CASE REPORT

Management of pregnancy in woman with suspected malignant deep infiltrating endometriosis fistulised to the uterine cervix

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SUMMARY

Deep infiltrating endometriosis (DIE) is a well-known cause of pelvic pain and infertility. Malignant transformation of DIE is rare but can be suggested by MRI. We report a case of a spontaneous pregnancy in a woman with suspicion of malignant transformation of DIE with fistulisation to the posterior uterine isthmus through to the cervical canal. The pregnancy was closely monitored and an uneventful caesarian section was performed at 34 weeks of gestation. This case raises the issue of the relevance of imaging techniques and management of pregnancy.

BACKGROUND

Apart from endometriomas, malignant transformation of endometriosis is an uncommon event occurring in approximately 1% of cases.^{1 2} Association of malignant transformation of deep infiltrating endometriosis (DIE) with pregnancy is extremely rare and raises specific concerns about the progression of the lesion which could have lethal consequences, the impact on pregnancy outcome and the route of delivery.^{3 4} We report a case of a spontaneous pregnancy in a woman with DIE with suspicion of malignant transformation fistulised to the posterior uterine isthmus through to the cervical canal.

CASE PRESENTATION

A 32-year-old woman presenting with chronic pelvic pain and a 10-year primary infertility related to DIE was referred to our department. On physical examination, a bulky 4 cm nodule involving the torus uterinum, uterosacral ligaments and the vagina was found. Rectal digitation suggested a lesion infiltrating the rectum and the right parametrium including the pelvic wall. MRI revealed a nodule measuring 48×38 mm confirming an endometriotic lesion composed of a cystic component and atypical solid formation measuring 31×16 mm. The solid component exhibited an intermediate signal intensity on T2-weighted images, type 3 enhancement curve, high signal intensity on diffusion-weighted images obtained at b value of 1000 s/mm² and a restricted apparent diffusion coefficient (ADC). The mass was fistulised to the uterine isthmus through to the cervical canal encompassing the ureter (causing a right ureterohydronephrosis) and the right uterine artery ([figure 1](#)). CA125 serum level was of 260 UI/mL. These findings were suggestive of a malignant transformation

of DIE. The patient was advised to undergo surgical management of the lesion consisting of biopsy with intraoperative histology completed by radical surgery with hysterectomy, bilateral salpingo-oophorectomy, colorectal resection, omentectomy and pelvic and para-aortic lymphadenectomy in case of malignancy, but initially refused. She was regularly monitored with additional MRI showing the same features with a slight increase in size. She finally accepted surgery 8 months after the first visit to the department. The day before the operation, a systematic plasmatic β -human chorionic gonadotropin test resulted positive. The patient decided to conserve the pregnancy. At 15 weeks of gestation, she refused the laparoscopic examination initially planned for biopsies. On MRI without gadolinium injection performed at 17 weeks of gestation, the mass was found to have increased in size up to 54 mm with a decrease in the cystic component but an increase in the solid component associated with an increase of the uterine isthmus fistulisation. MRI performed at 26 weeks of gestation showed a mass measuring 66 mm with increased solid component ([figure 2](#)) and a haemorrhagic component ([figure 3](#)).

Owing to the fistula, suspicion of malignancy and the potential risk of spontaneous bleeding or uterine rupture, elective caesarean section was performed at 34 weeks of gestation after corticotherapy for fetal pulmonary maturation. A male child weighing 2070 g with a 10/10 Apgar score was delivered. Intraoperative abdominal examination showed no abdominal spread of the disease but a retrouterine necrotic mass involving the uterus, the colorectum and ovaries. Exploration of the pouch of Douglas was not possible so we were unable to evaluate the entire mass and the fistula. Multiple biopsies of the peritoneum and of the upper part of the mass were performed along with peritoneal washing. Histology confirmed the presence of endometriosis without features of malignancy.

OUTCOME AND FOLLOW-UP

The patient had three postdelivery examinations and refused surgical evaluation at the 6-month postdelivery visit.

DISCUSSION

This case reports the management of a pregnancy in a woman with DIE fistulised to the uterine cervix. This formation of fistula, detected on MRI, is particularly interesting as it has not been described in the literature to date. Spontaneous

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Figure 1 T2-weighted MRI sequence before pregnancy: initial cystic component and atypical solid formation with intermediate signal intensity. Uterine isthmus fistulae.

fistula formation due to endometriosis is extremely rare and almost always involves the uterine wall. We could only find one reported case of peritonitis secondary to a colouterine fistula with pelvic sepsis.⁵ Rectovaginal and urinary fistula mostly occur after surgical resection for endometriosis.^{6,7}

We were concerned that this fistula in the posterior wall of the isthmus, weakening this part of the uterus, could present a risk of uterine rupture and possibly of pelvic haemorrhage and sepsis, especially in the setting of pregnancy.

The management of the pregnancy was further complicated by the suspicion of malignancy. Sampson first documented the association of endometriosis with malignant tumours in 1925.² Approximately 1% of women with endometriosis develop an endometriosis-associated malignancy.⁸ Endometriosis-associated ovarian and extraovarian cancers represent a specific entity with distinct imaging features and management options described in the literature.⁴ On MRI, these cancers typically manifest as solid lesions with intermediate signal intensity on T1-weighted and T2-weighted images.⁴ These pelvic lesions characteristically enhance after the intravenous administration of gadolinium-based contrast material and demonstrate restricted diffusion on diffusion-weighted images and ADC maps.^{4,9,10} In our case, we suspected malignant transformation on the solid component of the mass with intermediate-signal intensity on T2-weighted images, type 2 enhancements and restricted ADC. Another criteria for malignancy was the high signal intensity within the

Figure 2 T2-weighted MRI sequence at 26 weeks of pregnancy: increasing pseudo-tumoural solid component.

Figure 3 T1-weighted MRI sequence at 26 weeks of pregnancy: haemorrhagic component of the mass.

solid component at b value of 1000 s/mm², already described in the literature.¹¹

The pregnancy was managed without histological features of the mass as the patient refused any invasive procedure. During the pregnancy, we observed modifications to the aspect of the mass on each MRI: increased tissular component with decreased haemorrhagic component and aspects of serosal invasion in the posterior wall of the uterus and cervix. It can be difficult to distinguish the enlargement of adnexal masses due to malignant transformation from those related to decidualisation linked to pregnancy.¹² Indeed, decidual changes of the endometrial tissue in adnexal masses during pregnancy can manifest as mural nodules and mimic malignant transformation.¹³ In this case, we observed these decidual characteristics on the first MRI with hyperintensity on diffusion-weighted images and restricted ADC, suggesting malignant transformation before the beginning of the pregnancy.¹⁴

Other risk factors also justified our active management at 34 weeks of gestation. First, the risk of spontaneous bleeding by rupture of the uterine vessel due to parametrial invasion of the mass. There was also a risk of acute haemoperitoneum as the mass encompassed the right uterine artery.¹⁵ Endometriosis is now recognised as the main risk factor for spontaneous haemoperitoneum in pregnancy. Strikingly, the site of the lesion is almost invariably the posterior side of the uterus or on the parametrium.¹⁵ Uterine rupture is, as previously described, a potential risk during pregnancy. These risks linked with the fistula and right parametrial invasion, though unquantifiable, kept us on full alert.

Although DIE is a known cause of infertility especially when the colorectum is involved, the present case report confirms the possibility of spontaneous pregnancy even in patients with extensive lesions. Adamson *et al*¹⁶ developed an Endometriosis Fertility Index (EFI) to evaluate the risk of spontaneous pregnancy. The EFI score for our patient was 2 evaluating the occurrence of a spontaneous pregnancy at 3 years at under 9.9%. Moreover, Younis *et al*¹⁷ reported that a duration of infertility over 10 years was a negative predictive factor of fertility even using in vitro fertilisation (IVF). Finally, in accordance with previous studies,¹⁸ the best option for patients with colorectal endometriosis to conceive is surgery and IVF. However, this is only an option once suspicion of a malignant lesion has been excluded which was not possible in this case as the patient refused any procedure to assess the nature of the retrouterine mass.¹⁹

Learning points

- ▶ Spontaneous pregnancy with suspicion of malignant transformation of deep infiltrating endometriosis and fistulisation to the uterus is a rare event.
- ▶ MRI is a precious tool to monitor specific signs but can also lead to misdiagnosis.
- ▶ Management of pregnancy in this setting has to take into account many risks such as spreading of malignancy, uterine rupture and haemorrhage.

Competing interests None.

Patient consent Obtained.

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