



BENIGN CYSTIC MESOTHELIOMA OF THE PERITONEUM MIMICKING AN OVARIAN CYST ISSUES IN DIAGNOSIS

VINODHA M

Department of Obstetrics and Gynaecology, ESI-PGIMS R

Abstract : Benign cystic peritoneal mesothelioma is a rare intra abdominal tumour with a strong predilection for peritoneum of pelvic organs. Incidence is approximately 1 in 10,00,000 cases¹. We present a case of benign cystic peritoneal mesothelioma which mimicked an ovarian cyst both clinically and radiologically. Excision of the cyst was done. Histopathology proved to be benign cystic mesothelioma of the peritoneum.

Keyword : Ovarian cyst, Benign cystic peritoneal mesothelioma.

INTRODUCTION:

Benign cystic peritoneal mesothelioma arises from the peritoneum that covers the serous cavities. It occurs most frequently in women of premenopausal age group with history of abdominal surgery, endometriosis or pelvic inflammatory disease. Due to the rarity of this tumor and similarity of its presentation with ovarian cyst, a preoperative diagnosis of ovarian cyst is usually made. This case is presented to emphasize the diagnostic challenges faced by a clinician, when evaluating a pelvic mass.

CASE REPORT:

A 46 yr old perimenopausal woman, para2 live2, sterilized, with one prior lower segment caesarean section presented with complaints of abdominal distension and lower abdominal dull aching pain for 3 months, dyspepsia for 1 month. No menstrual complaints. No symptoms suggestive of bladder or bowel disturbance. No history of loss of appetite or loss of weight. General examination was normal. Lower abdomen was distended. Palpation revealed a mass of 20 weeks size, the mass was cystic in nature, mobile with smooth surface, not tender, lower border of the mass was not made out, dull on percussion. No evidence of free fluid. On bimanual examination the same mass was palpated in the left lateral and posterior fornices, uterus felt separately from the mass and was normal in size. Per rectal examination revealed the same mass, rectal mucosa was free.

USG ABDOMEN AND PELVIS- revealed a left ovarian cyst of size 21×18×17cm, clear cyst, no septations or solid components. Uterus and right ovary were normal.

Endometrial thickness was 8mm. No free fluid. **CT SCAN ABDOMEN AND PELVIS(Plain and Contrast)-** revealed a large abdominopelvic mass of size 22×21×15cm seen in left side displacing the bowel loops. No solid components, septations or calcifications seen. Left ovary not separately visualized from the mass, probably left ovarian cyst. Right ovary was normal. Bilateral kidneys and ureters normal. No evidence of free fluid. CA-125 was within normal limits (8.8U/mL). CEA, AFP and beta hCG levels were normal.

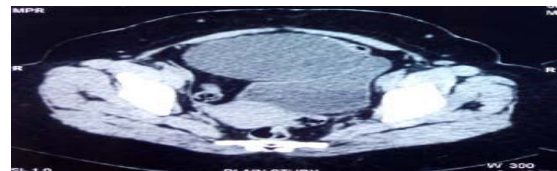


FIG-1 Plain CT showing the mass.

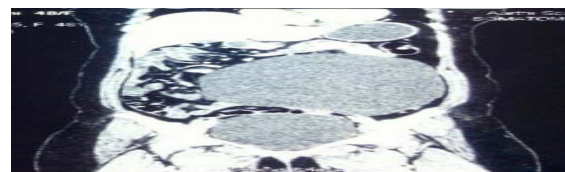


FIG-2 CECT- Coronal view of the cystic mass in the left side displacing the bowel loops to the right.

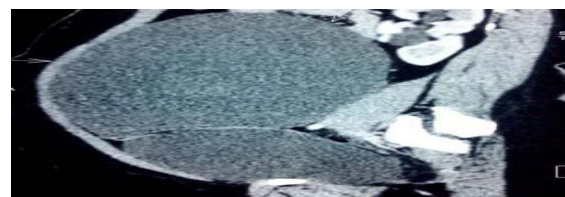


FIG-3 CECT- Sagittal view.

In view of provisional diagnosis of benign ovarian cyst, laparotomy was performed. Intraoperatively there was a huge clear cyst of size 20×20×18cm occupying whole of the pelvis and abdomen. The cyst was found to arise between the layers of sigmoid mesocolon. Uterus, both fallopian tubes and both ovaries were normal. Cyst decompressed and about 3.2 litres of clear fluid removed. Cyst wall was enucleated in toto. The post-operative period was uneventful. Histopathology of the cyst wall revealed **benign cystic mesothelioma of the peritoneum**. Patient was advised regular follow up in view of risk of recurrence.



FIG-4 Picture showing the cyst from the sigmoid mesocolon.

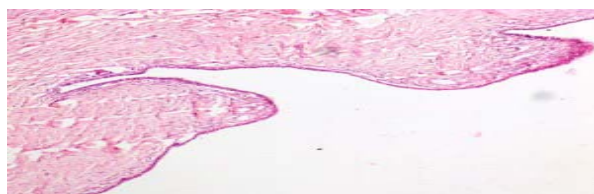


FIG-5 H&E Stain 100X : Fibrocollagenous wall with low cuboidal epithelium.

DISCUSSION:

Benign cystic peritoneal mesothelioma is a rare intra abdominal tumour with a strong predilection for peritoneum of pelvic organs. Incidence is approximately 1 in 10,00,000 cases. It was first described in 1979 by Menemeyer and Smith². It most commonly occurs in pre-menopausal women with a mean age of onset of 37 years.³ It is a localized tumor arising from the epithelial and mesenchymal elements of the mesothelial cells⁴. Most patients are diagnosed incidentally either on physical examination or as a finding on imaging, or during laparotomy for other indications.⁵ Differential diagnosis includes both benign and malignant lesions including cystic lymphangioma, endosalpingiosis, adenomatoid tumors, mesonephric duct remnants, malignant mesothelioma, sarcoma, and non-Hodgkin's lymphoma.^{3,5} Clinically, the differential diagnosis of benign cystic peritoneal mesothelioma from other ovarian tumors is important, since it can be treated with ovarian preservation techniques.⁵ The pathogenesis of this tumor is either reactive or neoplastic. The close relationship with inflammation, a history of prior surgery, endometriosis or uterine leiomyoma suggests that it is probably a peculiar peritoneal reaction to chronic irritation stimuli, with mesothelial cell entrapment, reactive proliferation and cystic formation.^{1,5} It could be neoplastic because of the slow progressive nature and marked tendency to recur after multiple surgical resections.¹ Malignant transformation of benign cystic peritoneal mesothelioma is very rare. However there is 30%-50% chance of recurrence. Therefore a lifelong regular follow up of these patients is required. The tumor is usually found on the surfaces of the pelvic viscera. In women it usually arises along the peritoneal surfaces of the uterus and rectum. Microscopic examination reveals cysts lined by a single layer of flattened or cuboidal mesothelial cells without atypia or mitosis.^{3,6} Focal reactive mesothelial changes such as hobnail-shaped cells and foci of mesothelial hyperplasia can be present.⁷ With immunohistochemical analysis, these cells stain positive for calretinin and cytokeratins, a reflection of their mesothelial origin.⁷

Treatment primarily involves surgery, as complete resection has been shown to be the only effective treatment.⁸ A case series by Sawh *et al*⁹ found that some stain positive for either estrogen or progesterone receptors, or both. Experimental treatment with anti-estrogen therapy (e.g., tamoxifen), GnRH agonists (e.g., leuprolide acetate), and intraperitoneal chemotherapy have been attempted with varying degrees of success.^{10,11} Diagnosis is difficult as there are no reliable clinical findings, features on imaging or tumor markers that are pathognomonic for this tumor.¹² While imaging abdomino pelvic mass, distinguishing ovarian from nonovarian masses is aided by determining the relationship of the mass to the anatomic pelvic structures, presence of ovarian parenchyma or ligament, and the effect of the mass on the ureter. CT imaging of the ovaries are based on ovarian vascular pedicle sign, embedded organ sign, phantom organ sign and beak sign. **Ovarian vascular pedicle sign** -the presence of asymmetrically enlarged ovarian vein in the pelvic mass indicates ovary as the organ of origin. **Embedded organ sign**-when part of the organ appears to be embedded in the tumour it is likely that the tumor originates from that organ. However to recognise this sign atleast some normal ovarian tissue should be present. **Phantom organ sign** when a large mass arises from a small organ, sometimes the organ becomes undetectable. The inability to detect ovary itself gives clue that the mass could be arising from the ovary, especially when the contralateral ovary is made out. **Beak sign** – when the tumor arises from the ovary, normal ovarian parenchyma extends and wraps around the tumor producing sharp angle between the ovarian parenchyma and the tumor. However all these signs may not be found in all cases especially when the tumor is large sized. Due to the existing limitations in imaging modalities, it becomes difficult to distinguish ovarian and non ovarian masses.

CONCLUSION:

Large pelvic masses in women may originate from the peritoneum, retro-peritoneum, ovaries, fallopian tubes, uterus, cervix, gastrointestinal tract or bladder. In spite of the advent of advanced diagnostic techniques, **distinguishing ovarian from non-ovarian masses, present a special diagnostic challenge**. A detailed history, thorough clinical examination, careful reading of radiological imaging is mandatory. We have to expect the unexpected and properly equipped to tackle the situation.

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