CASE REPORT

Peritoneal inclusion cysts in pregnancy, a diagnostic challenge

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SUMMARY

A cystic pelvic mass detected during pregnancy is not uncommon, but can be a diagnostic challenge. Most of these pelvic masses are benign ovarian cysts and resolve spontaneously. However, rare non-ovarian causes can complicate the diagnostic and therapeutic management. In this report an unusual case is presented of a 27-year-old pregnant woman with an atypical multicystic lesion in the pelvis, seen on routine first trimester ultrasound. A laparoscopic exploration was performed to rule out ovarian malignancy, and after histological analysis she was diagnosed with peritoneal inclusion cysts. The diagnostic and therapeutic challenges inherent to this rare non-ovarian disease are addressed in this case report.

BACKGROUND

Since the introduction of routine prenatal ultrasound, the detection of ovarian masses during pregnancy is not uncommon. The estimated incidence is reported between 1% and 10%.^{1–3} Most of these ovarian masses are functional and resolve spontaneously during the first or second trimester.^{3–5} The majority of these masses are benign cystic teratomas, cystadenomas, corpus luteum cysts and follicular cysts. Additionally, endometriomas, luteomas, haemorrhagic cysts, theca-lutein cysts, hyperstimulated ovaries, pseudocysts and leiomyomas are frequently seen in pregnancy.^{2–4 6 7} In general, the risk of a malignancy is low, estimated around 1–3% of all ovarian masses detected during pregnancy.^{1 4}

Ultrasound is a good diagnostic tool for determining the source of a cystic mass in pregnancy, and is usually sufficient to determine the management of ovarian masses. If ultrasound is inconclusive, MRI is preferred to CT due to its superior soft tissue resolution and the lack of ionising radiation.^{4 5}

Although most cystic masses detected during pregnancy are of ovarian origin, there are rare non-ovarian diagnoses that should be considered, especially when there is an atypical presentation on imaging.⁸ These diagnoses include omental cysts, mesenteric cysts, mucocoele of the appendix, paraovarian cysts and peritoneal inclusion cysts (PICs).^{8–10}

We present a case of PIC diagnosed during pregnancy and we will provide an overview of literature regarding diagnosis and treatment of this disease, complicated by pregnancy. There are only two cases of PIC in pregnancy previously described. In both cases the cystic mass was an incidental finding at caesarean section.¹¹

CASE PRESENTATION

A 27-year-old primigravid was referred to our outpatient clinic with multiple cysts posterior to the uterus seen on routine ultrasound during pregnancy at 14 weeks of gestation. She had no history of gynaecological disease, abdominal pain or abdominal surgery.

INVESTIGATIONS

Ultrasonography showed a multilocular abnormality of 9.5×5.5 cm in diameter, most suspect of a functional cyst of the ovary (figure 1). The cystic areas were homogeneous, and partially dense. The serum level of CA-125 was elevated up to 209 ku/L. Additional MRI of the pelvic region was performed to obtain more information about the exact localisation of the mass (figure 2). It showed a large multicystic lesion localised posterior, and at the left side of the uterus, presumably originating from the left ovary. The lesion was 10 cm in diameter, and consisted of multiple cysts varying in size, with thin septae. One of the cysts had higher signal intensity. There was no free fluid seen in the pelvis. A malignant cause seemed less probable.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of cystic masses is very extensive, and in this case complicated by atypical presentation on imaging. It concerned a persisting large mass, CA-125 was elevated and ovarian involvement was still uncertain. Therefore, the differential diagnosis of this pelvic cystic mass consisted of a benign, borderline or malignant tumour of the ovary, and non-ovarian causes such as omental cysts or PIC.

Figure 1 Transvaginal ultrasound showing multiple cysts posterior to the uterus, most suspect for functional ovarian cysts.

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Figure 2 Additional MRI showing a large multicystic lesion localised posterior, and at the left side of the uterus, presumably originating from the left ovary. The lesion was 10 cm in diameter, and consisted of multiple cysts varying in size, with thin septae.

TREATMENT

Owing to the fact that the differential diagnosis still included a malignancy, laparoscopic exploration of the abdomen was performed at 17 weeks of gestation, and revealed a multicystic lesion in the cul-de-sac, separately from the ovaries (figure 3). The multicystic lesion consisted of translucent and haemorrhagic cysts with a rigid wall. One separate cyst, located in the left paracolic gutter, was aspirated and sent for cytological analysis. Both ovaries and the appendix had a normal appearance. An origin of the masses could not be identified during the procedure. All cysts were drained, the visible cystic walls were partially removed, and multiple biopsies of the cystic wall were sent for histological analysis. There were no complications during the procedure, and the postoperative course was uneventful with good fetal condition. The results of cytological and histological analysis showed no evidence of a malignancy, and the cyst was diagnosed as PIC. Additional testing showed positive staining for calretinin and keratin, and also focal positive staining for estrogen receptor (ER) and progesterone receptor (PR), confirming the diagnosis.

OUTCOME AND FOLLOW-UP

After laparoscopy expectant management with frequent follow-up was performed. There were no further complications during pregnancy, and at 41 weeks of gestation a healthy daughter was born by vaginal delivery after induction. Third stage of delivery was complicated by a postpartum haemorrhage. At follow-up 6 weeks post partum there were no abdominal symptoms. Transvaginal ultrasound showed two normal ovaries, and a residual cystic lesion of 3.3 cm in diameter. Further follow-up will take place in 6–12 months.

DISCUSSION

PICs are rare, fluid filled lesions originating from the peritoneal mesothelium.^{9 12–14} They were thought to be neoplastic¹⁵ by some, but today they are mainly considered to be reactive

proliferations secondary to a peritoneal insult. PICs are therefore associated with a history of endometriosis, pelvic inflammatory disease or abdominal surgery.^{9 12 13} They predominantly occur in women at their reproductive age, and are most commonly localised in the pelvic peritoneum, particularly adhering to the reproductive organs, bladder, rectum and cul-de-sac.^{9 12 14–17} Most PICs are accidental findings, but they can be symptomatic and cause symptoms of abdominal distension and/or tenderness.^{6 12 13 16} On imaging, PICs appear as multilocular cystic lesions in the pelvic region. They pertain to the peritoneum, can have mass effect, are filled with fluid and can include haemorrhagic components. The classic appearance on ultrasound is a spider web or honeycomb pattern.^{11 17} However, PICs can present as very complex lesions on the ovarian surface, hence mimicking an ovarian malignancy.¹² MRI and CT can help define the anatomic relationships of the cystic mass with the surrounding organs.¹⁸ In pregnancy MRI is preferred over CT.^{4 5} The findings of a thickened wall, primary organ involvement, lymphadenopathy or ascites are in contradiction with the diagnosis of PIC.^{11 12 17} Owing to the fact that PICs are rare lesions and the difficulty of completely ruling out a malignancy on imaging, the diagnosis is almost never rendered preoperatively. Definitive diagnosis can be made based on cytological and histological examination after surgery.^{9 17}

Microscopic examination of the cystic walls shows lining by flattened or cuboidal mesothelial cells and fibrovascular tissue in the septae.^{14 15 17} The fluid derived from the cysts can be clear to yellow serous and can contain haemorrhagic components. Cytology can show non-specific reactive mesothelial cells and is usually non-diagnostic.¹⁷ Immunohistochemistry can be used for differentiation from other causes. PICs stain positive for calretinin and cytokeratins, and positive staining for ER/PR has been described in some cases.^{17 18}

In our case the diagnosis was complicated by pregnancy. There was no history of endometriosis, pelvic inflammatory disease or abdominal surgery. As described earlier, ultrasound imaging is usually sufficient to decide on the management of cystic lesions found during pregnancy.⁵ However, because of the size and complexity of the lesions, and the doubts about the origin further imaging was required. Normally, serum CA-125 levels are an important factor in the diagnostic process of a suspicious pelvic mass, and together with ultrasonic findings and menopausal status it is used to calculate the risk of malignancy index.^{3 17} In our patient serum CA-125 levels were elevated, but this is of limited value because it is also consistent with pregnancy. Especially in the early phase of pregnancy production of CA-125 by the decidua increases, therefore interpretation of serum levels in pregnant women is unreliable.^{3 4} In addition serum CA-125 can also be elevated due to benign ovarian lesions, such as functional ovarian cysts, endometriosis or other diseases that cause irritation of the peritoneum.¹⁹

Definitive diagnosis can be acquired through diagnostic laparoscopy, but safety regarding the pregnancy should be taken into account. Expectant management of non-suspicious lesions can be considered in pregnant patients because of the predominantly benign nature.^{1 6} However, it is not desirable to delay the diagnosis of a possible malignancy. Fortunately, ovarian cancer is rare during pregnancy, as it is in all women in their reproductive age.² Additional diagnostics through laparoscopy should be considered for suspicious lesions: for example, large masses, which persist after the first trimester or progress in size. After laparoscopy definitive diagnosis is more likely to be obtained through cytological and histological evaluation. This can be of great value in deciding on therapeutic management.⁴ When

Figure 3 (A–C) During laparoscopic exploration a multicystic lesion in the cul-de-sac, separately from the ovaries, consisting of translucent and haemorrhagic cysts was revealed.

performed in early second trimester, laparoscopy seems to be the safest and hardly increases the risk of pregnancy complications.^{3 4 6} The chance of pregnancy loss has decreased substantially compared with first trimester, and most functional cysts should have resolved by then.⁶

Most cysts found in pregnancy do not require surgical resection, especially in the first trimester, unless they cause pain or result in torsion, obstruction or rupture.^{4 6} However, emergency surgery because of such complications, especially in the third trimester of pregnancy, increases the risk of miscarriage, preterm birth and poor perinatal outcomes.^{2 4 7}

The recommended treatment for PIC consists of complete surgical resection of all visible cyst walls.^{11 17} Unfortunately recurrence after complete resection occurs in up to 50% of treated patients.¹⁶ In symptomatic patients with recurrence after multiple resections aggressive cytoreductive surgery can be considered as a last effort, as well as chemotherapy.^{11 17} In some cases the cystic wall contains ER/PR receptors, and hormonal management has been described as a successful way to decrease cyst size and symptoms, however long-term hormonal treatment is not desirable.¹⁷

Owing to the benign character of PIC, aggressive therapy is not always necessary, and observational management can be considered in asymptomatic patients. However, when there is any suspicion of a malignancy diagnostic surgery should be performed.^{12 17}

There is no consensus in the literature with respect to follow-up of patients treated for PIC, or asymptomatic patients.

In conclusion, this case presents a pregnant woman diagnosed with PIC. Non-ovarian causes such as PIC should be

considered in the differential diagnosis of cystic lesions found in pregnant women. Pregnancy complicates the diagnostic and therapeutic management of such cases, but MRI and, if necessary, diagnostic laparoscopy can safely be performed when there is any suspicion of a malignant cause or when benign cysts persist.

Learning points

- ▶ Peritoneal inclusion cysts (PICs) are a rare non-ovarian cause of cystic pelvic lesions.
- ▶ Rare non-ovarian causes such as PIC should be considered in the differential diagnosis of cystic lesions detected during pregnancy, especially when there is atypical presentation on imaging.
- ▶ When there is any suspicion of a malignant cause or when benign cysts persist, MRI and diagnostic laparoscopy can be safely performed during pregnancy, preferably in the early second trimester.
- ▶ Recommended treatment exists of surgical excision of all visible cyst walls. However, expectant management can be considered in asymptomatic patients.

Competing interests None.

Patient consent Obtained.

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