Case Report

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Pathology Section

Epithelioid Leiomyoma of Uterus: A Case Report

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ABSTRACT

Uterine smooth muscle tumours with distinctive epithelioid smooth muscle cells are called as epithelioid leiomyoma or leiomyoblastoma as the cells resemble smooth muscle cells of foetal uterus. We report a case of epitheliod leiomyoma of uterus in a South Indian peri-menopausal female. Pathological examination showed a single sub-serosal fundal mass measuring 15 x 14 x 10 cm in diameter. Cut section of the mass showed soft to firm grey white areas along with cystic spaces. Microscopy revealed sheets and peri-theliomatous arrangement of round to polygonal cells with eosinophilic cytoplasm and vesicular nucleus. Immunohistochemically, the polygonal cells stained strongly and positively with Smooth Muscle Actin (SMA) and desmin and negative for HMB-45 stain. The Ki-67 proliferative index was less than 1%.

Keywords: Caldesmon, Leiomyoblastoma, Smooth muscle actin

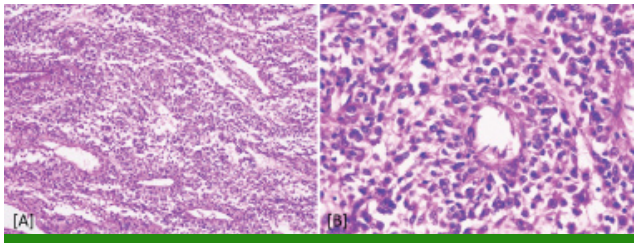
CASE REPORT

A 42-year-old peri-menopausal woman presented with abdominal distension to the Department of Gynaecology. Past menstrual history revealed normal menstrual cycles. Patient's general and systemic examination was normal. Ultrasound abdomen showed a bulky uterus with large hyperechoic areas measuring 12 x 10 x 10 cm with cystic spaces in the fundus. The ultrasound findings were suspicious for fibroid, with suggested histopathologic examination to rule out malignancy. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed. Post operatively the specimen was sent for histopathological examination after obtaining the informed consent.

Pathological examination showed a single sub-serosal fundal mass measuring 15 x 14 x 10 cm in diameter. Cut section of the mass showed soft to firm grey white areas along with cystic spaces and focal haemorrhagic areas [Table/Fig-1]. Microscopy revealed sheets and peri-theliomatous arrangement of round to polygonal cells with eosinophilic cytoplasm and vesicular nucleus with few showing resinoid changes. Mild pleomorphism of cells, with mitotic count of 0-1/50 High-power fields (HPF) was seen. Focal areas showed spindle shaped cells with elongated nucleus [Table/Fig-2]. Cystic myxoid degeneration and hyalinization was also seen. No necrosis or severe atypia was identified. A light microscopic diagnosis of epithelioid stromal neoplasm of uterus was made, although the differential diagnosis of Perivascular Epithelioid Cell Tumour (PEComas), and endometrial stromal tumour were considered.

Immunohistochemically, the polygonal cells stained strongly and positively with smooth muscle actin and positively with desmin. However, the polygonal cells were negative for HMB-45 stain. The Ki-67 proliferative index was less than 1% [Table/Fig-3].

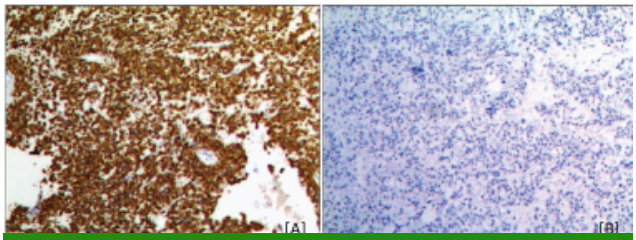
[Table/Fig-1]: Cut section of the fundal mass shows soft to firm grey white areas with cystic spaces.



[Table/Fig-2]: Microscopy showed (A) Peri-theliomatous arrangement of round to polygonal cells [Haematoxylin and Eosin in 10] (B) Round to polygonal cells with eosinophilic cytoplasm and esicular nucleus [Haematoxylin and Eosin in X 40].

DISCUSSION

Uterine smooth muscle tumours or leiomyomata are benign tumours with spindle shaped tumour cells that resemble



[Table/Fig-3]: Immunohistochemical characterization of tumour shows (A) strong positivity for SMA (B) negative staining for HMB-45.

those that of normal myometrium [1]. Sometimes, the leiomyomata consist of smooth muscle cells, which appear rounded resembling epithelioid cells. Uterine smooth muscle tumours with distinctive epithelioid smooth muscle cells, in more than 50% of tumour are called as epithelioid leiomyoma [2]. These are also called as leiomyoblastoma as the cells resemble smooth muscle cells of foetal uterus [1,3,4].

Uterine epithelioid leiomyomas are rare tumours characterized by round to polygonal cells along with mixture of clear cells with few areas showing transition to typical smooth muscle cells [1,3]. Immunohistochemical characterization of tumour includes reactivity for smooth muscle markers like actin, desmin and myosin. They are otherwise called as leiomyoblastoma because of immunohistochemical and ultra-structural resemblance to immature smooth muscle mesenchymal cells of foetal uterus during 14-26 weeks. The restricted immunohistochemical positivity for high molecular weight caldesmon and ultrastructural abundance of intermediate filaments along with intermingled dense body like structures of tumour cells suggest a resemblance to immature smooth muscle cells [3].

Our case had sheets and peri-theliomatous arrangement of round to polygonal cells with minimal mitosis. Differential diagnosis of Perivascular epithelioid cell tumour (PEComas), and endometrial stromal tumour were considered. Negative staining for melanocytic marker HMB-45 ruled out the possibility of PEComa [5]. Strong positive staining for SMA, and focal areas showing spindle cell morphology favoured a diagnosis of leiomyoma over endometrial stromal tumour [3]. With 12 months of follow-up, there was no recurrence or metastasis in our case.

The need to recognise epithelioid leiomyoma as a distinct entity is important because the rules to prognosticate the entity are different from standard leiomyoma. There are uncertainties in predicting the behaviour of epithelioid leiomyoma. All epithelioid leiomoyomata with tumour cell necrosis have a malignant behaviour, whereas epithelioid leiomomyomata with less than 5/10 hpf mitotic activity, minimal cytological atypia, no necrosis have a benign course [1,2]. However, the course of epithelioid leiomyoma with 2 or more of the following features is not established: size greater than 6 cm, 2-4 mitotic figures/10 hpf, moderate to severe cytological atypia and necrosis. Hence, epithelioid leiomyomas with 2 or more of these following features are classified as tumours of uncertain malignant potential and close follow-up is required [6].

CONCLUSION

Since, the prognostication criteria like mitosis, atypia in epithelioid leiomyoma is different from standard leiomyoma, extensive sampling and awareness of the different rules of prognostication is essential in reporting the variants of leiomyoma and leiomyosarcoma. Moreover, due to the uncertain course of the larger variant leiomyoma's, close follow-up is required in those subjects.

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