

A Rare case of Giant retroperitoneal uterine Leiomyoma with cystic degeneration mimicking huge ovarian cyst

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ABSTRACT

Introduction: Leiomyomas are the most common benign tumors of the uterus. Retroperitoneal uterine leiomyomas with cystic degeneration are very rare. They can pose diagnostic dilemma and can mimick ovarian cysts. **Case report:** We report the case of a 34 yrs old para 5 who presented to our hospital with complaints of distention whole abdomen, heaviness lower abdomen & decreased appetite. On examination a huge abdomino-pelvic cystic mass was felt. Clinically provisional diagnosis of ? Ovarian cyst was made. CT scan showed a huge retroperitoneal cystic mass. Laprotomy and proceed was planned. Intraoperatively a huge retroperitoneal cyst attached with uterus just below the left fundal structures by a pedicle and extending upto xiphisternum was found. Uterus was of normal size and both the ovaries and tubes were distinctly separate from the mass. Left ovary was slightly enlarged and unhealthy in appearance. Cyst excision followed by total abdominal hysterectomy and left sided salpingo-oophorectomy was done. Histopathological examination revealed degenerated leiomyoma with immunohistochemistry positive for smooth muscle actin. To the best of our knowledge this case is the largest retroperitoneal uterine leiomyoma with cystic degeneration in literature.

Conclusion: Retroperitoneal uterine leiomyoma with cystic degeneration are rare. They can pose diagnostic difficulty preoperatively and even peroperatively. As in our case the retroperitoneal leiomyoma with cystic degeneration was mimicking a huge ovarian cyst. Though CT scan was guiding towards retroperitoneal lesion but its cystic nature created diagnostic dilemma.

Keywords: cystic degeneration, leiomyoma, Retroperitoneal

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INTRODUCTION: Leiomyomas, the most common benign neoplasm of uterus arise from smooth muscle cells [1]. Retroperitoneal leiomyomas are however

very rare [2-5]. Retroperitoneal uterine leiomyomata with cystic degeneration are rarer and can be mistaken for ovarian cyst or retroperitoneal cysts either neoplastic or non neoplastic [6]. Fair number of cases of retroperitoneal leiomyoma are reported [3,7,8]. But only few cases of retroperitoneal leiomyoma with cystic degeneration are reported in literature [9,10]. In all these reported cases none had uterine attachment.

This rare entity can pose diagnostic challenge for clinicians as well as for radiologist. We report a rare case of a huge retroperitoneal uterine leiomyoma with cystic degeneration.

CASE REPORT: 34 yrs old P5L4 presented to gynecological OPD of our hospital with complaints of distention whole abdomen, heaviness lower abdomen and decreased appetite for last 5 months. There was no history of menstrual disturbance, any bladder bowel complaint, chronic cough with fever, blood in stool or hematemesis. She was P5L4A0, with all full term normal deliveries at home, last child birth was 6 yrs back. Her menstrual periods were regular, lasting for 5-6 days with average flow without pain. She denied any positive past or

family history. On examination there was no pallor, systemic examination was within normal limits. On per abdominal examination a huge abdomino-pelvic cystic mass extending upto xiphisternum was felt. On per speculum examination cervix was pushed anteriorly and towards right side, vaginal mucosa seen bulging through all fornices. On per vaginal examination normal size uterus and cervix were felt high up and towards extreme right side and the same cystic mass felt per abdominally was felt through all fornices. Blood investigations were within normal limits. Ultrasonography showed a large heterogenous cystic lesion with solid components filling the entire abdominal cavity. CT scan showed a very large (32 × 24 × 15 cms approx.) abdominopelvic, well defined, smooth margined lesion predominantly cystic with few small solid areas and multiple thick septations. The lesion was causing displacement of descending colon anteriorly and towards right side suggesting retroperitoneal location [Figures 1a & 1b]. Both solid components and septations showed post contrast enhancement. Uterus

and right adnexa were pushed rightwards, left sided adnexa not visualized.

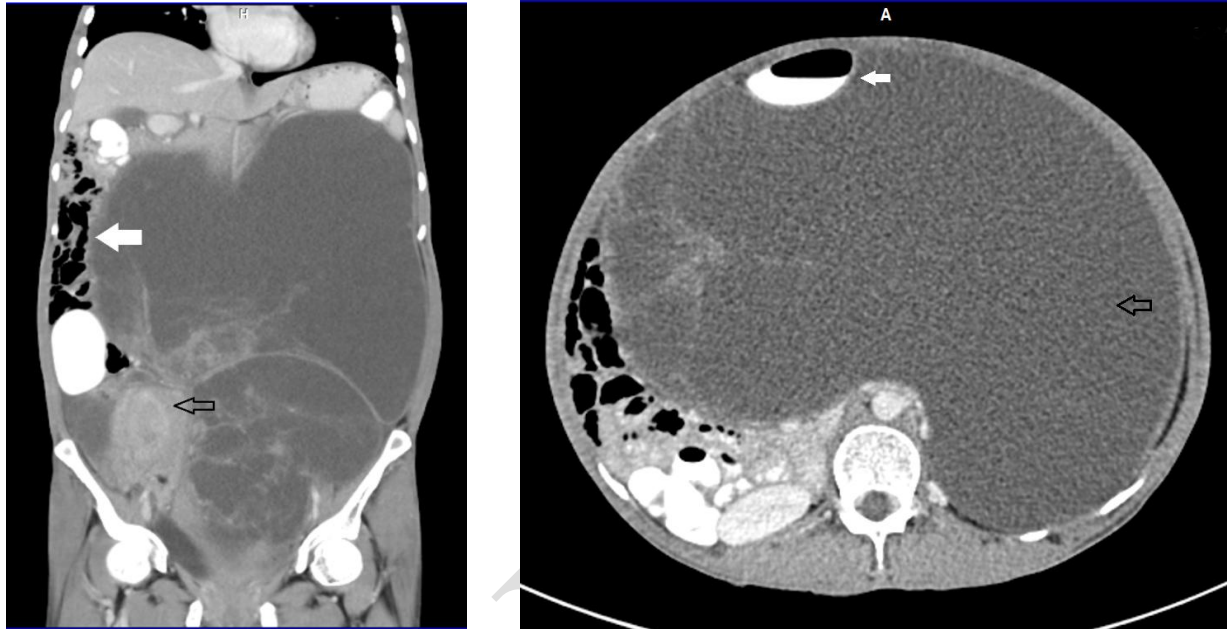


Figure 1: Contrast enhanced CT scan of retroperitoneal leiomyoma shows low attenuation huge mass occupying whole of the abdomen ;[a] black arrow shows uterus and white arrow shows compressed and displaced intestine [b] black arrow shows mass and white arrow shows anteriorly displaced descending colon

On laprotomy a huge cystic, lobulated retroperitoneal mass displacing the descending colon anteriorly and towards right side was seen .The general surgeons were involved, the mass was dissected off

retroperitoneally. Fluid was aspirated to reduce the bulk of mass. On tracing its origin and blood supply it was found to be attached with uterus on left side just below the left fundal structures [Figures 2a &2b].

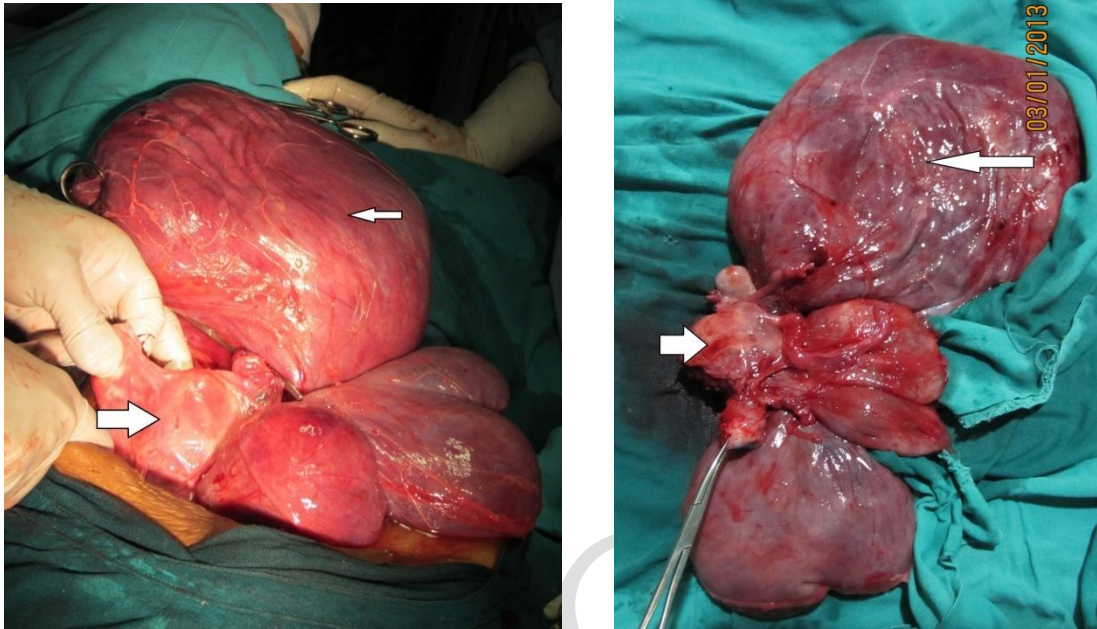


Figure 2: Large [22×17×6cm] retroperitoneal degenerated uterine leiomyoma (thin arrow) with other two smaller cysts and uterus (thick arrow) [a] intraoperatively and [b] in postoperative specimen

Uterus was of normal size. Bilateral ovaries were visualized separate from the mass. Two more cystic masses of approximately 10 cm size were found separate and below the above mass at the site of left broad ligament and were filling the whole pelvic cavity. Left ovary was enlarged and appeared unhealthy. Due to unavailability of frozen section examination facility, on clinical suspicion left salpingo-oophorectomy along with total hysterectomy was done following retroperitoneal dissection and removal of large cystic masses and the specimen in toto was sent for

histopathological examination. Postoperative period was uneventful, patient was discharged on 11th postoperative day. Histopathology report was suggestive of subserosal cystic palisaded leiomyoma with neural differentiation, rest of the findings were secretory endometrium, adenomyosis uteri, marked chronic cervicitis with no other remarkable finding in ovary and fallopian tubes [Figure 3]. To confirm the diagnosis immunohistochemistry was done which was positive for smooth muscle actin. KI-67 was positive up to only 1% suggestive of benign nature of the lesion. Patient was followed up

at one month, three month and six month and was found doing well.

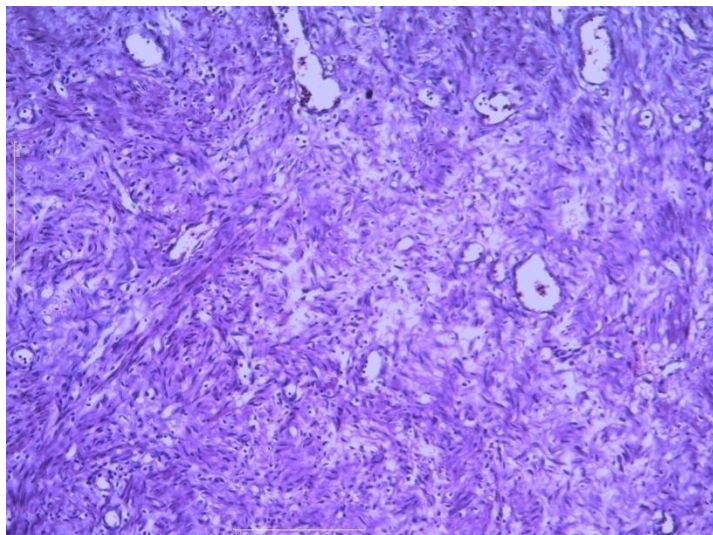


Figure 3: Histopathological examination of leiomyoma: H&E(10X) interlacing bundles of smooth muscles with compressed blood vessels

DISCUSSION: Leiomyomas are the most common benign tumour of the uterus and pelvis with an occurrence of 20%–30% in women of reproductive age [1,2,5]. They are benign tumors of smooth muscle cells but contain varying amounts of fibrous connective tissue [1,11]. In addition to the usual patterns of growth in the uterus, some unusual extra-uterine growth presentations are also seen like benign metastasizing leiomyoma, disseminated peritoneal leiomyomatosis, intravenous leiomyomatosis, parasitic leiomyomas and retroperitoneal growth [3,12]. Leiomyomas may enlarge enough to outgrow their blood

supply resulting in various types of degeneration, like hyaline, cystic, myxoid, or red degeneration and dystrophic calcification [1,8,9,11]. Hyaline degeneration is the most common change in leiomyomas. It is seen in 60% of leiomyomas [11]. Gradually the areas of hyalinization may undergo liquefaction and cystic cavities filled with clear liquid or gelatinous material are formed. Sometimes the cystic change is so great that the leiomyoma becomes truly a cystic tumor. Cystic degeneration is seen in 4% of leiomyomas [11].

Retroperitoneal location of leiomyomas are of extremely rare occurrence [2-5]. Pathologic

origin of these lesions are not certain. These retroperitoneal tumours are either metastatic or synchronous primary tumours. Tissue of origin is either smooth muscle elements or the embryonal remnants of müllerian or wolffian ducts ^[5,13].

In our case it was a secondary retroperitoneal leiomyoma. The huge degenerated leiomyoma had an uterine connection which suggests that it had been a secondary broad ligament fibroid to begin with, extending retroperitoneally later on. The cystic degeneration might have occurred after leiomyoma outgrew its blood supply. The retroperitoneum being a large potential space these cystic lesions can reach enormous size before becoming symptomatic ^[14].

Retroperitoneal cystic masses are rare ^[6,14]. These masses can be neoplastic like cystic lymphangioma, mucinous cystadenoma, cystic teratoma, cystic mesothelioma, müllerian cyst, epidermoid cyst, tailgut cyst, bronchogenic cyst, cystic change in solid neoplasms, pseudomyxoma retroperitonei and perianal mucinous carcinoma or they can be nonneoplastic like pancreatic

pseudocyst, nonpancreatic pseudocyst, lymphocele, urinoma and hematoma ^[6].

Mohammad ARAFA *et.al.* reported a case of extensive cystic degeneration in a pelvic leiomyoma mimicking ovarian cystic neoplasm but it was not connected with uterus ^[9]. Amr A.Soliman *et.al.* and Jifeng Feng *et.al.* have reported cases of huge degenerated and solid retroperitoneal leiomyomas respectively ^[3,4]. Uterine attachment was not found in any of the cases. So our case is one of the rarest of rare.

A case almost similar to ours has been described by Manoranjan Kar *et.al.*^[14]. In their case the presentation and gross appearance of the huge retroperitoneal cyst was almost similar but histopathology showed a benign multiloculated cystic lesion lined by flattened endothelium resembling cystichaemangioma. Immunohistochemistry was not available at their set up.

For diagnosis of uterine fibroids USG is the primary modality; however unusual locations and presentations may lead to a diagnostic dilemma ^[11]. CT and MR imaging are valuable tools for assessment of retroperitoneal masses describing their location, extent, internal characteristics and

involvement of adjacent structures [15]. Complete excision with or without abdominal hysterectomy is the main stay of treatment for these lesions [4,5].

CONCLUSION

Retroperitoneal leiomyomata is a rare entity. Huge retroperitoneal uterine leiomyoma with cystic degeneration is rarer. Degenerated uterine leiomyoma in unusual locations can pose diagnostic challenge preoperatively as well as intraoperatively. Thorough radiographic imaging is important in such cases. In these unusual cases histopathology and immunohistochemistry help to make final diagnosis. The treatment of retroperitoneal leiomyoma is complete excision with or without abdominal hysterectomy.

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