CASE REPORT

Intravenous Leiomyomatosis: A Silent Killer

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Abstract

We present a case of 30 years female presenting with irregular menstrual bleeding, diagnosed as extensive leiomyomatosis on ultrasonography, confirmed on histopathology and immunohistochemical markers Desmin and CD34. Intravenous leiomyomas are benign tumors of smooth muscle origin. Long-term follow of patients is necessary of patients diagnosed to have these tumors.

Keywords: Intravenous leiomyoma, benign, immunohistochemistry (IHC), Desmin, CD_{34}.

INTRODUCTION

Intravenous leiomyomatosis is a rare uncommon type of uterine tumor defined as "an intraluminal growth of benign smooth muscle cells in either venous or lymphatic vessels outside the confines of or even in absence of leiomyoma."[1]

Though it was reported by Birch-Hirshfield in 1896, it was in 1959 that the microscopic features were described by Marshall and Morris.[2,3]

CASE HISTORY

A 32-year-old female presented with menorrhagia and abdominal mass. Ultrasonography of abdomen revealed multiple intramural and serosal fibroids in the uterus. Rest of the pathological and biochemical investigations were within normal limits. Hysterectomy with bilateral salphingo-oophorectomy was performed. Specimen was grossed according to standard protocol. Paraffin sections were stained with hematoxylin and eosin. IHC was done using antibodies against Desmin and CD_{34}.

Specimen consisted of bulky distorted uterus and cervix measuring 24 × 20 × 11 cm (Fig. 1).

External surface of uterus showed multiple serosal fibroids, largest measuring 15 × 13 × 10 cm and smallest measuring 1 cm across. Tiny linear grey white nodules were also seen on the external surface and the broad ligament.

Cut section of uterus showed multiple intramural fibroids. Cut section of fibroids showed grey white whorled appearance (Fig. 2). On squeezing the uterus, multiple worm like protrusions was seen arising from the blood vessels of the myometrium, broad ligament and isthmus.

Hematoxylin and eosin stained section studied from myometrium showed benign proliferation of smooth muscle cells (Fig. 3), without any cytological atypia consistent with leiomyoma. Secondary changes like hyaline change and clear cell change were seen.

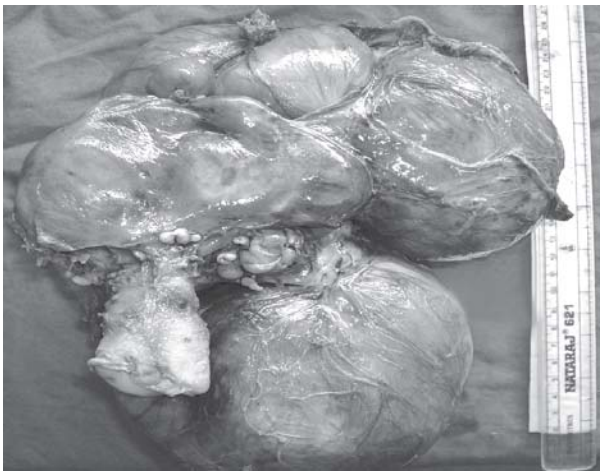

Fig. 1: Specimen of bulky distorted uterus and cervix measuring 24 x 20 x 11 cm

There were islands of tumor cells in the venous channels. These tumor cells were attached to the vessel wall, at places they were separated from the vessel wall by cleft like spaces within the tumor cells were freely floating. Tumor emboli were also seen in the blood vessels of the broad ligament and on the serosal surface of uterus.

Secondary changes like hyaline change and clear cell change were seen in the intravascular component. Mitosis was less than 3 per 10 high power fields.

Immunohistochemistry was done using antibodies against Desmin and CD_{34}. Smooth muscle nature of tumor cells was confirmed by Desmin positivity. CD_{34} was positive in the endothelium of the vascular channels (Fig. 4).

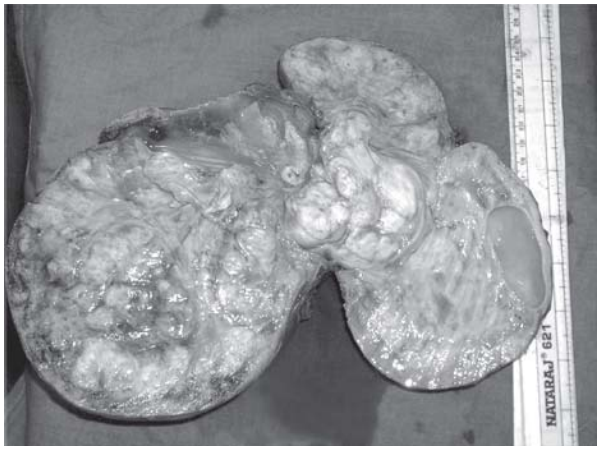

Fig. 2: Cut section of uterus with fibroids showing grey white whorled appearance

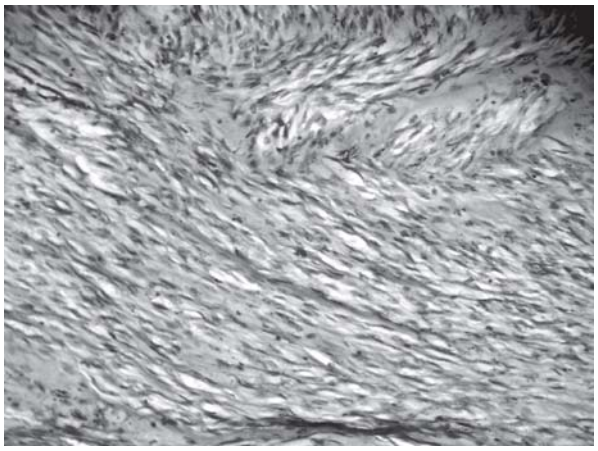

Fig. 3: Section studied from myometrium showed benign proliferation of smooth muscle cells, without any cytological atypia consistent with leiomyoma. H and E stain—100X

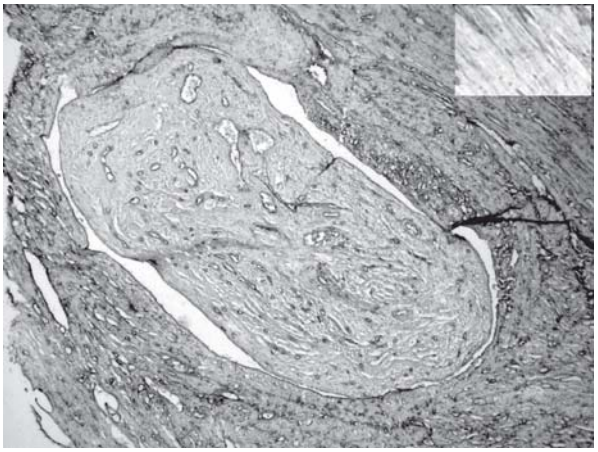

Fig. 4: CD_{34} positive in the endothelium of the vascular channels containing the vascular leiomyoma—400X Inset shows Desmin positivity of tumor cells. IHC—100X

DISCUSSION

Intravascular leiomyoma is a distinct variant of leiomyoma occurring exclusively in females of reproductive age group, presenting with symptoms of uterine fibroid such as pelvic pain and abnormal uterine bleeding.[1]

Though benign, these tumors behave in a malignant fashion by extensive spread into the pelvic veins with pelvic recurrences, obstruction of inferior vena-cava, extension into the right heart through inferior vena-cava and pulmonary metastasis.[1,4]

Microscopy of the intravascular component is similar to the conventional leiomyomas, other microscopic variants such as clear cells type, epithelioid type, myxoid change, lipoleiomyomatosis, intravenous leiomyoma with bizzare nuclei, extensive hyalinization, necrosis, intravenous leiomyoma with endometrial glands, and intravenous leiomyomas with extensive thrombosis have been described.[1,5,6] Our case showed extensive hyaline change, clear cell change and thick walled blood vessels in the leiomyoma.

On microscopy, intravenous leiomyoma has to be differentiated from leiomyoma with artifactual retraction from surrounding myometrium, leiomyoma with vascular invasion, leiomyoma with perinodular hydropic change and low grade endometrial stromal sarcoma.[4]

Immunohistochemistry aids in the diagnosis of the intravascular leiomyoma and differentiating from other tumors. Most of these tumors are positive for smooth muscle specific actin, Desmin and Caldesmon. The intravascular presence of the tumor is demonstrated by vascular markers like CD_{34}, ulex europeus.[1]

Recurrence rate of 30% has been reported in patients with intravenous extension.[1] Histogenesis of this tumor is very uncertain. Some authors suggest origin of these tumors from uterine smooth muscle, while others suggest the origin from smooth muscle of blood vessels.

Intravenous leiomyomas are histologically benign tumors with quasimalignant behavior with potential to spread to distant organs along the blood vessels giving rise to life-threatening complications. Careful follow-up of these patients throughout life is essential.

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