11-1-2022

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Mariam Ayyash

Madison Miller

Miriana Hijaz

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Mariam Ayyash ,[1] Madison Miller,[1] Miriana Hijaz[2]

[1]Department of Women's Health, Obstetrics and Gynecology, Henry Ford Health System, Detroit, Michigan, USA
[2]Department of Women's Health, Division of Gynecologic Oncology, Henry Ford Health System, Detroit, Michigan, USA

Correspondence to
Dr Mariam Ayyash;
mayyash1@hfhs.org

Accepted 18 October 2022

To cite: Ayyash M, Miller M, Hijaz M. *BMJ Case Rep* 2022;**15**:e252017. doi:10.1136/bcr-2022-252017

SUMMARY

Microperforate hymens are rare anatomical variants with an unknown incidence and very few reported cases. Borderline ovarian tumours are similarly uncommon, with an incidence of approximately 0.002%–0.006%. The concurrent presence of a microperforate hymen and a borderline ovarian tumour is therefore exceedingly unique with no documented cases to date. In this report, we review the case of a nulliparous woman in her late 20s who initially presented with an inability to have penetrative intercourse. A subocclusive hymenal variant was noted on examination and further imaging work-up resulted in the incidental discovery of a large ovarian mass subsequently noted to be a borderline ovarian tumour. Herein, we review contemporary approaches to the diagnosis and management of both hymenal variants and borderline ovarian tumours, and discuss fertility-sparing strategies for young women diagnosed with ovarian neoplasms.

BACKGROUND

Microperforate hymens are exceptionally rare, partially occlusive hymenal variants with an unknown incidence rate.[1–3] Although there is a paucity of literature available on microperforate hymens, documented cases were typically diagnosed in adolescent patients seeking evaluation for symptoms such as primary amenorrhoea, atypical menstrual bleeding (characterised by filiform or watery-appearing menses), prolonged spotting, bleeding 'from only one side of the vagina', cyclic abdominal pain, difficulty with tampon insertion, recurrent vulvovaginitis, and changes in urinary and bowel habits.[1 4 5] When a microperforate hymen is suspected, imaging with translabial or transperineal ultrasound can help to differentiate these variants from other anomalies, such as transverse vaginal septa, Mayer-Rokitansky-Küster-Hauser syndrome and vaginal agenesis.[6]

As subocclusive hymenal variants can lead to the partial or complete obstruction of menstrual efflux, it is not uncommon for patients to develop haematocolpos, haematometrocolpos or haematosalpinx.[7 8] In fact, all reported cases of pelvic masses in patients with known hymenal variants were found to be benign accumulations of blood.[9] To our knowledge, this is the first report of an ovarian neoplasm—ultimately identified as a borderline serous ovarian tumour—discovered in a patient undergoing evaluation of symptoms related to a microperforate hymen.

CASE PRESENTATION

A nulliparous woman in her late 20s presented to an outpatient gynaecologist with an inability to have penetrative vaginal intercourse. She reported coitarche 2 months prior to presentation. The patient and her partner attempted but never achieved penetrative intercourse on two occasions; both times, intercourse was suspended due to severe dyspareunia. She experienced sexual arousal and endorsed a desire to have penetrative intercourse on both occasions. She reported regular menstrual cycles and denied dysmenorrhoea, menorrhagia or intermenstrual spotting. She attempted tampon insertion once a few years prior to presentation, but was unable to do so secondary to pain. Her gynaecological, medical, surgical, familial and social histories were otherwise unremarkable. The abdominal examination revealed no masses or distention. On pelvic examination, an intact vaginal hymen was noted with two small perforations located just beneath the urethra. Neither a digital nor speculum examination could be performed.

INVESTIGATIONS

Translabial ultrasound was ordered to better typify the patient's hymenal variant and screen for any additional genital anomalies. The ultrasound revealed a large, vascularised presacral mass of uncertain origin extending into the right adnexa (figure 1). MRI was obtained for further characterisation that redemonstrated the 11.7×9.1×8.3 cm pelvic mass likely arising from the right ovary/adnexal region with features favouring ovarian neoplasm (figure 2).

Gynaecological oncology was consulted and recommended CT scan of the chest, abdomen and pelvis, as well as tumour markers. Tumour markers were significant for an elevated cancer antigen 19-9 of 257.1 and cancer antigen 125 of 4233 (table 1). A CT scan showed a non-enlarged uterus with a large, heterogeneous-appearing pelvic mass 11.7×9.7 cm in size. No additional masses or enlarged lymph nodes were visualised (figure 3).

DIFFERENTIAL DIAGNOSIS

Following the completion of these diagnostic studies, the patient met with a gynaecological oncologist and was counselled that the differential diagnosis of her pelvic mass included both benign and malignant aetiologies.

TREATMENT

Given the patient's strong desire for future fertility, a plan was made to proceed with a stepwise, diagnostic approach starting with laparoscopy with intra-abdominal/ peritoneal biopsies and concurrent hymenectomy. Pending pathology results,

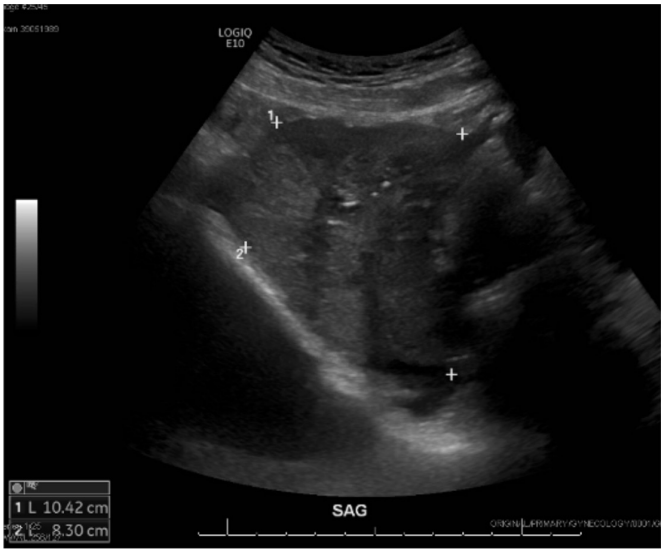

Figure 1 Translabial ultrasound image demonstrating a large pelvic mass, measuring 10.4×8.9×9.3 cm.

final recommendation for surveillance versus definitive surgical management would be offered. Two weeks later, the patient underwent the above-listed procedures. Examination under anaesthesia was consistent with a microperforate hymen, and an uncomplicated hymenectomy was performed (figure 4). Upon abdominal entry with the laparoscope, a large mass seemingly originating from the right ovary with fatty-appearing papillary-like projections that extended to the posterior cul-de-sac was observed (figure 5). The uterus, left ovary, left fallopian tube, omentum, bowel, liver and abdominal wall appeared grossly normal. The papillary-like projections extending from the ovarian lesion were biopsied using bipolar energy, and care was taken to ensure that the mass remained intact. Omental biopsies and some of the free fluid noted in the pelvis were also collected. All surgical specimens were sent to pathology for evaluation. Although frozen pathology was initially concerning for 'adenocarcinoma', the final pathology report suggested 'at least serous borderline tumour, invasive disease cannot be excluded'.

Given the patient's young age and strong desire for future fertility, she was counselled on the option of either radical or conservative fertility-sparing surgical management. Following extensive discussion about the risks and benefits of both approaches, she elected for the fertility-sparing strategy and underwent consultation with both reproductive endocrinology and infertility and colorectal surgery specialists. She was counselled on, but ultimately declined, embryo cryopreservation. Five weeks after her initial diagnostic laparoscopy and hymenectomy,

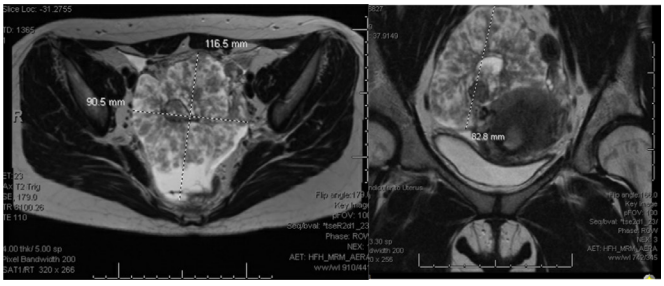

Figure 2 MRI demonstrating an 11.7×9.1×8.3 cm pelvic mass, assumed to be arising from the right ovary/adnexa.

Table 1 The patient's tumour marker values and reference ranges

Tumour marker	Value	Normal reference range
Inhibin A (pg/mL)	13	<98
Inhibin B (pg/mL)	14	<153
CA 19-9 (U/mL)	257.1*	<45.1
CA 125 (U/mL)	4233*	<49
CEA (ng/mL)	0.7	<6.1
bHCG (mIU/mL)	<10	<10
LDH (IU/L)	180	<250
AFP (ng/mL)	1.8	<8.1

*abnormal
AFP, alpha-fetoprotein; bHCG, beta-human chorionic gonadotropin; CA, cancer antigen; CEA, carcinoembryonic antigen; LDH, lactate dehydrogenase.

she was boarded for exploratory laparotomy. The gross appearance of the mass was stable from prior (figure 6) and the lesion was noted to be adhered to the posterolateral aspect of the uterus but not fixed within the pelvic cavity. Uncomplicated right salpingo-oophorectomy was performed, resulting in complete resection of the pelvic mass, as well as peritoneal biopsies, infragastric omentectomy and right pelvic lymph node dissection. Frozen pathology was consistent with 'at least borderline tumour', which prompted our staging biopsies and pelvic lymph node biopsies. Final pathology demonstrated 'serous borderline tumour of the ovary with negative peritoneal and lymph node biopsies'.

OUTCOME AND FOLLOW-UP

Given her final diagnosis of stage 1C3 serous borderline tumour of the ovary, recommendation for surveillance was made and the patient was scheduled for a 6-month follow-up with gynaecological oncology.

DISCUSSION

The presented case represents the confluence of two particularly rare gynaecological anomalies: (1) a subocclusive hymenal variant and (2) an incidentally discovered borderline ovarian tumour (BOT).

The vaginal hymen is formed by the invagination of the posterior wall of the urogenital sinus and typically ruptures during the perinatal period, resulting in a thin mucous membrane remnant within the vaginal orifice.[10] Incomplete dissolution of the hymen can result in a number of hymenal variants, including microperforate hymens. Unlike patients with completely imperforate hymens who classically present during puberty with cyclic abdominal pain and dark-coloured or blue-tinged vaginal bulges, patients with subocclusive hymenal variants may have normal or partial menstrual efflux.[11] Therefore, the initial diagnosis of these variants may occur later in adolescence or even adulthood

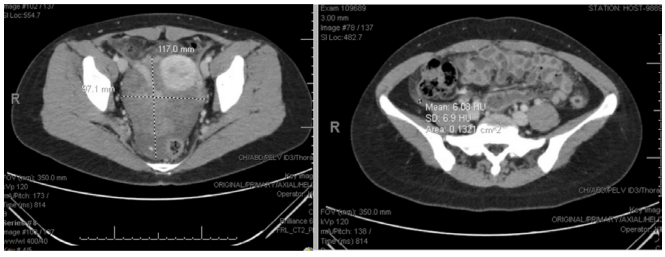

Figure 3 CT imaging showing a non-enlarged uterus with a large, heterogeneous-appearing pelvic mass.

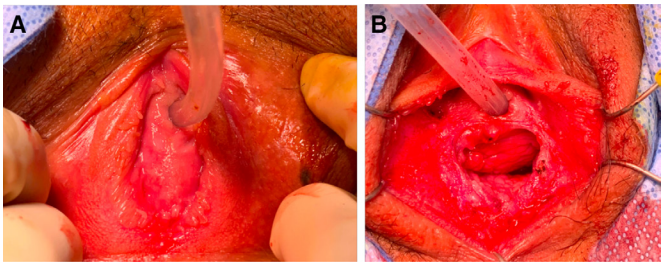

Figure 4 Microperforate hymen with two small microperforations located just inferior to the urethra (A). Vaginal introitus status after uncomplicated hymenectomy (B).

when patients present because they have difficulty placing or removing tampons or are unable to have penetrative vaginal intercourse.[1 11] When a hymenal variant is suspected, the physical examination plays a critical role in characterising the specific variant. Furthermore, imaging via translabial/transperineal ultrasound or MRI may aid in the differentiation of imperforate or subocclusive hymens from other anatomical anomalies such as labial adhesions, transverse or longitudinal vaginal septa, distal vaginal atresia, cervical atresia and obstructed uterine horns.[11]

Ultimately, surgery is required for the definitive management of an imperforate or subocclusive hymenal variant. Hymenectomy allows for complete efflux of menses and reduces patients' risk for the accumulation of menstrual fluid and subsequent distension of the vagina, uterus or fallopian tubes. Hymenectomy may also resolve other symptoms associated with hymenal variants, including recurrent vulvovaginitis, dysmenorrhoea, urinary retention, dysuria, constipation and dyschezia. Patients should be counselled that those with isolated hymenal variants who undergo hymenectomy are not at increased risk of infertility, long-term sexual dysfunction or adverse obstetic outcomes. It is recommended that hymenectomy be carried out with an initial cruciate incision using either sharp or cautery dissection with care taken to avoid damage to the urethra. Following excision of the redundant hymenal tissue, the mucosal edges should be re-approximated using a 3-0 or 4-0 absorbable suture in an interrupted fashion, and the vagina should be copiously irrigated.[11] Patients typically do not require dilator use, pelvic floor physical therapy or topical oestrogen after hymenectomy; however, these interventions may be considered if there is postoperative concern for adhesions or stenosis.

Approximately 15% of all primary ovarian neoplasms are considered BOTs, possessing features that are intermediate to those of benign and overtly malignant ovarian neoplasms of similar cell types.[12] Ovarian cancer encompasses multiple types of cancers, including epithelial and non-epithelial cancers, and small cell carcinomas and carcinosarcomas, which are histologically composed of both an epithelial and a sarcomatous

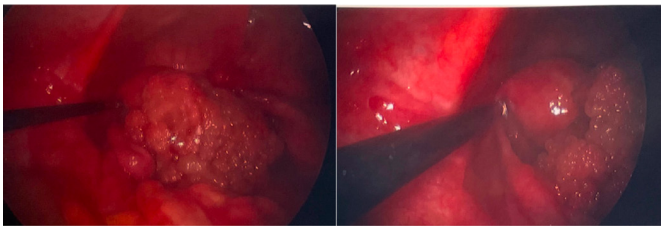

Figure 5 Diagnostic laparoscopy revealed pelvic mass seemingly originating from the right ovary with fatty-appearing papillary-like projections, fixed to the posterior cul-de-sac.

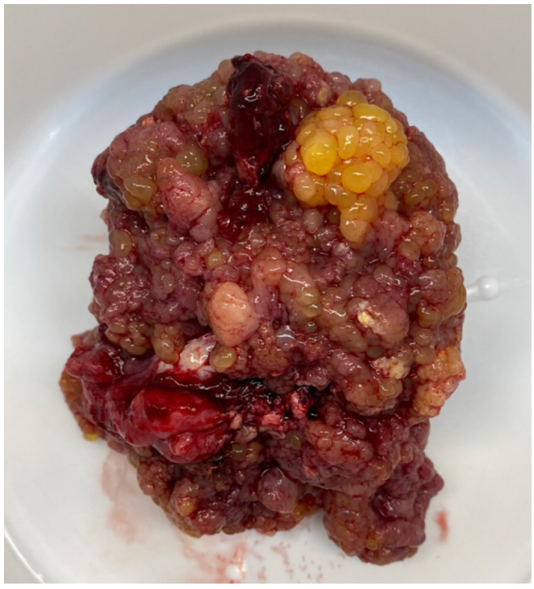

Figure 6 Complete excision of right fallopian tube, ovary and associated mass.

component.[13] Serous BOTs are the most common and best understood histological subtype, accounting for approximately 65% of all BOTs.[14] The vast majority of serous BOTs are stage I at the time of diagnosis, contributing to the overall excellent prognosis of these tumours with 5-year and 10-year survival rates of 99% and 97%, respectively.[14–16] However, it has been estimated that the 10-year survival rate for even advanced stage (stage II or higher) BOTs is between 70% and 90%.[17]

As approximately one-third of all BOTs are diagnosed in patients younger than 40 years, the decision to proceed with a complete staging procedure versus a more conservative surgical approach may be significantly influenced by a patient's desire for future fertility.[18] The fertility-sparing approach to the management of apparent unilateral serous BOTs involves either unilateral salpingo-oophorectomy or 'cystectomy'. Performance of pelvic washings, omental biopsy and biopsy of any visible peritoneal lesions is recommended with either procedure.[19] Fertility-sparing surgery with or without platinum-based chemotherapy remains the standard of care of non-epithelial ovarian cancers, providing a high chance of cure at all stages.[20] The majority of young women who are treated with that strategy may expect recovery of ovarian function, usually within a few months after treatment.[21] Fertility seems to be only marginally affected by treatment, with many reports of successful pregnancies and over 85% have been shown to regain menstrual function once chemotherapy was completed.[22 23] The efficacy of unilateral salpingo-oophorectomy versus cystectomy is highly debated, and there are mixed data on the rate of recurrence and subsequent fertility with one approach over the other. A systematic review of more than 100 relevant publications determined that the pooled estimate for spontaneous pregnancy rate following conservative surgical management (either unilateral salpingo-oophorectomy or cystectomy) of an early stage BOT was approximately 54%. For this same pooled cohort, the risk of lethal recurrence was low at an estimated 0.5%.[24] Patients who have undergone fertility-sparing surgery with borderline disease confirmed by final pathology are recommended to follow up with gynaecological oncology every 3–6 months with physical/pelvic examination and cancer antigen 125 (if initially elevated) at every visit for 5 years. Thereafter, annual visits are recommended with imaging as clinically

indicated. For patients who have elected for a fertility-sparing approach, the National Comprehensive Cancer Network presently recommends consideration of completion surgery—with removal of the contralateral fallopian tube and ovary and/or uterus—upon the conclusion of childbearing.[25]

Learning points

- A broad differential is always critical to consider in a woman with suspected hymenal variant.
- When a hymenal variant is suspected, the physical examination plays a critical role in characterising the specific variant and follow-up imaging is significant to aid in the differentiation of imperforate or subocclusive hymens from other more involved anatomical anomalies.
- Young patients with ovarian mass would benefit from a stepwise, diagnostic approach to allow for fertility preservation opportunities during the diagnostic and treatment process.

Acknowledgements The authors thank Stephanie Stebens, at Sladen Library, Henry Ford Hospital, for her input in reviewing this manuscript.

Contributors MA, the corresponding author, cared for the patient and identified the case for publication. MA and MM drafted the case report. MH cared for the patient and provided revisions to the article for approval for publication.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

ORCID iD

Mariam Ayyash http://orcid.org/0000-0002-0313-7438

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