

Plexiform Tumorlets of Uterus: A Case Report

¹Ujjawal Khurana, ²Gunjan Raghubanshi, ³Sunita Arora,
⁴RPS Punia, ⁵Harsh Mohan

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¹Assistant Professor, ²Ex Demonstrator, ³Ex Assistant Professor, ⁴Professor, ⁵Retd. Head and Professor

¹Department of Pathology and Lab Medicine, AIIMS, Bhopal, Madhya Pradesh, India

^{2, 4, 5}Department of Pathology, Government Medical College and Hospital, Chandigarh, India

³Department of Obstetrics and Gynecology, Government Medical College and Hospital Chandigarh, India

ABSTRACT

Plexiform leiomyoma, a variant of epithelioid leiomyoma of the uterus is an incidental finding in hysterectomy specimens. Small plexiform leiomyomas that are detected only on microscopic examination are referred to as plexiform tumorlets. A case of plexiform tumorlets mimicking as metastatic lobular carcinoma of the breast is discussed.

Keywords: Lobular carcinoma, Menorrhagia, Plexiform leiomyoma, Tumorlet.

INTRODUCTION

Plexiform leiomyoma, a variant of epithelioid leiomyoma of the uterus are incidental findings in hysterectomy specimens. Small plexiform leiomyomas that are detected only on microscopic examination are referred to as plexiform tumorlets. Microscopically, the “Indian-file” pattern of epithelial-like tumor cells of plexiform leiomyomas may be confused with metastatic lobular breast carcinoma which is one of the most frequent extragenital neoplasms metastasizing to the uterus. This case presents multiple plexiform tumorlets mimicking as metastatic lobular carcinoma breast.

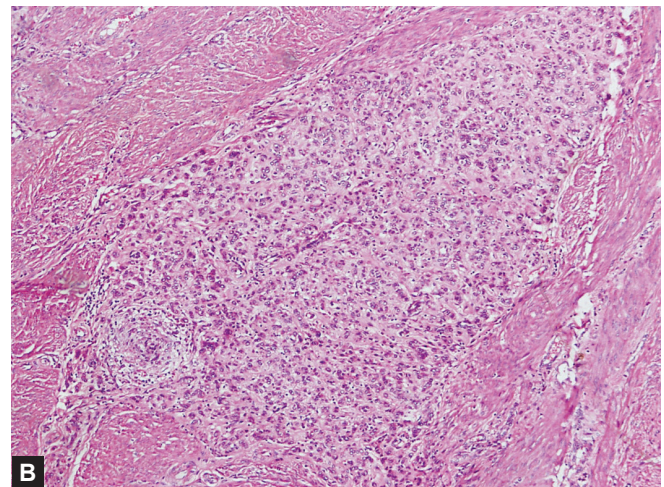
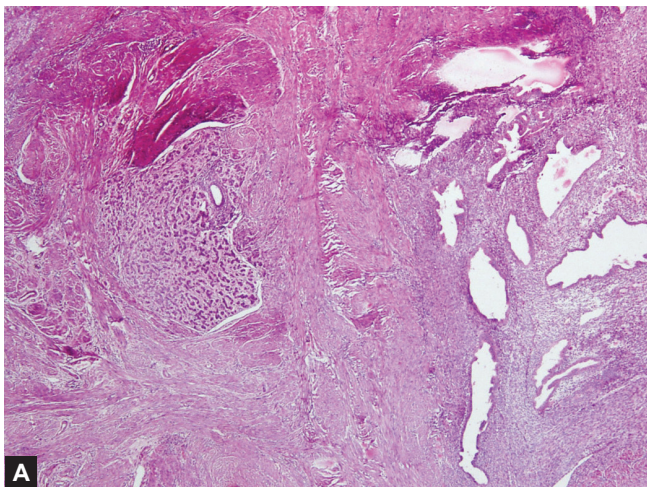
CASE REPORT

A 38-year-old woman, P₂L₂, presented with menorrhagia of two years duration. Her symptoms did not improve on medications. Since her family was complete, a total vaginal hysterectomy was performed.

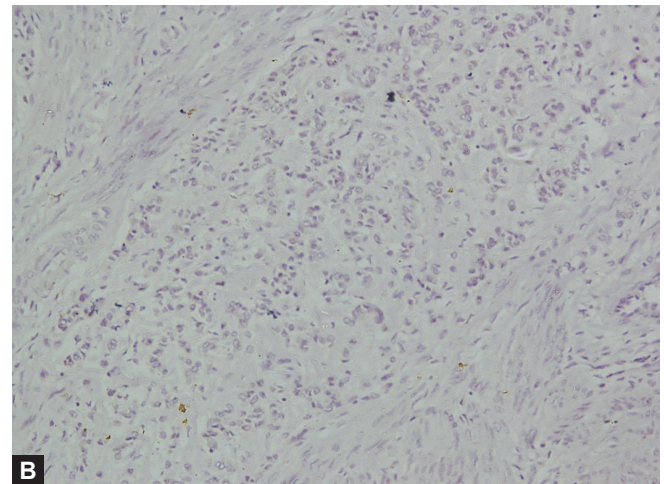
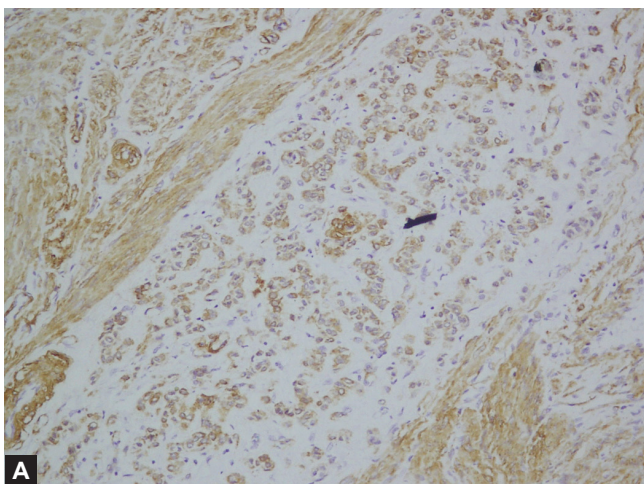
On gross examination, the total hysterectomy specimen measured 9.5 × 6 × 4 cm. It was unremarkable externally. On cut section, the endometrium/myometrium thickness measured 0.4/1.8 cm. Myometrium showed trabeculations. On serial

sectioning, a single intramural fibroid measuring 0.6 cm was identified.

Microscopically, the endometrium was in secretory phase. The myometrium showed adenomyosis and minute foci of tumors. These foci were well-circumscribed. The tumor cells were arranged in cords and nests and the stroma was hyalinized. The tumor cells were having round nuclei and moderate amount of pale to eosinophilic cytoplasm (**Figs 1A and B**). The cord-like arrangement of tumor cells was highly reminiscent of the Indian-file pattern seen in invasive lobular carcinoma of the breast. No mitosis or necrosis was seen. A possibility of metastasis from invasive lobular carcinoma was kept on the basis of this morphology. However, on clinical examination no breast lump was found. Meanwhile, special stains showed granular cytoplasmic positivity of tumor cells for periodic acid-Schiff and negative result on mucicarmine stain. Immunohistochemical examination revealed diffuse strong cytoplasmic positivity for smooth muscle actin and focal desmin positivity while cytokeratin staining was negative (**Figs 2A and B**). Thus a diagnosis of plexiform tumorlets of epithelioid leiomyoma was given on histopathology.



Figs 1A and B (A) Microscopic plexiform tumorlet in adenomyotic foci completely surrounded by normal myometrium. Also endometrium in secretory phase is seen (H&E, 50X). (B) Another tumorlet from the same case. Note the cord-like arrangement and epithelioid appearance of the tumor cells (H&E, 100X)



Figs 2A and B (A) Tumor cells show strong diffuse cytoplasmic positivity for smooth muscle actin. The surrounding myometrium is the positive internal control (H&E, 100X). (B) Tumor cells are negative for cytokeratin (H&E, 100X)

The patient has been maintaining well six months post-surgery.

DISCUSSION

Plexiform tumorlets are rare lesions with only few cases reported so far. These were first described in 1958 by Borghard-Erdle and Hirsch who believed it to be a glomus tumor. Generally, an incidental microscopic finding in a uterus removed for metromenorrhagia and uterine fibroids, they have mostly been seen in middle-aged women. It has been suggested that this association with adenomyosis or uterine fibroids is purely coincidental.^{1,2}

Small plexiform leiomyomas detected only on microscopic examination have been called plexiform tumorlets. The name 'plexiform tumorlet' has been given in analogy with the carcinoid tumorlets of lung.² These tumorlets are frequently solitary and submucosal in location though they can also be multifocal as was seen in our case.³ These can be located anywhere in the myometrium and even in the endometrium. They are defined by a plexiform architecture and a size smaller than 10 mm.^{1,4}

On microscopy, these are well-circumscribed tumors, completely surrounded by normal myometrium and are composed of round or polygonal tumor cells with scanty to moderate eosinophilic or clear cytoplasm. These cells

are arranged in nests or serpiginous cords.³ The typical plexiform pattern is produced by the hyalinization of the stroma present around these cords and nests.⁵ These are believed to be a subtype of epithelioid leiomyomas which are distinguished from the usual leiomyoma by the predominance of rounded or polygonal rather than spindle-shaped cells and by a clustered or cord-like pattern. This round or polygonal shape of the tumor cells gives the appearance of epithelial cells, hence the name 'epithelioid leiomyoma'. To call it so, these cells should be present in at least 50% of the tumor.⁴

On special stains, these are PAS positive diastase sensitive because of glycogen content and they are negative for mucin stains because of lack of mucin.³ Immunohistochemically, the tumor cells are negative for cytokeratins, synaptophysin, chromogranin, EMA, S-100 protein, and inhibin. However, they are strongly positive for smooth muscle actin and desmin indicating that they are of myoid and not epithelial nature.

The differential diagnosis of plexiform tumorlets of epithelioid leiomyomas may include metastatic lobular carcinoma of breast as was in the present case. Microscopically, the Indian-file pattern of epithelial-like tumor cells of plexiform leiomyomas may be confused with metastatic lobular breast carcinoma which is one of the most frequent extragenital neoplasms metastasizing to the uterus.² The "plexiform pattern" or "sex-cord like pattern" is encountered in a variety of uterine conditions such as endometrial stromal nodule, endometrial stromal sarcomas, poorly differentiated carcinoma of the endometrium and adenomyosis.^{2,6} Plexiform tumorlets and uterine PEComas also show considerable morphologic and immunophenotypic overlap. Fadare and Liang have proposed that positivity for CD1a immunohistochemical stain in uterine PEComas may be useful to differentiate it from plexiform tumorlets.⁷

Ultrastructural studies have shown that plexiform tumorlets show features of smooth muscle differentiation.³ Studies have also demonstrated increased expression of *HMGA2* gene responsible for cell proliferation and differentiation and up-regulation of *COL1A* responsible for synthesis of extracellular matrix.⁸ Plexiform tumorlets invariably are considered to be benign lesions though very rare cases of malignant plexiform tumors have also been cited in the literature.^{2,9}

Circumscribed margins, extensive hyalinization, and predominant clear cell type morphology are associated with benign behavior. Lack of mitosis, atypicality and necrosis further favor benignancy.³ It has been proposed that these are a subtype of the ordinary leiomyomas and as with the ordinary leiomyomas benign evolution without recurrence seems the rule. However, long-term follow-up of patients is not documented.

To conclude, the entity is discussed and reported so as to avoid mistaking this benign totally surgically resectable entity for metastatic tumor deposits.

Address for Correspondence

Ujjawal Khurana
Assistant Professor
Department of Pathology and Laboratory Medicine
All India Institute of Medical Sciences (AIIMS)
Bhopal, Madhya Pradesh, India
ujjawalkhurana@rediffmail.com
ujjawal.patho@aiimsbhopal.edu.in

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