

## CASE REPORT

**Cystic Clear Cell Epithelioid Leiomyoma: An Unusual Presentation***Varun Kumar Singh<sup>1\*</sup>, Tanvi Shetty<sup>1</sup>, Brij Mohan Kumar Singh<sup>1</sup>**<sup>1</sup>Department of Pathology, Melaka Manipal Medical College, Manipal Campus, Manipal Academy of Higher Education, Manipal-576104 (Karnataka) India***Abstract:**

Epithelioid leiomyoma is a rare variant of leiomyoma. The tumour includes three distinct entities, leiomyoblastoma, clear cell leiomyoma and plexiform leiomyoma. It is histologically characterised by epithelioid cells, clear cells or plexiform pattern in varied proportions. We report a case of a 49-year-old female with complaints of palpable mass per abdomen and spotting per vagina. Radiology revealed a complex cystic mass with obscured right ovary. She underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy. Pathological examination showed a posterior wall lesion with central 9 × 8 cm cyst with multiple septations and surrounding rim of solid areas. Microscopic examination showed a clear cell epithelioid leiomyoma. No mitosis/tumour cell necrosis was seen. The clinical course of epithelioid leiomyoma is still unclear owing to absence of large studies. Small size, clear cell histology, mitosis of < 2/10 High Power Field (HPF) and hyalinization are features of a benign course, whereas cellular atypia, mitosis of > 5/10 HPF are definite indicators of a malignant outcome. An epithelioid leiomyoma with, size >6 cm moderate atypia, necrosis and mitosis of 2-5/10 HPF are termed as smooth muscle tumours of uncertain malignant potential and should be under close follow up.

**Keywords:** Epithelioid Leiomyoma, Clear cell, Smooth Muscle Tumour, Uterus

**Introduction:**

Epithelioid smooth muscle tumours of the uterine corpus encompass varied benign and malignant entities. Unlike the epithelioid tumours in gastrointestinal tract, ones occurring in the female

genital tract are still sparsely reported. Epithelioid variant of leiomyoma, comprises of three entities namely leiomyoblastoma, clear cell leiomyoma and plexiform leiomyoma [1-2]. All these entities contain epithelioid cells, clear cells and plexiform pattern in varied proportions. Epithelioid leiomyomas are usually large tumours and show varied degree of degenerative changes, which mimic malignancy on radiological investigations. Prognosis of these tumours largely depends upon the nuclear atypia, necrosis and mitotic activity which are used to differentiate them from Smooth Muscle Tumours of Uncertain Malignant Potential (STUMP) and leiomyosarcoma [2-3].

**Case Report:**

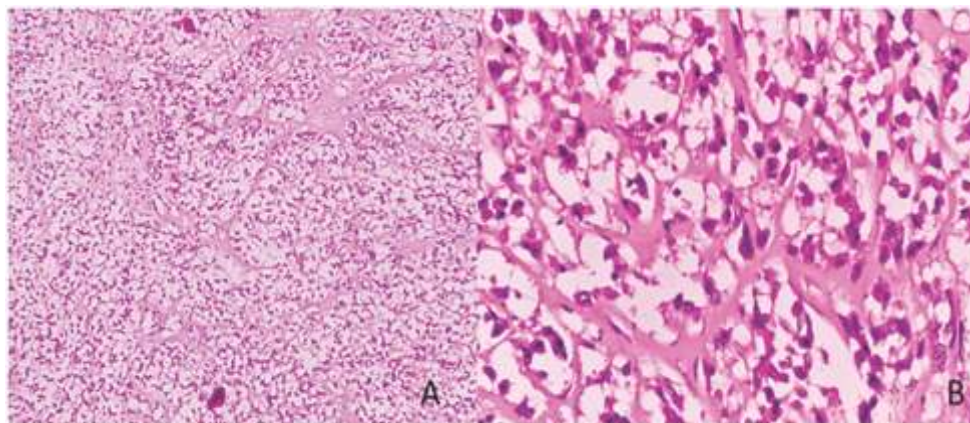
A 49-year-old post-menopausal female presented with palpable mass in the abdomen with a history of increase in size for last 3 months and spotting per vagina. There was no associated pain, vomiting, loss of appetite or weight loss. Pelvic ultrasonography revealed a large, predominantly cystic mass, approximately 9 × 8.5 × 7.0 cm in size, with multiple internal septae in the right pelvis and abdomen. The right ovary was not separately visualised, hence a clinical suspicion of an ovarian cystic mass or a degenerated fibroid was suggested. Patient underwent a total abdominal hysterectomy and bilateral salpingo-oophorectomy. On gross examination, the ovaries were unremarkable and the uterus was asymmetrically enlarged. Cut

section showed a cystic cavity with multiple internal thin septae measuring 9 × 8 cm and peripheral grey white solid areas with a circumscribed outer rim of myometrium (Fig. 1). Endometrial cavity was reduced to a slit like space. Histopathological examination showed a tumour composed of polygonal cells with abundant eosinophilic cytoplasm and vesicular nuclei. Extensive clear cell change comprising of > 50% of the tumour (Fig. 2), mild anisonucleosis and foci of

hyaline change (Fig. 3). An extensive sampling was done for mitotic activity, necrosis and nuclear atypia. No mitotic figures, necrosis was noted, and focal mild nuclear atypia was seen. The periphery of the tumour showed merging of the epithelioid cell pattern into the classic spindle cell pattern of a leiomyoma. Ovaries and tubes showed normal histology. Correlating the gross and microscopic features a diagnosis of cystic clear cell epithelioid leiomyoma was made.

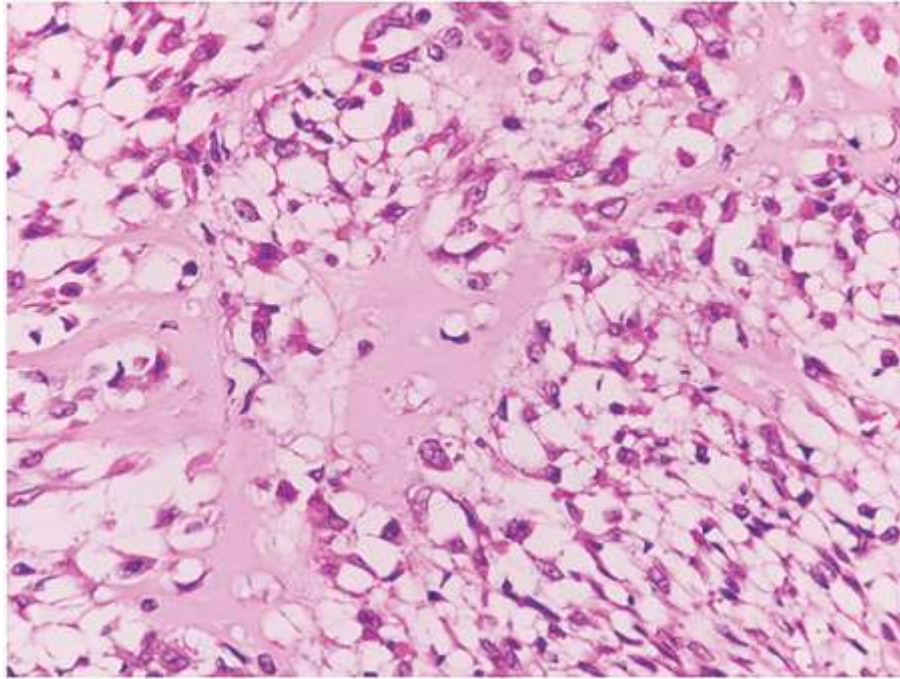


**Fig. 1: Cut Section of Uterus Showing Leiomyoma with Cystic Cavity, Multiple Internal Thin Septae, and Peripheral Grey White Solid Areas Circumscribed By Outer Rim of Myometrium**



**Fig. 2a: Tumour Composed Predominantly of Clear Cells (H&E, 40×).**

**Fig. 2b: Few Tumour Cells show Eosinophilic Cytoplasm Mild Nuclear Atypia along with Hyalinization and Clear Cell Change (H&E, 200×).**



**Fig. 3: Few Cells show Peripherally Pushed Nuclei Assuming A Signet Ring Morphology, along with Areas of Stromal Hyalinization (H&E, 200×)**

#### **Discussion:**

Epithelioid leiomyoma is a rare variant of leiomyoma reported in uterine corpus. These occur in over a broad age range of 30-78 years with a peak in the fifth decade. Per vaginal abnormal bleeding is the most common presenting feature. Other features include abdominal pain, menorrhagia and abdominal mass in a fraction of cases [1-2]. Grossly, these tumours are usually solitary, yellow or grey with haemorrhage and cystic spaces; they are less firm than usual leiomyoma and can occur in any part of the uterus [2]. The cysts are usually small, but large cystic change is also documented in these tumours [4-5]. These leiomyoma variants are usually large with a median size around 7 cm [1-2]. The present case had a similar presentation with an age of 49 years and a large complex cyst measuring 9 cm across in greatest dimension. Histological

spectrum of epithelioid leiomyoma comprises of patterns described as leiomyoblastoma, clear cell leiomyoma and plexiform leiomyoma in the literature. A mixture of these three patterns in various proportions is seen in an epithelioid leiomyoma providing the rationale behind combining all three under a single umbrella [1-2]. Leiomyoblastoma primarily is composed of polygonal eosinophilic cells and resembles the foetal smooth muscles [1-2,6]. Plexiform leiomyoma primarily shows extensive hyalinization with cords and nests of round eosinophilic cells, any tumour < 1 cm in size is referred to as a plexiform tumourlets [2]. Clear cell leiomyoma is the most common histological pattern amongst the three. The cells are polygonal, show cytoplasmic clearing, and well defined cell borders. The clear

cytoplasm is due to presence of glycogen. The nucleus can be pushed to periphery and can assume as signet ring morphology. The epithelioid cells/clear cells show foci of transition to the classical spindle cell leiomyoma morphology [2, 4, 7]. Such epithelioid morphology has been documented not only in uterine corpus but also in other sites like vagina, broad ligament, intravenous and cotyledon leiomyoma [8-14]. The present case showed predominant clear cell morphology, which merged into the spindle cell foci in the periphery. The present case also showed a large complex cystic change, which is an unusual presentation described only in a handful of case reports [4-5].

The behaviour of epithelioid leiomyoma is uncertain, primarily owing to lack of studies focused on this variant. Few histological features, which point to a benign course, include a small tumour, lack of atypia, lack of necrosis, infrequent mitosis, circumscribed margins, extensive hyalinization, and predominant clear cell morphology [2, 15]. Tumours with moderate to severe atypia, tumour necrosis and 2-5 mitosis/10

High Power Field (HPF) can be classified as STUMP and warrants a careful follow-up [15]. A mitosis rate of > 10/10 HPF points to a diagnosis of leiomyosarcoma even in absence of tumour necrosis and cellular atypia, however, some reports state that any mitosis rate of > 5/10 HPF is enough to predict a malignant behaviour [1-2, 15]. An uncertainty regarding outcome still shrouds around epithelioid leiomyoma with necrosis, size > 6 cm, mitotic rate of 2-4/10 HPF and moderate to severe atypia [1-2]. The present case had benign histological features and mitosis but had a larger size. The patient was followed up for 9 months without any recurrences.

#### Conclusion:

The present case highlights an unusual presentation, cystic clear cell epithelioid leiomyoma, which is sparsely documented in the English literature. The complex cystic structure and irregular solid areas raise red flags, and warrants extensive sampling to rule out a malignant aetiology.

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\***Author for Correspondence:** Dr. Varun Kumar Singh, 305, Melaka Manipal Medical College, Manipal campus, Madhav nagar, Manipal, Udupi, Karnataka, India 576104. Email: varun.singh@manipal.edu, Cell: 9506423336