Uterine Symplastic Leiomyoma: A Rare Smooth Muscle Tumor

Saeed Alam¹ and Huma Mushtaq²

¹Professor of Pathology, Islamabad Medical & Dental College, Islamabad
²Assistant Professor Pathology, Islamabad Medical & Dental College, Islamabad
(Bahria University, Islamabad)

Abstract
Leiomyomas are the commonest smooth muscle tumors of uterus and are further classified according to their pathological features. Majority of the leiomyomas usually do not have a diagnostic problem but some of its subtypes mimic malignancy and so are of great interest. Symplastic leiomyoma is a rare variant of leiomyoma and such leiomyomas require a precise histopathological evaluation so that they are not misinterpreted as leiomyosarcomas due to their similarity of microscopic features. Here we present a case report of Symplastic leiomyoma in a young lady with primary infertility. The patients presenting with this type of leiomyoma should be followed up carefully and total hysterectomy should be considered if fertility preservation is not a consideration.

Key words: Symplastic leiomyoma, uterus

Introduction
Leiomyomas are the commonest smooth muscle tumors of uterus and are further classified according to their pathological features.¹ Most of the diagnostic terminologies reflect the morphological details of a lesion, but they do not provide a proper information regarding the clinical details or prognosis of that disease which can guide in postoperative management.² Majority of the leiomyomas usually do not have a diagnostic problem but some of its subtypes mimic malignancy and so are of great interest.³ Symplastic leiomyoma, also referred to as Bizarre, Pleomorphic, or Atypical leiomyoma, is a rare variant of leiomyomas. Such leiomyomas require a precise histopathological evaluation so that they are not misinterpreted as leiomyosarcomas due to their similarity of microscopic features.³

Symplastic leiomyomas microscopically reveal cells with moderate to severe cytological atypia but there is absence of cell necrosis and mitotic index is less than 10/10 hpf. Even though it has high cellularity along with numerous widely distributed bizarre cells and low mitotic count it is categorized as a benign tumor.⁵ Symplastic leiomyoma has been diagnosed in sites other than the uterus such as in vagina, nasal cavity and scrotum.⁶ This tumor leads to challenging treatment issues involving fertility preservation especially when diagnosed in younger females. The incidence of symplastic leiomyoma is very low, therefore there is not enough evidence to support myomectomy alone as an appropriate management option and its impact on fertility is not well known yet.⁷ Certain immunohistochemical profiles and genetic aberrations suggest that uterine leiomyosarcoma can arise from pre-existing areas of Leiomyoma that have a symplastic or cellular morphology. Generally it is rare for leiomyomas to progress to Leiomyosarcomas and the frequency of leiomyosarcomas is only 0.1–0.3% as compared to leiomyomas.⁸ It is becoming increasingly important to differentiate between benign and malignant uterine smooth muscle tumors in order to plan a proper treatment strategy. It is important to have conservative set of measures for women who have benign uterine pathology and want to preserve fertility.⁷ Here we present a case report of symplastic leiomyoma in a young lady with primary infertility.

Case report
A 32 year old female presented with complaints of heavy menstrual bleeding and mild anaemia. Ultrasound examination showed a uterine leiomyoma with a diameter of 9 x 6 cm. Myomectomy was carried out. Grossly the tumor was circumscribed and cut surface showed an ill-defined whorled pattern with no recognizable areas of necrosis.(figure1) Histopathological evaluation of the specimen revealed atypical cells having pleomorphic nuclei and infrequent mitoses (figure 2)

Figure 1: Gross section of Symplastic Leiomyoma
Discussion

Symplastic leiomyomas are a rare type of uterine tumors and are about 0.5% of uterine mesenchymal neoplasms. They belong to a grey zone from a clinical and morphological point of view. The characteristic features of this variant include pleomorphic tumor cells with distinct atypical nuclei and low mitotic rate. Some of the pleomorphic cells are multinucleated or multilobulated and these changes can be focal, multifocal or diffuse throughout the mass. Other features seen in these tumors are degeneration, edema and hyaline change but there is no coagulative tumor cell necrosis. Depending upon the features of nuclear pleomorphism, mitotic count and areas of coagulative necrosis the tumors are categorized from benign to potentially malignant or malignant leiomyosarcomas. Such symplastic leiomyomas can be misdiagnosed as leiomyosarcoma but distinction between atypical leiomyoma and leiomyosarcoma is made on mitotic activity usually equal to or more than 10 /10 hpf in the later. In this case report the patients age was 32 years which is comparable to the results seen in a study by Downes et al in which patients age ranged from 25 to 51 years (mean 40.7). In another series of 5 cases of symplastic leiomyomas reported by RE Fechner the patients age group was in a range of 32 to 47 years. In different studies patients presented with different clinical features. In this case report patient presented with mass abdomen and menorrhagia for a period of 2 years. Similarly in another case report by Downes et al, the patient presented with enlarged uterus, heavy menstrual bleeding and pelvic pain. In a series of three cases reported by RE Fechner the presenting complaints were dysmenorrhea, menstrual irregularity (three cases) and pelvic pain. The size of the tumor in our case was 9 X 6 cm which is comparable to other case report by Downes et al in which the range was 1 to 14 cm and mean was of 4.2cm. The features of Symplastic leiomyomas raise the question of postoperative recurrence and the possibility of distant metastasis. Most of the time these tumors are managed conservatively with minimally invasive surgical techniques. In our case the patient wanted to preserve fertility and myomectomy was the optimal choice. As substantial data regarding management and long term follow up is not available some studies have concluded that the behavior of those symplastic leiomyomas who have undergone myomectomy is still not clear. According to some sources there is no recurrence after conservative techniques like hysteroscopic myomectomy or morcellation techniques but other studies have concluded that the behavior of those symplastic leiomyomas that have under gone myomectomy is still not clear due to the limited data available regarding the follow up of such patients. On the other hand some authors have presented cases with metastatic leiomyoma, and some even after total hysterectomy had malignant transformation and also local recurrence at the site of myomectomy. In order to differentiate between benign and malignant smooth muscle tumors and also to confirm smooth muscle tumors of uncertain potential, mainly histological criteria is used. Further confirmation is done through different immunohistochemical markers like p16, p53, Ki 67, Calponin h1 and Desmin expression. In case of Leiomyosarcoma there is over expression of p16, p53 and Ki 67 antigen along with reduced expression of Calponin h1 and Desmin therefore differentiating this malignant form from tumors of uncertain malignant potential and benign variants of leiomyoma.

Conclusion

Symplastic leiomyoma is a rare benign uterine smooth muscle tumor. It closely mimics its malignant counterpart therefore proper evaluation of its microscopic features should be done. Patients presenting with this type of leiomyoma should have a thorough follow-up and total hysterectomy should be considered if fertility preservation is not a consideration.

References

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