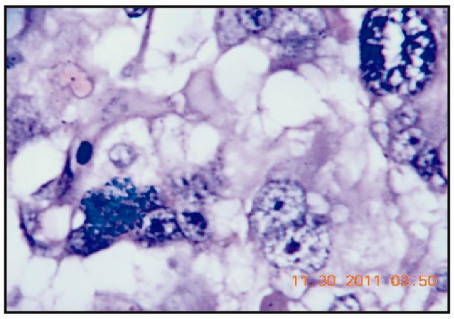

Fig 4 -High power view showingbizzare nuclei coarse chromatin andabnormal mitosis.(400 x)

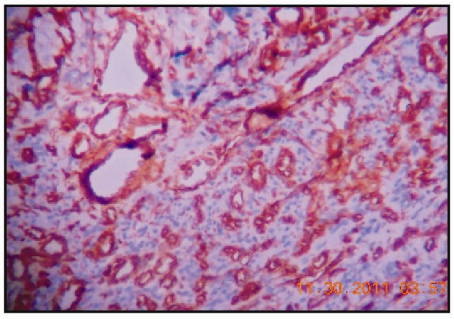

Fig -5 Vimentin positivity (100 x)

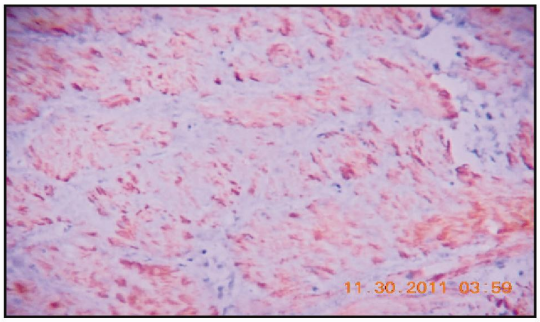

Fig 6- Desmin positivity (100x)

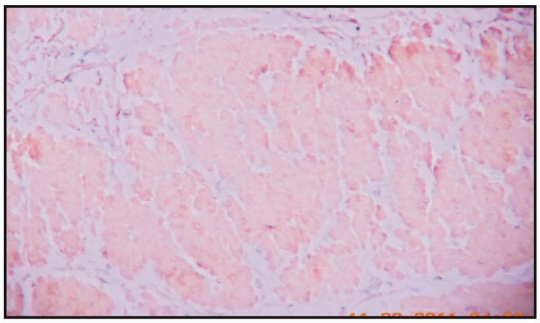

Fig 7- Smooth Muscle Actin positivity (100x)

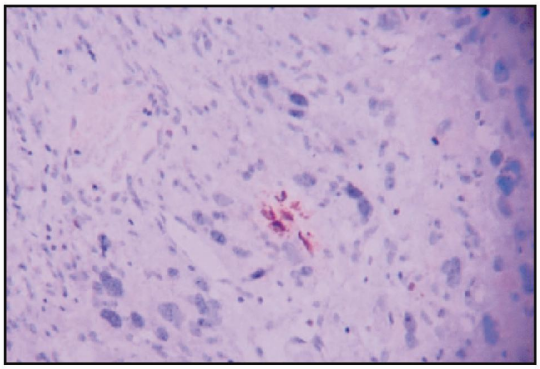

Fig-8 Ki -67 Proliferative index of < 0.5 %. (100 x)

Discussion

Uterine smooth muscle tumors of uncertain malignant potential are rare neoplasms that cannot be reliably diagnosed as benign or malignant and are hence designated as STUMP. Uterine leiomyosarcoma represent <2 % of all uterine malignancy but is the most common uterine sarcoma, constituting 25 %of mesenchymal tumours of the uterus and STUMPS are more uncommon(4,5)

According to Joseph et al 83% of patients are premenopausal females that present with non-specific abdominal symptoms with or without a history of abnormal menstrual cycle(6). Statistically, a patient presenting with an enlarged globular uterine tumor is likely to have a final diagnosis of benign uterine leiomyoma(7). According to Bell et al, three histopathological criteria were examined: coagulative tumour cell-necrosis (CTCN), degree and extent of atypia and mitotic index (MI).Based on these parameters, the definition proposed by Bell et al. is as follows(8):

A.Leiomyosarcomasare defined as tumors with at least two of the following three features: diffuse cytological atypia, tumor cell necrosis, and ≥ 10 mitosis events per 10HPFs.

B.Leiomyomas are defined as tumors with no or mild cytological atypia, no tumor cell necrosis, and < 5 mitosis events per 10HPFs.

C. STUMPs are defined as tumors with following features: (1) focal moderate to severe cytological atypia, no tumor cell necrosis, and < 5 mitosis events per 10HPFs, or (2) no or mild cytological atypia, tumor cell necrosis, < 10 mitosis events per 10HPFs.

Other studies by O'Connor et al and Marisa et al. have given similar criteria with mild variations.

However STUMPS need to be differentiated from symplastic leiomyomas that are smooth muscle tumors with multiple, gigantic nuclei with abundant nuclear chromatin in an otherwise typical leiomyoma with foci of bizarre and pleomorphic tumor cells with atypical multilobed nuclei. Most of the nuclear features appear to be degenerative, with smudged chromatin, vacuolization, and pyknosis. Multinucleated cells may be found focally, multifocally, or diffusely throughout the neoplasm, and occupy more than 25% of the tumor in most cases. These tumors often show degeneration, edema, and hyaline change, but not coagulative tumor cell necrosis, with the symplastic (bizzare) cells predominantly at the edge of the degenerating areas. Mitotic figures are often lacking and if present they are never atypical(9).

Out of the three criteria proposed by Bell et al, coagulative tumor cell necrosis and extensive severe atypia seemed to correlate with malignant behavior(1). However, Burns et al, reported the importance of coagulative tumor cell necrosis as

the single most powerful factor for malignant behavior among morphological features(10). The role of Ki-67as a proliferative marker and the importance of p53 over-expression has been stressed upon in the recent years. Over-expression of p53 and a high Ki-67 labelling index are frequently associated with leiomyosarcoma, and therefore these markers may be useful IHC parameters to distinguish between cases of malignant smooth muscle tumors and those of uncertain or borderline histology(11). During the 2 year(1),8 years(3),15 years(8) follow up of stumps in various studies ,only 1-2 cases of stumps recurred as malignant tumors or showed metastatic deposits. As mentioned above, previous studies suggest that STUMPs are usually clinically benign, but they should be considered as tumors of low malignant potential because they can occasionally recur or metastasize to distant sites, years after hysterectomy(2). Therefore, patients diagnosed with STUMPs should receive long-term follow-up. Our patient continues to be under close surveillance 18 months post diagnosis.

Conclusion

Smooth muscle tumours of uncertain malignant potential have an unpredictable clinical course, and can grossly have a varied presentation. Relapses generally appear to occur after a long disease-free interval of up to several years and therefore, patients diagnosed with smooth muscle tumours of uncertain malignant potential should receive long-term follow-up.

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