Case Report

Indian Journal of Pathology: Research and Practice
Volume 12 Number 3, July - September 2023
DOI: http://dx.doi.org/10.21088/ijprp.2278.148X.12323.3

Leiomyoma with Bizarre Nuclei: An Uncommon Variant of Uterine Smooth Muscle Neoplasm

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How to cite this article:
Sunil V. Jagtap, Swati S Jagtap, Pranjal Shah, *et al.*/ Leiomyoma with Bizarre Nuclei: An Uncommon Variant of Uterine Smooth Muscle Neoplasm/Indian Journal of Pathology: Research and Practice 2023;12(3) 105–108.

Abstract

Leiomyoma with bizarre nuclei (LBN) is an uncommon variant of uterine smooth muscle neoplasm. A 52 year female presented with complained of intermittent pelvic pain, abnormal uterine bleeding, and abdominal discomfort of 6 month duration. On ultrasonic evaluation showed well defined uterine masses suggestive of leiomyomas. The systemic examinations were normal. On histopathologycal finding reported as uterine leiomyoma with bizarre nuclei. Patient responded well to treatment. On follow up there was no evidence of recurrence. We are presenting this uncommon case of leiomyoma with bizarre nuclei for its clinical, radiological and histopathological findings. On histopathological findings this tumor should be properly differentiated form various uterine spindle cell tumors with nuclear atypia for better management of patients.

Keywords: Leiomyoma with bizarre nuclei; Smooth muscle tumors; Uterine tumors; Histopathology.

INTRODUCTION

In 1909, Kelly and Cullen, described Leiomyoma with bizarre nuclei (LBN) as a benign uterine leiomyoma with "sarcomatous degeneration.[1] The WHO adopted the terminology "leiomyoma with bizarre nuclei" and classified it as a leiomyoma variant and the other terminologies were no longer recommended.[2]

In uterine myometrium, spindle cell tumors with nuclear atypia are noted in various conditions like leiomyoma with bizarre nuclei, fumarate hydratase deficient leiomyoma, intravenous leiomyomatosis, perivascular epithelioid tumor, inflammatory myofibroblastic tumors, uterine smooth muscle tumors of uncertain malignant potential (STUMP) and leiomyosarcoma. On histopathological findings these tumors should be properly differentiated for better management of patients.

The 2020 WHO classification defined LBN as a leiomyoma subtype with bizarre cells arranged in

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Received on: 23.03.2023 **Accepted on:** 15.06.2023

a multifocal to diffuse distribution in a background of typical leiomyoma.[3]

Oliva E. described the specific criteria to diagnosis LBN is mainly based on clinical information, gross appearance, and strict utilization of morphologic criteria, including cytologic atypia, mitotic activity, and tumor cell necrosis.[4]

Herewith we are presenting this uncommon case of leiomyoma with bizarre nuclei.

CASE REPORT

A 52 year female presented with complains of intermittent pelvic pain, abnormal uterine bleeding, and abdominal discomfort of 6 month duration. On ultrasonic evaluation showed well defined uterine masses suggestive of leiomyomas. The systemic examinations were normal. There was no any history of hormonal therapy. The past and family history was non contributory. Patient underwent panhysterectomy. On histopathologycal finding reported as uterine leiomyoma with bizarre nuclei.

Gross examination, specimen of panhysterectomy showed uterus with cervix measured 10×8×5 cm and weighs 200 gms. External surface of uterus at fundus and body at posterior lateral wall showed bossilation. External surface of cervix appeared unremarkable. On cut section, endometrial canal measured 6.5 cm. Average endometrial thickness was 0.2 cm. The wall of uterus showed one intramural and two submucosal fibroids. The largest submucosal fibroid measured 2.7 in diameter. Cut surface showed grey white, firm, whorled, circumbscibed tumor with areas of hemorrhages (Fig. 1).

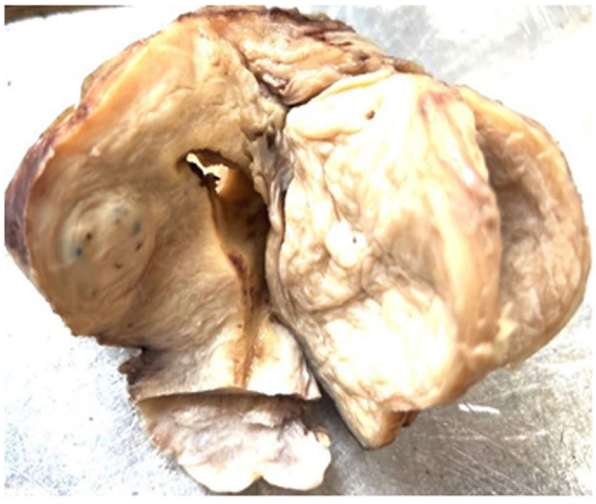

Fig. 1: Specimen of pan hysterectomy shows grey white, firm, whorled, circumbscibed tumors with areas of hemorrhages.

Average myometrial thickness was 4 cm. The cervical canal measured 3 cm in length. The right ovary measured 4×3×1 cm and cut section showed a cyst measured 2.6x2x1.5 cm and filled clear fluid. The left sided ovary measured 2.3x1.6x0.5 cm. The both fallopian tube were normal. A left peritubal cyst was noted.

On histopathologycal finding reported as chronic nonspecific cervicitis, nabothian cyst with inflamed granulation tissue, foci at hemorrhage and congestion. Mild endometrial hyperplasia with mild chronic nonspecific endometritis was noted. Three leiomyomas, one intramural, and two submucosal locations were noted. The intramural largest tumor showed a tumor of benign uterine smooth muscle having expansive margins. Tumor was arranged in a diffuse, sheath, whirling patterns. Tumor showed multifocal, or diffused bizarre cells many scattered bizarrely shaped, hyperchromatic, multilobulated nuclei with nuclear pseudoinclusions and occasional small nucleoli and eosinophilic cytoplasm, on a background of a typical leiomyoma (Fig. 2, 3, 4).

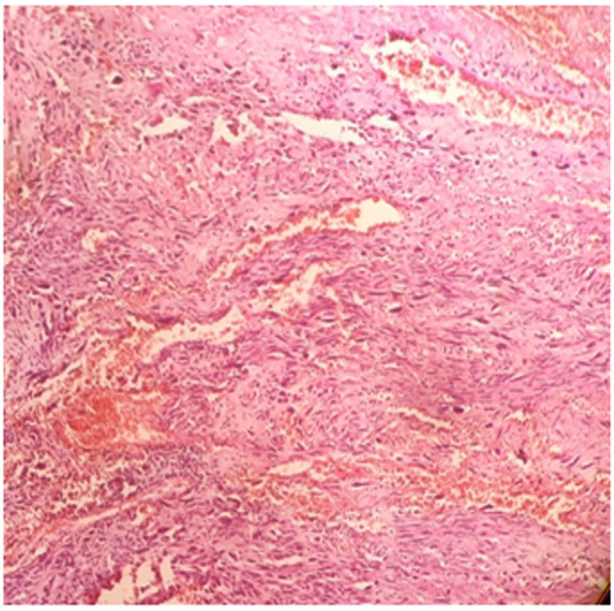

Fig. 2: Photomicrograph showing leiomyoma: tumor arranged in a diffuse sheath, whirling patterns having show multifocal bizarre cells. (Haematoxylin & Eosin stain, x10)

The low mitotic activity (<2 mitoses/10 high power fields) was noted. The staghorn vessels and thick walled vessels were noted. There was no necrosis. The areas of oedema, hemorrhage and congestion were noted. On histopathologycal finding reported as leiomyoma with bizarre nuclei.

Patient responded well to treatment. On follow up, there was no evidence of recurrence. The systemic examination was normal.

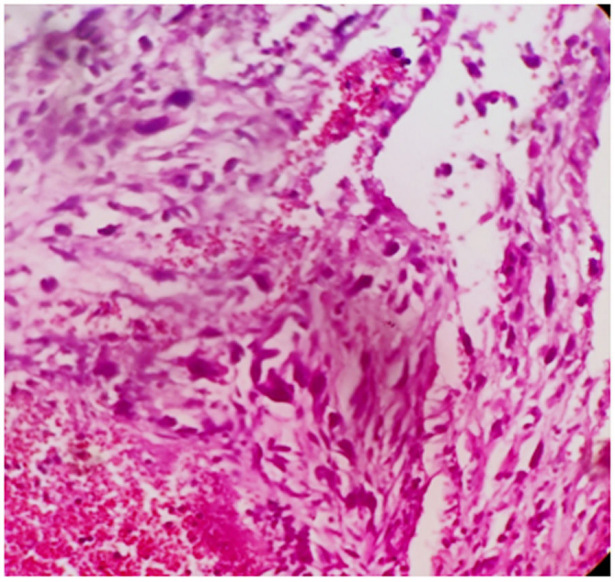

Fig. 3: Photomicrograph showing multinucleated bizarre cells that may have a diffuse distribution, prominent nucleoli, and karyorrhectic nuclei. (Hematoxylin & Eosin stain, x100)

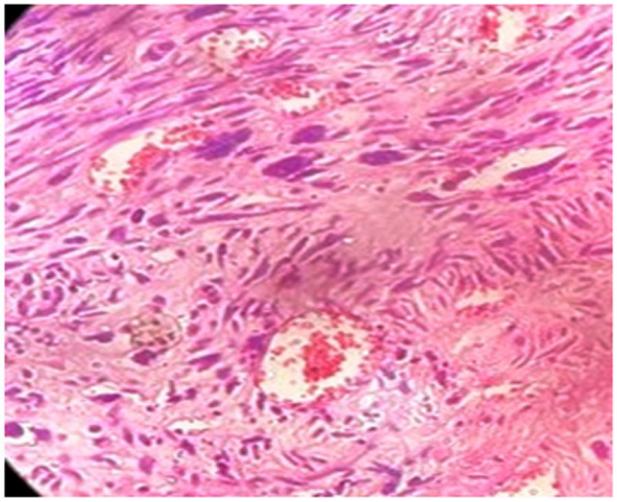

Fig. 4: Photomicrograph showing mononucleated or multinucleated bizarre cells in a background of typical leiomyoma. (Haematoxylin & Eosin stain, x100)

DISCUSSION

The leiomyomas are commonest benign tumors arising from uterine smooth muscle cells.[5]

LBN is usually an incidental finding in myomectomy or hysterectomy for leiomyoma cases. During grossing in LBN sampling and morphological examination remain the key to diagnosis. Clinically the most cases of LBN are asymptomatic or may present with pelvic pain, abnormal uterine bleeding, menorrhagia etc. The avarage age of presentation LBN is at 45 year. It is about 10-15 years younger than those with leiomyosarcoma.[6]

On gross examination tumors are non encapsulated, well circumscribed. The cut surface is white or tan, soft to firm consistency, and shows less bulging. The average size is of 7 cm.

The leiomyomas on histopathological features are of conventional types (90%), other subtypes are cellular, epithelioid, myxoid, leiomyoma with bizarre nuclei, fumarate hydratase (FH) deficient, intravascular, epithelioid, myxoid, lipoleiomyoma, mitotically active, leiomyomatosis.[7,8]

LBN is a variant of uterine leiomyoma, which has replaced the previous category of atypical leiomyoma. Leiomyoma with bizarre nuclei (LBN), is an uncommon tumor with histologic features (mononucleated or multinucleated bizarre cells that may have a diffuse distribution, prominent nucleoli, and karyorrhectic nuclei that may mimic atypical mitoses) that must be distinguished from uterine smooth muscle tumors of uncertain malignant potential (STUMP) and leiomyosarcoma.

LBN have expansive margins, a multifocal distribution with a high density of the bizarre cells. LBN have significant cytologic atypia, but high mitotic rate and tumor cell necrosis are absent. The density and distribution of nuclear atypia vary from case to case. The 2020 WHO classification established an objective criterion, and indicated a mitotic index of 0–4/10 hpf for LBN and 5–9/10 hpf for STUMP.

LBN has a biological behavior consistent with a benign tumors. LBN presents with occasional recurrence and rare reports of malignant transformation. Travaglino A *et al* reported the risk of local recurrence is 1.9% and no evident risk of recurrence outside uterus.[9]

The differentiating LBN from other benign leiomyoma subtypes, uterine smooth muscle tumors of uncertain malignant potential (STUMP), Intravenous leiomyomatosis with nuclear atypia, leiomyosarcoma (LMS) can be diagnostically challenging related to its overlapping features. In post-operative hysterectomy specimens, pathological evaluation, morphological diagnosis remains the gold standard. The mitotic count and tumor necrosis are the most important features that differentiate LBN from smooth muscle tumor of uncertain malignant potential (STUMP) or LMS. Discriminating between these two lesions therefore highly relevant in terms of prognosis and patient management. For the uterine smooth muscle tumors, the definitive diagnosis of is important for the determination of the prognosis of the patient and the most appropriate therapeutic approach.

The treatment for uterine leiomyomas can vary depending on the clinical menifestations, size, number, type, age of patient and location. Uterine leiomyoma may have various effects on pregnancy and fertility. Uterine leiomyoma complicated by gestational trophoblastic disease is very rarely noted.[10,11]

Surgical removal by laparoscopic, transvaginal or transcervical approaches as hysterectomy or myomectomy is mostly done. Uterine fibroid embolization, are other modalities. Sabrina Croce *et al*, observed that LBN is associated with a favorable outcome even in those patients only treated by myomectomy and highlights that a conservative approach can be useful in patient of reproductive age.

The various recent studies have described that immunohistochemical, biomarkers, molecular high-resolution mapping for gene mutations, gene expression, gene methylation, and genomic DNA copy number changes characteristics of LBN and LMS could be a future research direction.

CONCLUSION

LBN is a rare, benign and unique leiomyoma variant of uterine smooth muscle tumor. The diagnosis of LBN remains a challenge due to its nuclear atypia and its histologic heterogeneity reminiscent of leiomyosarcoma. We are presenting this uncommon case of leiomyoma with bizarre nuclei for its clinical, radiological and histopathological findings.

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