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CASE REPORT

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A borderline mucinous ovarian tumour treated by ovarian cystectomy in a 16-year-old patient

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Abstract We present a case of a mucinous borderline ovarian tumour in a 16-year-old patient. It was removed by ovarian cystectomy before histological confirmation of its borderline nature.

Keywords Borderline · Ovarian tumour · Cystectomy · Mucinous tumour · Low malignant potential

Case report

A 16-year-old student presented to the casualty department in the early hours of the morning complaining of lower abdominal pain and vaginal bleeding in her first pregnancy. Her last menstrual period had been 3 months earlier, and a urine pregnancy test was positive. She gave a history of slight painless bleeding the previous day. This had become gradually heavier, with passing of large clots and increasing abdominal pain. She had had no shoulder-tip pain or fainting episodes. There was no previous surgical or medical history of note, and she was not on any medication apart from folic acid.

By the time she arrived in the emergency department, her bleeding had started to settle, and the pain was only mild. On vaginal examination, her uterus was tender, compatible with a gestation of around 10 weeks, and the cervical os was closed. The patient was haemodynamically stable with no active vaginal bleeding.

A provisional diagnosis of threatened miscarriage was made, and the patient was allowed to go home. An appointment to attend the early pregnancy assessment unit the following day for a transabdominal ultrasound scan was arranged. This scan showed an empty uterus with a clear endometrial echo. In the right adnexa, there was a complex multiloculated mass measuring $16.4 \times 11.5 \times 6.6$ cm^3.

A laparotomy was performed. A large right ovarian cyst was found. Both fallopian tubes and the left ovary were inspected and found to be normal. There was no evidence of an ectopic pregnancy, and the rest of the pelvis was otherwise normal. Therefore, a diagnosis of likely complete miscarriage and coexistent right ovarian cyst was made.

Thirty millilitres of peritoneal fluid was obtained and sent for cytological analysis. An ovarian cystectomy was performed. The cyst was removed intact and sent for histological processing. The remainder of the ovary was reconstructed. The patient recovered well and was sent home 5 days later. The following week, her pregnancy test had become negative, which confirmed she had had a complete miscarriage.

The histology report described a thin-walled multilocular ovarian cyst weighing 250 g and filled with mucoid material. There were no solid areas, and the capsule was intact. Tissue had been processed in 10 blocks. Microscopically, the cyst was lined by tall and uniform columnar epithelial mucinous cells. Occasional areas of slight nuclear hyperchromasia and frequent mitoses were seen. However, there was one area of rather complex thickened epithelium with mild to moderate nuclear pleomorphism and frequent mitoses. There were also buddings of the same atypical epithelium towards the adjacent stroma. There was no significant stromal reaction around the latter glands, thus confirming the absence of any stromal invasion. These features gave the cyst the appearance of a borderline mucinous ovarian tumour.

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After consultation with a tertiary referral centre for gynaecological oncology, a decision to manage this case conservatively was reached. The patient will be followed up in the outpatient clinic at six-monthly intervals for at least 5 years. These visits will involve a physical examination, a pelvic ultrasound scan, and measurement of serum Ca125 levels.

Discussion

Borderline ovarian tumours, described in 1929 by Taylor [1], consist of a subgroup of ovarian tumours characterised by the presence of branching papillae, epithelial stratification, nuclear atypia, mitotic activity, and the absence of stromal invasion in the primary tumour [2]. These ovarian tumours comprise 7.5–15% of all epithelial ovarian neoplasms. It is important to separate them from the invasive tumours because of their superior prognosis. The overall 5-year survival rate for patients with borderline tumours is 86.2%, compared with 46.1% in patients with invasive disease [2]. Survival for patients with stage I borderline neoplasms is 99%, while survival for advanced-stage disease without invasive implants is 92% [3]. Frank malignant transformation appears to occur in less than 1% of borderline tumours [3]. This rate is similar to the rate quoted for the malignant transformation of fibroids (0.2–0.7%), and the latter are not even considered tumours with low malignant potential (LMP). Therefore, conservative treatment of these lesions may be justified.

Compared with invasive epithelial ovarian cancers, borderline tumours occur in younger women, present at an earlier stage, and have a favourable prognosis. For historic reasons, these tumours are still often confused with carcinomas, resulting in mismanagement of many patients and the saying that ''there is no borderline tumour, just borderline management.''

Much of the morbidity and mortality associated with borderline tumours is directly associated with the treatment rather than with the disease itself [4]. The standard surgical treatment of borderline ovarian tumours has followed the guidelines for invasive carcinoma, including total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, and peritoneal washings [2]. In the case of borderline mucinous tumours, particularly bilateral cases, appendicectomy is recommended [5]. More conservative procedures preserving fertility are now accepted in younger patients.

The LMP category of ovarian tumours is one of the most controversial topics in gynaecological oncology and pathology and is confusing to both clinicians and patients. This category of ovarian tumours, also referred to as borderline tumours or, most recently, ''atypical proliferative tumours,'' was introduced in 1971 by the International Federation of Gynaecology and Obstetrics (FIGO) [6]. These tumours are recognised to generally behave in a benign fashion, occasionally following an indolent malignant course.

The borderline category of ovarian tumours has played an important role in the evolution of our understanding of ovarian carcinomas by segregating a group of tumours that, in contrast to typical ovarian carcinomas, do not invade the ovarian stroma and therefore are considered to be noninvasive. More importantly, these noninvasive tumours have a superior prognosis when compared with ovarian carcinomas stage for stage. Unfortunately, the category has also led to a clinical dilemma because many patients with borderline tumours are young and wish to preserve their fertility. Because borderline tumours are regarded as a subset of carcinomas, their treatment has often been more aggressive than is necessary.

Clearly, in a 16-year-old young woman, there is always a desire to preserve fertility as well as ovarian steroidogenesis. Therefore, an ovarian cystectomy was performed in our patient. A diagnosis of mucinous borderline ovarian tumour is uncommon in patients of this age. The presence of a large (> 10 cm) cystic ovarian mass in the absence of solid elements would point towards benign ovarian pathology. Tumour markers (α-FP, βHCG, Ca125) are raised in pregnancy and were therefore unhelpful in this case. Healy et al. found serum anti-inhibin to be commonly elevated with mucinous borderline ovarian tumours [7]. In these circumstances and in the presence of abdominal symptoms, a laparotomy and ovarian cystectomy would seem an adequate choice.

Although an adnexectomy is a reasonable option in the presence of a large ovarian cyst, several factors need to be considered. In an emergency situation and without a histological diagnosis, careful consideration of whether to perform a cystectomy or an adnexectomy needs to be exercised. In general, the patient's age is a very important factor. Ovarian cysts are common in women of reproductive age. If surgically possible, a cystectomy and reconstruction of the remaining ovary would be preferable in the presence of a large ovarian cyst in a young woman. Although women may conserve their fertility and steroidogenesis potential with only one ovary remaining, it is also true that the risk of losing the other ovary later on in life needs to be considered. Why remove an ovary if a cystectomy will suffice? Once histological confirmation of an LMP tumour is reached, there is no evidence to suggest that a second operation to complete an adnexectomy has any long-term survival advantage over long-term follow-up. There is also some evidence that ovarian cystectomy for stage 1 LMP tumours probably does not affect prognosis [8, 9]. If the woman had completed her family, then perhaps a total abdominal hysterectomy and bilateral salpingo-oophorectomy would be preferable because this, with optimal staging and debulking, is the recommended treatment for patients with LMP ovarian tumours who have completed childbearing [10].

Exploration of the abdominal cavity excluded the presence of an ectopic pregnancy. Cytological analysis of peritoneal fluid was negative, there were no macro-

scopic suspicious deposits, and the contralateral ovary appeared normal. These findings, along with the histological diagnosis of a borderline mucinous ovarian tumour with an intact capsule, allowed grading the tumour as stage IA. In view of this unusual diagnosis, an additional independent review of the pathologic specimen was sought, as recommended by the National Institutes of Health Consensus Statement [10]. This review confirmed the initial diagnosis.

Mucinous borderline stage I tumours have a survival rate close to 100%. In contrast, stages II and III mucinous borderline tumours have a poor survival but are nearly always associated with the syndrome of pseudomyxoma peritonei [5]. It is now clear that pseudomyxoma peritonei is a condition of appendiceal origin in virtually all cases. Therefore, apparent advanced-stage ovarian mucinous borderline tumours with pseudomyxoma peritonei represent secondary ovarian involvement from a gastrointestinal neoplasm and should not be classified as ovarian. In addition, there is a small group of mucinous carcinomas typically from the pancreas and biliary system that present with relatively bland-appearing metastases to the ovaries that closely simulate mucinous borderline tumours. Once these metastatic carcinomas and mucinous tumours associated with pseudomyxoma peritonei are removed from the borderline category, the remaining mucinous borderline tumours are always confined to the ovaries and have a benign behaviour.

Although formal staging was not performed in this case, the therapeutic benefit of reexploration is of questionable value as there was no evidence of gross residual disease at the time of the initial surgery [11]. A retrospective study by Querleu et al. suggests that whenever restaging of borderline tumours is to be considered in an individual patient, laparoscopy provides a suitable alternative approach to laparotomy [2]. Although more research is needed in this area, a laparoscopy rather than a laparotomy would seem an attractive alternative if operative restaging were to be considered.

Long-term follow-up is also a controversial issue. Traditionally, clinical history, pelvic examination, Ca125 levels, and transvaginal ultrasound scanning at regular intervals for 5 years has been the norm. However, there is no evidence available yet that this approach can be effectively used to reduce mortality from ovarian cancer or that it will result in decreased rather than increased morbidity and mortality [7].

In a 16-year-old woman with a diagnosis of mucinous borderline ovarian tumour, conservative treatment is indicated because of the likely benign nature of the disease. This allows preservation of fertility and future ovarian steroidogenesis. There may be an argument for appendicectomy to exclude primary appendiceal mucinous adenocarcinoma with secondary ovarian involvement. However, this is more common when there is bilateral ovarian disease. Long-term follow-up is controversial, although some authorities recommend that it be undertaken in an attempt to identify cases of invasive change as early as possible. Ultrasound scans and Ca125 monitoring seem to be a sensible approach but may not translate into long-term benefit.

References

1. Taylor HC Jr (1929) Malignant and semi malignant tumours of the ovary. Surg Gynecol Obstet 48:204–230
2. Querleu D, Papageorgiou TH, Lambaudie E, Sonoda Y, Narducci F, LeBlanc E (2003) Laparoscopic restaging of borderline ovarian tumours: results of 30 cases initially presumed as stage IA borderline ovarian tumours. Br J Obstet Gynaecol 110:201–204
3. Kurman RJ, Trimble CL (1993) The behaviour of serous tumours of low malignant potential: are they ever malignant? Int J Gynecol Pathol 12:120–127
4. Silva EG, Kurman RJ, Russell P, Scully RE (1996) Symposium: ovarian tumours of borderline malignancy. Int J Gynecol Pathol 15:281–302
5. Riopel MA, Ronnett BM, Kurman RJ (1999) Evaluation of diagnostic criteria and behaviour of ovarian intestinal-type mucinous tumours: atypical proliferative (borderline) tumours and intraepithelial, micro-invasive, invasive, and metastatic carcinomas. Am J Surg Pathol 23:617–635
6. International Federation of Gynaecology and Obstetrics (1971) Classification and staging of malignant tumours in the female pelvis. Acta Obstet Gynecol Scand 50:1–7
7. Healy DL, Burger HG, Mamers P et al (1993) Elevated serum inhibin concentrations in postmenopausal women with ovarian tumours. N Engl J Med 329:1539–1542
8. Lin-Tam S, Cajigas HE, Scully RE (1988) Ovarian cystectomy for serous borderline tumours: a follow-up study of 35 cases. Obstet Gynecol 72:775–80
9. Tazelaar HD, Bostwick DG, Ballon SC, Hendrickson MR, Kempson RL (1985) Conservative treatment of borderline ovarian tumours. Obstet Gynecol 66:417–22
10. National Institutes of Health Consensus Development Conference Statement (1994) Ovarian cancer: screening, treatment and follow-up. Natl Inst Health 12:10–12
11. Ehlen TG (2000) Management of low malignant potential tumour of the ovary. J Soc Obstet Gynaecol Can 22(1):19–21