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Case report

A case report of mucinous cystadenoma with contralateral serous cystadenofibroma in identical twin

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ABSTRACT

Introduction: There are limited reports on ovarian neoplasm occurring among identical twins. Most previous reports showed ovarian teratoma found in both twins. Herein, we report for the first time a case of ovarian mucinous cystadenoma with contralateral serous cystadenofibroma found in twin siblings.

Case report: One patient suffered from abdominal distension and the following computed tomography found ovarian mucinous cystadenoma. During the laparoscopy, another ovarian mass was found in the contralateral ovary. The histopathology revealed ovarian mucinous cystadenoma with contralateral serous cystadenofibroma. The twin sister had no symptoms but underwent gynecological screening. She also showed a similar finding, mucinous cystadenoma with serous cystadenofibroma on the contralateral ovary. Both patients underwent laparoscopic bilateral ovarian cystectomy.

Conclusion: This is the first clinical report on left ovarian mucinous cystadenoma with right serous cystadenofibroma in twin siblings. Our cases support awareness of ovarian tumors in twin sisters.

1. Introduction

The incidence of identical adnexal tumor found in twin sisters is rare. The most common adnexal tumor in reproductive age women is mature cystic teratoma, and accordingly, there were only cases of mature cystic teratoma reports in twin sisters [1–3]. To the best of our knowledge, there has been no report of mucinous cystadenoma and serous cystadenofibroma occurring simultaneously in twin. Both tumors are not common with mucinous ovarian cystadenoma accounting for approximately 10% of ovarian benign neoplasm while the incidence of ovarian cystadenofibroma is only 1.7% [4]. Despite the low incidence, surgical treatment is required for mucinous tumor for its malignant potential. For ovarian cystadenofibroma, it is important to differentiate from malignant neoplasm as it presents as a multicystic mass with solid components. Pre-operative evaluation including pelvic magnetic resonance imaging (MRI) is used to differentiate from malignant tumor. For early diagnosis, it may be important to consider the possibility of mucinous cystadenoma concurrently appearing with serous cystadenofibroma in the identical twin. We herein report the first case of identical twin with left ovarian mucinous cystadenoma with right serous cystadenofibroma who were treated with laparoscopic surgery.

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2. Case presentation

2.1. Case 1)

A 28-year-old woman, gravida 0 para 0, visited our gynecologic outpatient department for abdomen distension for about one month. Her history was noncontributory and she had no remarkable family history. Physical examination revealed a distended abdomen and a palpable mass in the whole abdomen. A pelvic examination revealed no significant findings. A transabdominal ultrasound showed a huge pelvic cyst measuring 28 cm × 19 cm with multiple septations. An abdomen computed tomography of the pelvic revealed a left ovarian tumor (29.2 cm × 21.2 cm x 10cm) (Fig. 1). The level of cancer antigen (CA-125) was 37.4 U/ml and 17.4 U/ml for CA 19-9 (normal range <35 U/mL). The preoperative level of Anti-Müllerian hormone (AMH) was 8.68 ng/ml. She underwent laparoscopic left salpingo-oophorectomy. During the laparoscopy, besides the mucinous cyst on the left ovary (Fig. 2A), a 2-cm-sized mass was found on the right ovary (Fig. 2B). Right ovarian cystectomy was also performed without surgical complication. Microscopic and histopathological examinations revealed mucinous cystadenoma for the left ovary (Fig. 3L) and serous cystadenofibroma for the right ovary (Fig. 3R). After the surgery, she was discharged three days following the operation. Regular pelvic examination using ultrasonography was performed and there was no sign of recurrence. The level of AMH decreased from 8.68 ng/ml before the operation to 3.19 ng/ml at 1 year after the surgery. However, the AMH was recovered to preoperative level at a 2-year follow-up measuring 9.27 ng/ml. The level of CA-125 was measured at 11.1 U/ml after the surgery.

2.2. Case 2)

A 28-year-old woman, gravida 0 para 0, who was the monozygotic twin sister of the patient in Case 1, visited our gynecological department for ovarian screening 1 month after the surgery of Case 1. Transvaginal sonography showed an 8cm sized multiseptated cyst on the left ovary and a 4cm sized hypoechogenic cyst on the right ovary. Serum levels of CA-125 and CA 19-9 were 18.28 U/ml and 29.60 U/ml respectively. The pelvic magnetic resonance imaging (MRI) showed a 4.7cm sized unilocular cyst with 5 mm sized solid nodule suspicious of cystadenofibroma (Fig. 4A) and an 8cm sized multilocular cyst with multiple internal septations in the left ovary with internal mural nodules (Fig. 4B). During the laparoscopy, besides the mucinous cyst on the left ovary (Fig. 5A), a small sized mass was found on the right ovary (Fig. 5B). And Laparoscopic bilateral ovarian cystectomy was performed without surgical complication. Microscopic and histopathological examinations revealed mucinous cystadenoma for the left ovary (Fig. 6L) and serous cystadenofibroma for the right ovary (Fig. 6R). The level of AMH was 23.55 ng/ml before the operation to 10.43 ng/ml 1 year after the surgery.

3. Discussion

The incidence of adnexal tumors in twin sisters is rare. The most common adnexal tumor in reproductive age women is mature cystic teratoma and accordingly, there were only cases of mature cystic teratoma reported in twin sisters, including monozygotic twins [1–3]. The genetic basis of mucinous cystadenoma and serous cystadenoma remains unknown while environmental factors could potentially cause the concurrent neoplasm. Shared environmental factors can include parental factors, such as socioeconomic status, lifestyle, and experiences shared by siblings during childhood and adolescences, and screening in adult life. The effects of

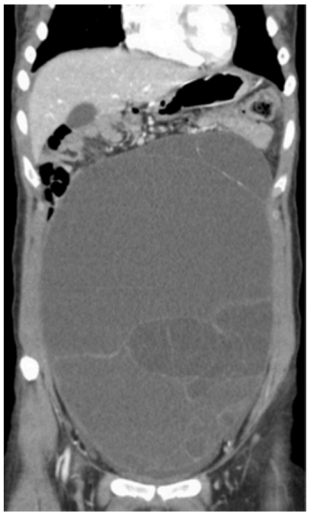

Fig. 1. CT finding for case 1. About 28 cm sized multilocular cyst with multiple internal septations in left ovary
CT: Computed-tomography.

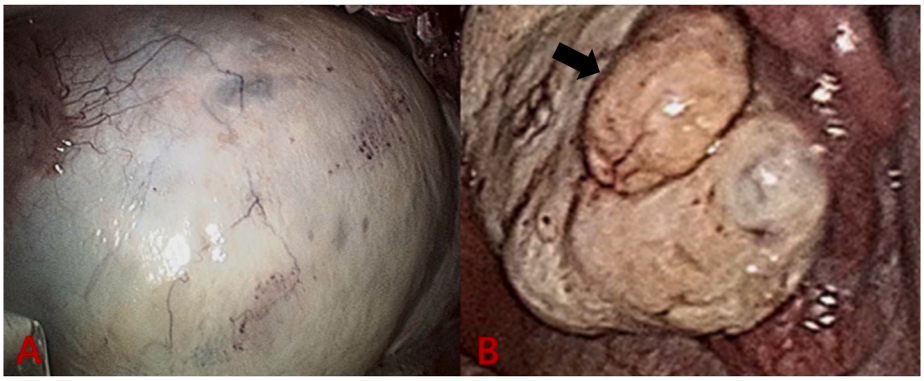

Fig. 2. Operation findings for case 1. A: left ovary, B: right ovary.

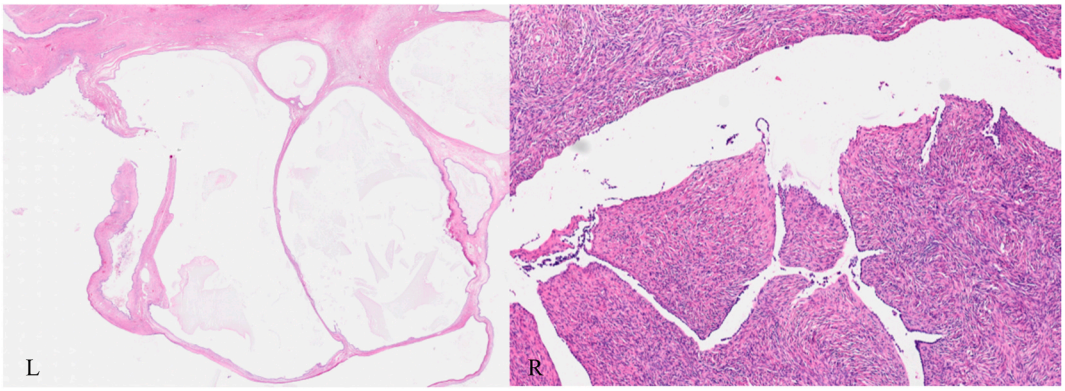

Fig. 3. Histology (H&E stain, 200x) for case 1. L: left ovary - Multilocular cystic neoplasm lined by a single layer of mucin producing epithelium. Mucin material is noted in the cystic lumen. R: right ovary - Part of cyst wall and fibrotic nodules in the cyst. Stroma of nodules and walls are composed of fibroblasts. Lining or covering epithelium is flat.

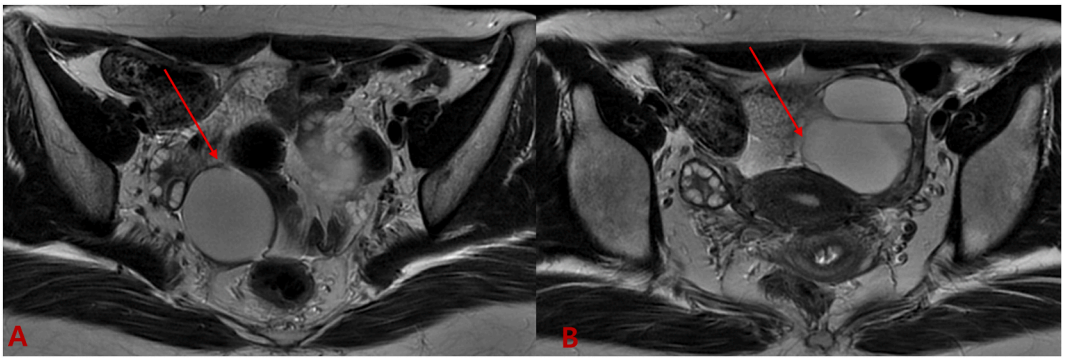

Fig. 4. MRI finding for case 2. 8cm sized multilocular cyst with multiple internal septation and mural nodule in the left ovary, 4.7 cm sized unilocular cyst in the right adnexa. MRI: Magnetic resonance imaging, L: left, R: right.

environmental factors on the development of various disease have been evaluated extensively in many studies. However, the evidence collectively suggest that both genetic and environmental factors may be involved in the development of tumors [5,6]. Moreover, according to a recent prospective study of twins regarding cancer, there was no evidence of shared environmental association for the development of many cancer in twins [7]. From the previous study, the effect of environmental factors may play a role in the development of adnexal tumor in twins, and yet the effects may be modulated by the genetic factors shared by the twins. The twin sisters in our study shared the similar environmental factors including parental factors. The living environment may have influenced the concurrent disease and yet the direct effect is difficult to measure.

Our case is unique and it is the first case to report the co-presence of mucinous cystadenoma with contralateral serous

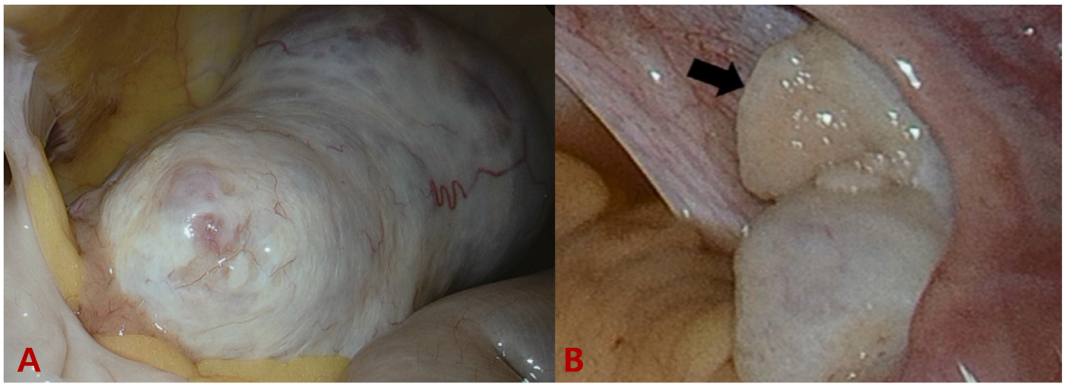

Fig. 5. Operation findings for case 2. A: left ovary, B: right ovary.

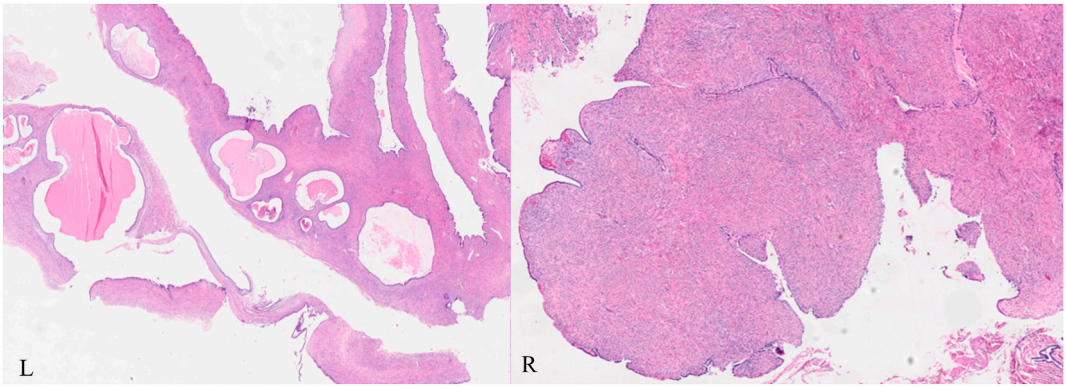

Fig. 6. Histology (H&E stain, 200x) for case 2. L: left ovary - Multilocular cystic neoplasm lined by a single layer of mucin producing epithelium. R: right ovary - Protruding fibrotic nodules into cyst. Lining or covering epithelium is flat. Slit single lining cysts are entrapped in fibrotic nodules.

cystadenofibroma in twin sisters. According to a prospective study of 200,000 twin individuals, a cumulative incidence of all cancer was 32% with 38% of monozygotic twins diagnosed with same cancer type. For most cancer types, the cumulative risks were higher in monozygotic than dizygotic twins. The heritability of ovarian neoplasm was reported to be 39% [7]. Mucinous tumors represent a spectrum of malignant behavior and have benign, borderline, and invasive histologic variants. Among benign ovarian neoplasm, mucinous cystadenomas account for approximately 10–15% of all cases [1,2].

Non-gynecological mucinous tumor has been previously reported in a twin study. Appendiceal mucinous adenoma was found in identical twins one of which developed pseudomyxoma peritonei with a possible link to *KRAS* mutation. The different types of mutations in *KRAS* and the different allelic status of the adenomatous polyposis coli (APC) locus in the tumors from both twins suggest that mutation in *KRAS* and loss of heterozygosity of APC occurs somatically in adenomas and is independent of the identical genetic background of the twins [8].

There was another case report of an intraductal papillary mucinous tumor of the pancreas in twin brothers. In this case, there was also *KRAS* gene mutation, which was suggested to be related to the development of papillary mucinous pancreatic tumor [9]. *KRAS* mutation is also suggested to be early even in mucinous ovarian tumorigenesis [10] Although, the examination of gene mutation was not evaluated in our case, this presence of mucinous adnexal tumor in twin sisters may suggest that genetic mutation such as *KRAS* mutation may be an important event to develop a mucinous adnexal tumor in twin siblings.

There were previously no case reports of ovarian serous cystadenofibroma in twins. This is the first case report of an ovarian mucinous tumor with contralateral serous cystadenofibroma in twin sisters. Although ovarian serous cystadenoma is a common benign tumor, ovarian cystadenofibroma is relatively rare. The actual incidence of ovarian cystadenofibroma is unknown. Cho et al. estimated that these tumors account for approximately 1.7% of all benign ovarian neoplasm [4]. Primary ovarian cystadenofibroma is encountered in women aged between 15 and 65 years [11]. Ovarian cystadenofibroma originates in the epithelium and is composed of cystic and solid fibrotic tissues, which are classified according to the epithelial cell types present, as serous, endometrioid, mucinous, clear cell, and mixed categories [12]. Ovarian cystadenofibroma presents as a multicystic mass with solid components, preoperative differential diagnosis is important to distinguish it from malignant neoplasm.

Given the rarity of ovarian cystadenofibroma, its incidence may be difficult to detect. Therefore, ovarian cystadenofibroma is one of many tumors subject to differential diagnosis in which preoperative diagnosis by MRI is useful. In addition, frozen sections may be

helpful in the intraoperative assessment of ovarian masses to guide appropriate surgical management. In our case, pre-operative MRI evaluation revealed a 4 cm-sized unilocular cyst with a solid portion suggestive of cystadenofibroma in preoperative MRI evaluation. In another patient, however, the mucinous cyst was approximately 30 cm in size which limited the evaluation of the contralateral ovary. In this patient, a solid mass was found in the laparoscopy, which was sent for frozen section evaluation showing cystadenofibroma.

In conclusion, it is the first case to report the presence of mucinous cystadenoma with cystadenofibroma in twin siblings. This case suggests that when evaluating mucinous tumor or cystadenofibroma, it is important to evaluate the twin sibling for the possible presence of the same disease.

Ethical statement

This retrospective study was approved by the Institution Review Board of Kyungpook national university hospital, and the requirement for informed consent from the patients was waived by the IRB.

Author contribution statement

All authors listed have significantly contributed to the investigation, development and writing of this article.

Data availability statement

Data included in article/supplementary material/referenced in article.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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