Case Report

A Leiomyomatous neoplasm of uterus - report of a case highlighting diagnostic difficulties and the role of immunohistochemistry

Priya S.[1*], Lakshmikanth R. M.[2], Nirmala V.[3]

[1,2]Assistant Professor, Department of Pathology, Chennai Medical College Hospital and Research Centre, Trichy, Tamilnadu, INDIA.
[3]Professor, Department of Pathology, PSG Institute of Medical Sciences and Research, Peelamedu, Coimbatore, Tamilnadu. 641 004, INDIA.
Email: drspriya78@gmail.com

Abstract

Introduction: A case of uterine leiomyoma with unusual morphological features is reported. The tumor arose from the outer anterior wall of the uterus, growing into the broad ligament, between the uterus and bladder. Macroscopically the tumor was large, partially cystic and had areas resembling lymphangioma. Solid areas of the tumor consisted of spindle mesenchymal cells in fascicles and round epithelioid cells in a perivascular arrangement. Role of immunohistochemistry in the evaluation of the cystic lymphangioma -like component and in the differential diagnosis of combined mesenchymal – epithelioid cell tumours of uterus is discussed.

Keywords: Combined Smooth Muscle - Endometrial Stromal tumors, Cystic degeneration, Epithelioid Leiomyoma, Immunohistochemistry.

[*]**Address for Correspondence:**
Dr. Priya S., Assistant Professor, Chennai Medical College Hospital and Research Centre, Trichy, Tamilnadu, INDIA.
Email: drspriya78@gmail.com
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INTRODUCTION

Leiomyoma of uterus is perhaps one of the commonest tumors encountered in routine practice of diagnostic pathology and the surgical pathologist is more than familiar with its morphology and variants. One such variant which could at times pose a diagnostic challenge is the epithelioid leiomyoma, defined as a smooth muscle neoplasm composed of uniform round cells with eosinophilic, vacuolated or clear cytoplasm.[1,2] Transition from conventional leiomyomatous component is often seen.[1] These neoplasms are generally described as circumscribed solid masses resembling conventional leiomyoma, though softer in consistency.[1,2] cystic degeneration is known to occur, as in conventional leiomyomas[3], and can be massive.[4] Microscopically, the combination of epithelioid/clear cell and spindle shaped smooth muscle cells make it necessary to distinguish epithelioid leiomyomas from PEComas(Perivascular epithelioid cell tumours) and mixed endometrial stromal – smooth muscle tumours[5,6]. Here we report a case of leiomyoma with epithelioid features which presented as a large cystic mass projecting from the outer uterine wall into the broad ligament. The cyst-wall contained multiple thin walled spaces resulting in a honeycomb appearance resembling lymphangioma grossly as well as microscopically. A solid nodule projected into the largest cystic cavity, consisting of spindle and round "epithelioid" cells. Immunostaining revealed the true nature of cystic spaces and that of the bimodal cell populations of the tumour.

MATERIAL AND METHOD

Clinical Summary

A 52 year old woman presented with vaginal bleeding of 15 days duration following five months of amenorrhoea. Ultrasound revealed a cystic mass of varied echo texture extending between the uterus and the bladder. Exploratory laparotomy was performed and the mass was

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seen in between bladder and the uterus, adherent to the uterus with the left round ligament stretched over it. The mass was clamped and removed, followed by total abdominal hysterectomy and bilateral salphingoophorectomy. The cystic mass and uterus with bilateral tubes and ovaries were fixed in 10% neutral buffered formalin and sent to the histopathology laboratory. Samples were embedded in paraffin, sectioned, stained with hematoxylin and eosin and the sections were examined under light microscope. Immunohistochemical studies were performed using the standard supersensitive polymer- HRP IHC detection system. The antibodies used were Anti- Actin, smooth muscle(1A4), (Biogenux, San Ramon, CA, USA, prediluted), Anti-Desmin (33),(Biogenux, San Ramon, CA, USA, prediluted), Anti-CD10 (56C6), (Biogenux, San Ramon, CA, USA, prediluted),Anti- CD34 (endothelial cell) (QBEND/10),(Biogenux, San Ramon, CA, USA, prediluted), Anti-Melanoma (HMB45),(Biogenux, San Ramon, CA, USA, prediluted),Anti-Melan- A (MART-1) (A103),(Biogenux, San Ramon, CA, USA, prediluted).

OBSERVATION AND RESULTS

Gross Appearance

The resected mass was partly cystic and measured 10 X 6.5 X 3.6 cms. Cut surface showed multiple cystic spaces with a 4cm firm white whorled nodule projecting into the largest of them. The locules had smooth lining and contents were clear, fluid. The cyst wall in areas had multiple tiny spaces with clear fluid within, resulting in a honey comb appearance "Fig.1". A raw area corresponding to the site of dissection from anterior uterine wall was present on the external surface.

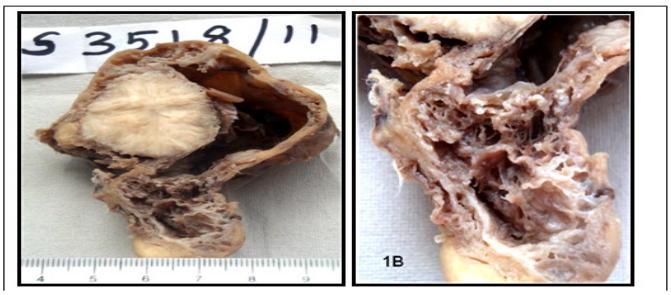


Figure 1A: Gross appearance of the resected mass the mass is partly cystic,multiloculate with a nodule projecting into the largest locule

Figure B: Multiple tiny spaces giving rise to honey combed appearance

Microscopic Appearance

H&EThe neoplasm consisted of fascicles of spindle cells separated by abundant hyaline matrix, interspersed with groups and cords of uniform round cells with eosinophilic to vacuolated cytoplasm intimately associated with vasculature "Fig.2 A&D".The spindle and epithelioid components merged with each other. In addition there were multiple large cystic spaces, and focal collections of smaller vascular channels "Fig. 3". The tumor had areas consistent with conventional leiomyoma as well as epithelioid areas resembling an epithelioid variant or an endometrial stromal element. The spindle and round cells showed diffuse strong and unequivocal expression of desmin and smooth muscle actin "Fig.2 B &E". CD10 expression was observed only in a few of the perivascular round cells and in a rare spindle cell "Fig.2 C&F". The large cystic spaces showed no endothelial lining on staining for CD 34. But the focal collections of small vessels were lined by CD 34 + cells "Fig.3 B&C". Mitotic figures were not seen. Nuclear atypia and necrosis were absent.

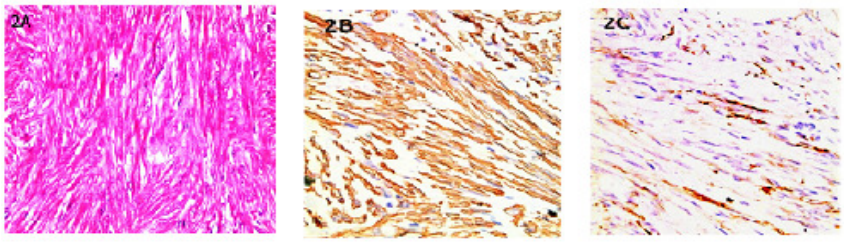


Figure 2A: Spindle cell component, composed of fascicles of cells. H and E X400

B: Diffuse, strong expression of Desmin in the spindle cells X 400

C: Rare focal positivity for CD 10 in spindle cells X 400

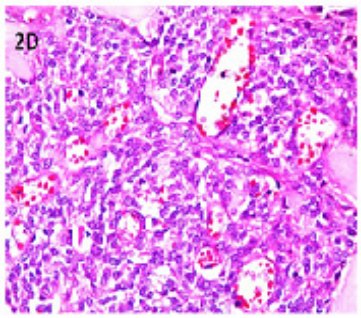
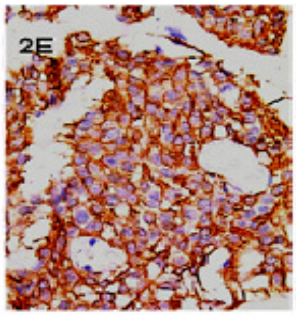
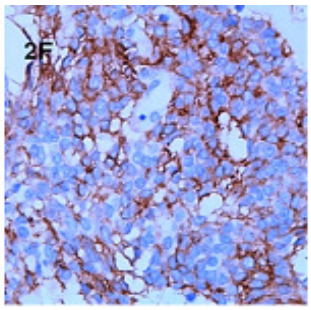


Figure 2 D: Epithelioid component composed of round to polyhedral cells having eosinophilic to vacuolated cytoplasm H and E X400



E: Desmin expression in the round cells is diffuse and strong X 400



F: Some of the round cells are CD 10 positive X 400

DISCUSSION

Multiloculated cystic epithelioid leiomyomas are unusual, and those resembling lymphangiomas, exceptionally rare. Magro and Manusia reported one such case in which the larger cystic cavities were considered to represent intratumoral degeneration, as they lacked an endothelial lining and basement membrane[7]. Here we present another case that had identical macroscopic and microscopic features. The peculiar gross morphology and the presence of multiple cystic cavities raised the suspicion of an angiomatous component in association with an otherwise clearly leiomyomatous neoplasm. As in the previous case, immunostaining for endothelial markers revealed absence of endothelial lining suggesting that the cavities were degenerative in nature. Microscopic lymphangioma like collections of endothelial lined small vascular channels were observed in the present case. Interestingly, the earlier authors have also described such foci in their tumour, which they interpreted as a histological variation in leiomyoma. The solid portions of the tumor consisted of fascicles of spindle cells and round to oval epithelioid cells, the latter closely mingled with rich vasculature. The light microscopic appearance on routine staining suggested either a leiomyoma variant or a combined smooth muscle – endometrial stromal tumor. Distinction between the two is necessary because endometrial stromal tumors can be aggressive and can respond to anti - estrogen therapy. Light microscopic features such as a lack of clear demarcation of the two components and small volume of the epithelioid component help to distinguish between the two to some extent. Immunostaining for muscle and endometrial stromal cell markers can also be applied in this context, but interpretation needs care. Both endometrium and myometrium arise from Mullerian ducts and can express identical antigens [8]. It has been generally agreed that pure smooth muscle neoplasms, while showing diffuse expression of desmin and smooth muscle actin, rarely express CD10, the endometrial stromal cell marker[9]. But co- expression of CD10 can be found in the epithelioid component of otherwise typical cellular leiomyomas[8]. Desmin expression in neoplastic endometrial stromal cells tends to be absent or occasional focal, if present[6]. The tumor reported here expressed desmin and smooth muscle actin throughout, with some focal co- expression of CD10 in part of the round cell component. This immune profile favours a smooth muscle origin, supported by the merging of the spindle and round cell areas into one another. Perivascular distribution of epithelioid cells along with the lymphangiomyomatous areas which have already been described raised the possibility of a perivascular epithelioid cell tumor(PEComa)[5].The location of the tumour also favored this diagnosis. The cells did not express melanocyte markers (HMB45, Melan-A), thus excluding the possibility.

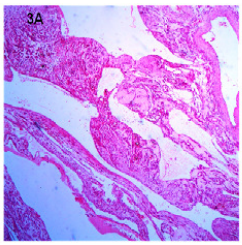
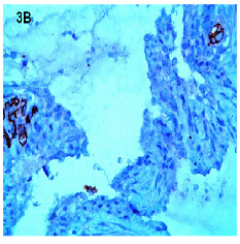
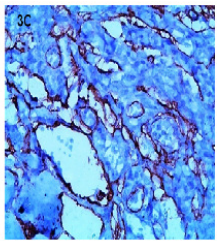


Figure 3A: Multiple cystic spaces imparting a ''lymphangiomatous'' appearance. H and E X 100



B: CD 34 immunostaining of cystic spaces – no endothelial lining is seen Endothelial cells of blood vessels are strongly positive and serve as internal control X 400



C: CD34 immunostaining showing ramification of capillary sized vessels imparting a ''lymphangiomatous'' pattern X400

CONCLUSION

In conclusion we report a common neoplasm, garbed in unusual morphology, transformed into a diagnostic puzzle. The case further illustrates the role of immunohistochemistry in such a context. Judicious interpretation of the immunostaining patterns with standard markers is essential when dealing with mixed epithelioid- mesenchymal uterine tumours.

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