Case report

Incidental placenta increta at the time of prophylactic hysterectomy for Lynch syndrome: Insights into individualized decision-making and surgical timing

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1. Introduction

Lynch syndrome is the most common cause of hereditary endometrial cancer and the second most common cause of hereditary ovarian cancer (Crispens, 2012; Meyer et al., 2009; Lachiewicz et al., 2014). Prophylactic hysterectomy and bilateral salpingo-oophorectomy (BSO) should be discussed with patients as a risk-reducing option for endometrial and ovarian cancers at the completion of childbearing (Lancaster et al., 2015; Committee on Practice B-G, Society of Gynecologic O, 2014). However, the precise timing of prophylactic hysterectomy and BSO, including within the postpartum period is not well defined.

Here, we report a case of a woman with Lynch syndrome who underwent prophylactic hysterectomy and BSO approximately 15 weeks postpartum. Her course was notable for persistent postpartum bleeding and intraoperative hemorrhage at the time of hysterectomy. Final pathology revealed an incidental placenta increta and focal endometrial hyperplasia with atypia.
and the patient did well postoperatively. Final pathology revealed hyalinized and infarcted chorionic villi with dystrophic calcifications and extensive myometrial involvement of hyalinized tissue with areas of focal hemorrhage. These findings were consistent with placenta increta (Fig. 1A–C). There was also a focus of endometrial hyperplasia with atypia in proximity to the increta (Fig. 1D). Both ovaries and fallopian tubes were normal and pelvic washings were negative for malignancy.

2. Discussion

Lynch syndrome is an autosomal dominant, familial cancer risk syndrome, characterized by a germline mutation in DNA mismatch repair genes: MLH1, MSH2, MSH6, PMS1, and PMS2 (Crispens, 2012; Lachiewicz et al., 2014). The risk of endometrial cancer, a sentinel cancer for many women diagnosed with Lynch syndrome, is equivalent to or greater than the risk of colorectal cancer (Crispens, 2012). Since Lynch syndrome is associated with hereditary endometrial and ovarian cancers, gynecologic oncologists play a critical role in performing screening and risk-reducing prophylactic procedures. Consensus guidelines recommend that women with known or suspected Lynch syndrome undergo annual endometrial sampling, starting at ages 30–35 or when there is any change in bleeding patterns (Committee on Practice B-G, Society of Gynecologic Oncology, 2014; Burt et al., 2013). Routine pelvic ultrasound and CA125 for ovarian cancer surveillance are not endorsed by the National Comprehensive Cancer Network (NCCN) guidelines for the management of Lynch syndrome (Burt et al., 2013).

Prophylactic hysterectomy and BSO is an option for reducing the risks of both endometrial and ovarian cancers in women who have completed childbearing or in the early mid-40s (the mean age of endometrial (ages 47–49) and ovarian (ages 42–49) cancers is younger than the general population) (Committee on Practice B-G, Society of Gynecologic Oncology, 2014). Preoperative endometrial sampling and intraoperative evaluation of the uterine specimen are warranted since occult preinvasive lesions and endometrial, ovarian, and fallopian tube cancers have been detected at the time of prophylactic hysterectomy (Lachiewicz et al., 2014; Karamurzin et al., 2013). To our knowledge, the finding of placenta increta at prophylactic hysterectomy for Lynch syndrome has not been reported. This unexpected finding, in addition to the focal endometrial hyperplasia, likely contributed to the patient’s postpartum and intraoperative bleeding. Interestingly, an antenatal diagnosis of abnormal placentation was not made despite serial imaging and the diagnosis of hyperplasia was not made on preoperative curettage. These factors point to the importance of thorough pathological evaluation of the final specimens. The disrupted interface between the normal endometrium and myometrium could be related to Lynch syndrome, however, we did not find any literature to support this intriguing possibility. Although the precise explanation for the finding of placenta increta is not known, in this case risk factors included advanced maternal age and postpartum D&C for retained products (Publications Committee SFM-FM and Belfort, 2010).

For MSH6 mutation carriers, the cumulative risk by age 70 of endometrial cancer is approximately 60% and ovarian cancer is approximately 24% (Burt et al., 2013; Baglietto et al., 2010; Barrow et al., 2013). This patient was highly motivated to undergo genetics counseling and testing during her second and final planned pregnancy, based on her knowledge of her strong family history of Lynch-associated gynecological cancers in multiple female family members in their 40s and 50s. Once her test results returned as positive for a deleterious MSH6 mutation, she expressed a strong desire for risk-reducing surgery as soon as possible after delivery. The patient underwent extensive counseling about the risks, benefits, and alternatives of prophylactic surgery in the postpartum period. A shared decision was made to allow the uterus to involute and the patient to breastfeed for at least 3 months postpartum without discomfort from surgery.

As awareness about hereditary gynecologic cancer risks increases and genetics testing becomes increasingly routine, it is likely that prophylactic gynecologic surgeries will increase. The primary goal of prophylactic surgery is to prevent the development of cancer, while minimizing risks of complications and negative effects on quality of life. Counseling should include desires for future fertility, the medical management of menopause, and the timing of prophylactic surgery after completion of childbearing.
The possibility of discovering an occult cancer or other unanticipated pathology should also be stressed.

There is limited information in the literature about the decision-making process for prophylactic surgery women diagnosed with Lynch syndrome and what exists is qualitative (Etchegary et al., 2015). Guidelines that recommend prophylactic surgery do not specify precise timing after completion of child-bearing (Committee on Practice B-G, Society of Gynecologic Oncology, 2014). Here, we illustrate the importance of weighing the patient’s desires and concerns about her future risk of developing cancer and the immediate risks of unexpected findings and complications from surgery performed in the postpartum period. Hysterectomy proximate to delivery poses a higher risk of perioperative complications. The physiologic changes associated with pregnancy contribute to the risk of complications from decreased visibility due to the large size of the uterus, increased risk of hemorrhage from the large blood vessels and blood volume supplying the gravid uterus, and other factors related to cardiac output and coagulation (Cunningham et al., 2013). After the uterus involutes, minimally invasive options for hysterectomy (vaginal or laparoscopic), which are associated with better outcomes and fewer complications (Nieboer et al., 2009; ACOG Committee Opinion, 2009), become available. To maximize outcomes, we recommended a minimally invasive approach to be performed after 6 weeks post delivery. Although hysterectomy was performed 15 weeks postpartum in this case, the finding of an occult placenta increta demonstrates the potential risks of peripartum pathology and complications beyond 6 weeks.

3. Conclusion

Timing of prophylactic procedures in the postpartum period should be included in the shared decision making process for the management of Lynch syndrome and other hereditary cancers. It is important to assure extensive work up and counseling regarding symptoms and risks of detecting incidental findings that include peripartum pathologies and occult cancer.

Conflict of interest statement

The authors have no financial conflicts of interest to disclose.

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