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Mucinous Cystadenoma Coexisting with Stromal Tumor with Minor Sex-Cord Elements of the Ovary
: A Case Report

Mucinous neoplasms occur rarely in association with cystic teratoma, Sertoli-Leydig cell tumor, granulosa cell tumor or carcinoid tumor. Several cases of an ovarian stromal tumor with minor sex-cord elements have been reported in the literatures. However, there has been no report about an ovarian mucinous neoplasm coexisting with a stromal tumor with sex-cord elements yet. We report a case of an ovarian neoplasm composed of both mucinous cystadenoma and stromal tumor with minor sex-cord elements in a 58-yr-old female. The ovary including the mass measured 5 cm in size. On section, it revealed an unilocular cyst (4.5 cm in diameter) filled with mucinous fluid. There was a round, yellow, solid nodule, 1.5 cm in diameter within the wall. Microscopically, the cyst was lined by a single layer of endocervical mucinous epithelium and the nodule was composed of spindle cells showing an intersecting and whorled arrangement. There were cell nests showing polygonal shape with abundant cytoplasm among the spindle cells. They showed immunoreactivity for inhibin and did not have any connection with the adjacent mucinous epithelium. Therefore, we interpret the mucinous cystadenoma as having arisen de novo.

Key Words: *Ovary; Cystadenoma; Adenocarcinoma, Mucinous; Sex Cord-Stromal Tumor*

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INTRODUCTION

Ovarian stromal tumor with minor sex cord elements is a rare neoplasm, first described by Young and Scully as a predominantly thecomatous or fibromatous tumor containing scattered sex-cord elements less than 5% of the tumor on each slide examined (1). The sex-cord elements are various in shape, and may show well-differentiated granulosa cells, totally indiscriminate cells, or solid tubular structures like immature testicular tubules. There have been several cases of ovarian stromal tumor with minor sex-cord elements reported in the literatures (1-4). Mucinous neoplasms occur rarely in association with cystic teratoma, Sertoli-Leydig cell tumor, granulosa cell tumor and carcinoid tumor (5). To our knowledge, a mucinous neoplasm coexisting with an ovarian stromal tumor with minor sex cord elements has not been reported in the literature until now. So we report the first case of mucinous cystadenoma coexisting with an ovarian stromal tumor with minor sex-cord elements.

CASE REPORT

Clinical history

A 58-yr-old woman was transferred to our hospital for proper management of an ovarian mass. Six yr ago she was found to have the left ovarian cyst of 4.0 cm in size on transvaginal ultrasonogram during a routine health examination (Fig. 1). She had been relatively well until the present admission without significant change of the ovarian mass on regular transvaginal ultrasonogram. The patient's menarche had occurred at the age of 16 and her menses were regular. She did not have a history of endocrine abnormalities, such as precocity, virilization, or an abnormal serum level of sex hormones. Under the clinical impression of a benign ovarian cystic neoplasm, she received the left oophorectomy. The operation findings were normal except for the ovarian mass.

Gross finding

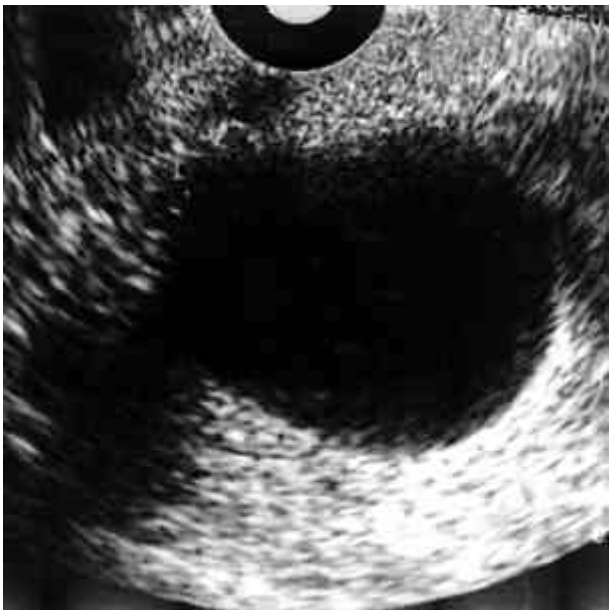

Fig. 1. Transvaginal ultrasonogram reveals a hypoechoic cystic mass of the left ovary, measuring 4.0 cm in size.

Grossly, the ovary including the mass measured 5 cm in maximal dimension and weighed 67 g. The external surface of the ovary was smooth. On section, it revealed an unilocular cyst (4.5 cm) with a leakage of mucinous fluid. There was a round, pale yellow to white solid nodule (1.5 cm) with a slightly whorled pattern in the wall without connection into the cyst (Fig. 2).

Microscopic finding

Microscopic examination revealed that the cyst was lined by a single row of uniform mucin-filled tall columnar cells with basal nuclei, resembling endocervical epithelium, the findings of which were consistent with a mucinous cystadenoma (Fig. 3A). The mucinous epithelium abutted onto the adjacent solid area was mainly composed of spindle cells (Fig. 3B), and there was no evidence of transition between them. A high-power view revealed that the solid tumor was composed of two elements. The majority consisted of spindle cells showing an intersecting fascicular and whorled arrangement with deposition of collagen tissue (Fig. 3B). The minority was composed of scattered clusters of relatively uniform large cells arranged as round nests and cords amidst a spindle cell background located at the junction between the above-mentioned spindle cell area and mucinous cystadenoma. The cells had angulated to round nuclei, indistinct nucleoli, and relatively abundant clear cytoplasm (Fig. 3C).

Immunohistochemical finding

The spindle cells showed immunoreactivity for inhibin, α-smooth muscle actin, vimentin and S-100 protein. The large uniform cells in cords or nests were strongly immunoreactive for inhibin and S-100 protein, but negative for α-smooth muscle actin, vimentin and placental alkaline phosphatase (Fig. 4).

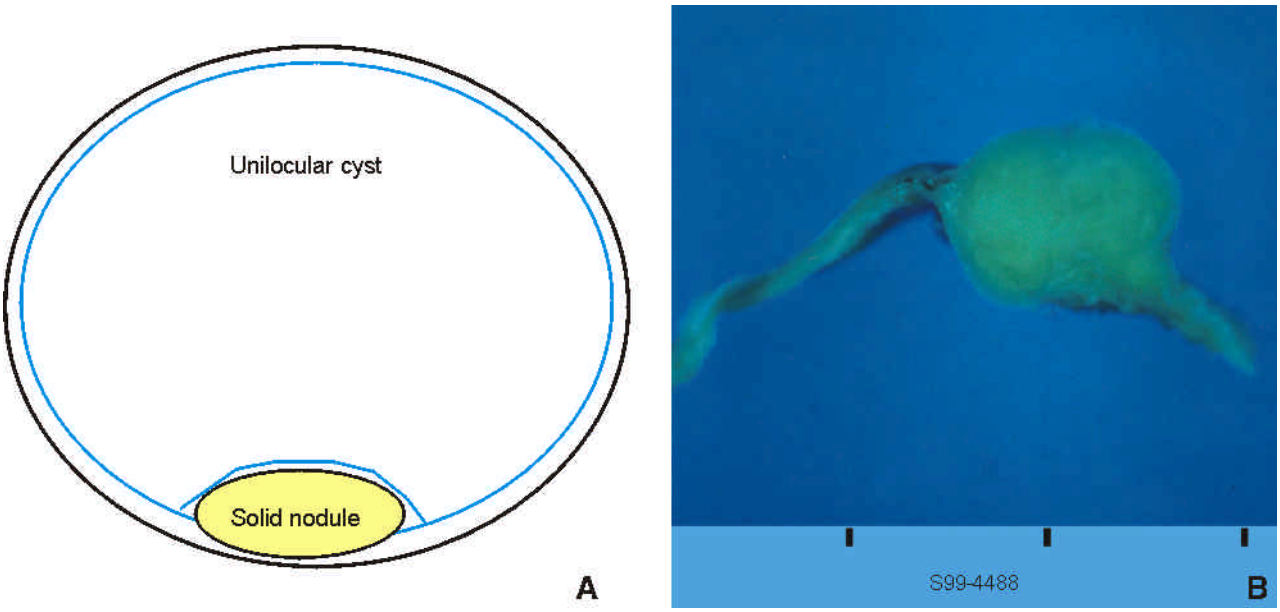

Fig. 2. Schematic view of the cut surface of the left ovary shows a unilocular cyst with a round solid mass (A). The cut surface of solid nodule reveals a pale yellow and slightly whorled pattern (B).

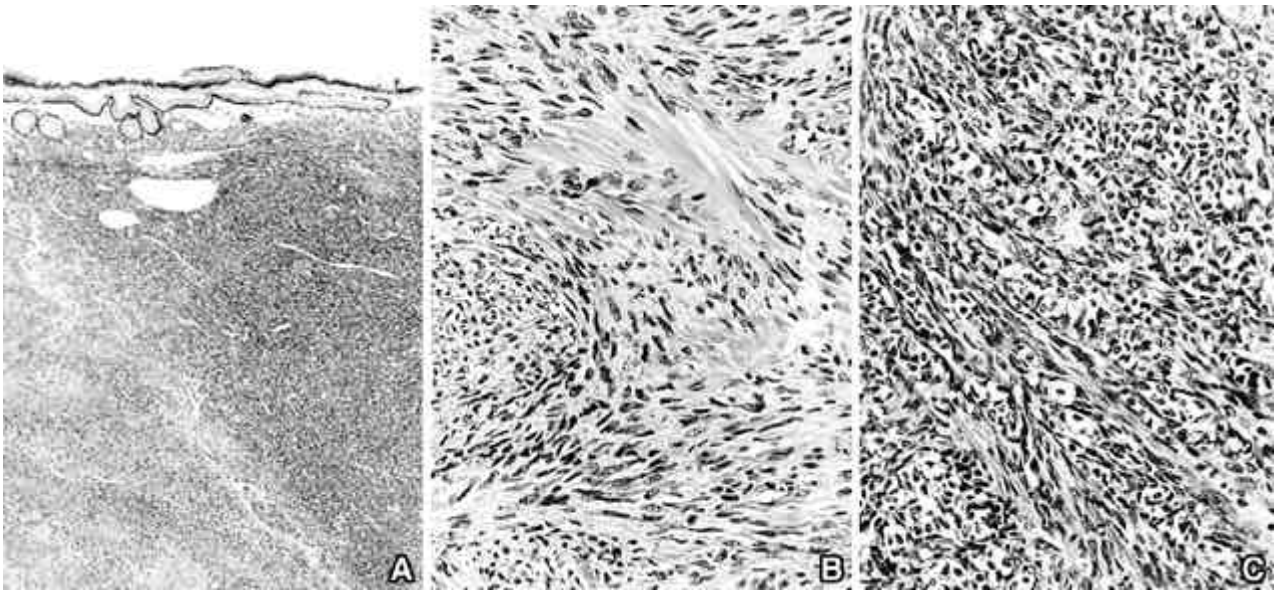

Fig. 3. The cyst is lined by a single row of uniform, endocervical type, mucin-filled columnar cells with basal nuclei; the finding of which is consistent with mucinous cystadenoma (A, H&E, ×40). The solid tumor is composed of fascicles of spindle cells (B, H&E, ×200) and scattered nests or cords of large polygonal cells. The large cells have angulated to round nuclei, indistinct nucleoli, and a clear cytoplasm (C, H&E, ×200).

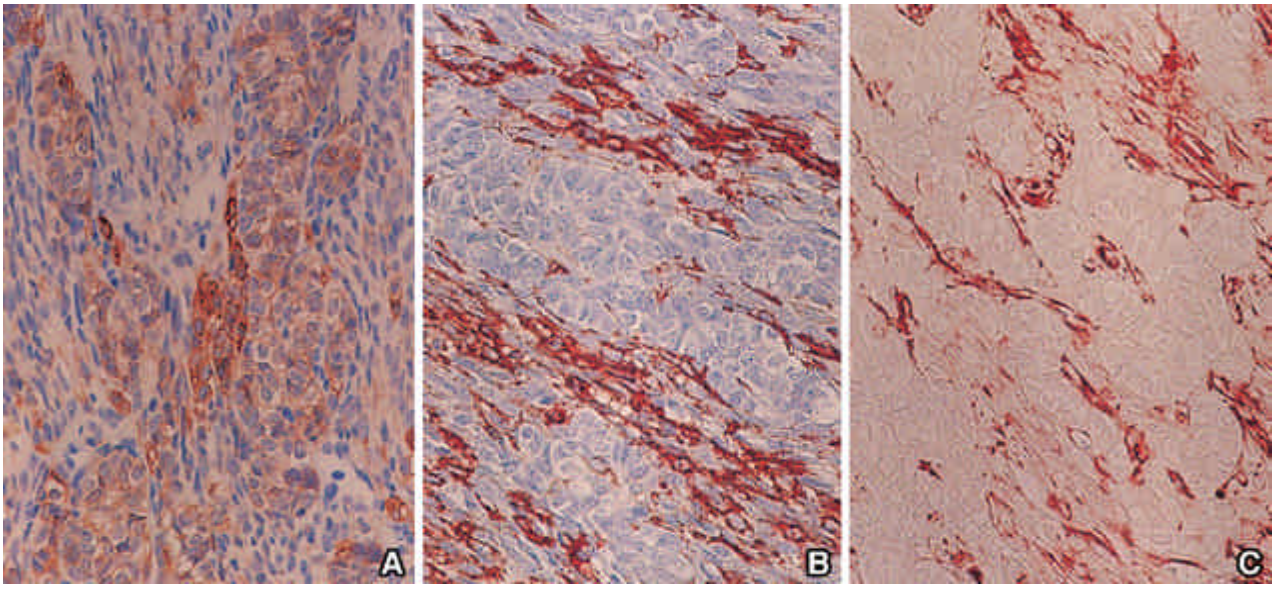

Fig. 4. Large uniform cells show strong immunoreactivity for inhibin (A), but are negative for α-smooth muscle actin (B) and vimentin (C) (×400).

DISCUSSION

Ovarian sex cord-stromal tumors are generally classified as three types: granulosa-stromal cell group (ovarian cell types), Sertoli-Leydig cell group (testicular cell types), and unclassified cell group, with regard to the differentiation of sex-cord elements toward the ovarian or testicular line (6). The stromal component is usually fibromatous or thecomatous. Our case showed an ovarian stromal tumor with scattered large uniform cells in cords or nests and a coexisting mucinous cystadenoma. These large uniform cells were initially considered as either sex cord elements or germ cells. But the possibility of a germ cell origin could be ruled out as these cells did not have prominent nucleoli and were negative for placental alkaline phosphatase and positive for inhibin and S-100

protein (7). Therefore, we thought the possiblity of a sex-cord origin for these cells. They were reminiscent of Sertoli cells as they showed a tubular or cordal arrangement in a background of spindle cell stroma, and they were also reminiscent of granulosa cells as they were immunoreactive for inhibin and S-100 protein. But the characteristic histologic and cytologic features of granulosa cells, such as the Call-Exner body or nuclear groove were absent. We concluded these large cells to be unclassified sex cordal elements owing to a lack of evidence of differentiation along testicular or ovarian lines. In addition, because the total area of these indifferent sex-cord elements was less than 5% of the tumor, our case most likely corresponds to ovarian stromal tumor containing minor sex-cord elements as described by Young and Scully (1).

In terms of mucinous component in this tumor, the diseases which would be most likely confused with this case are Sertoli-Leydig cell tumor with heterologous elements, gonadal teratoma with predominant mucinous elements (8) and mucinous cystadenofibroma. Heterologous elements occur in about 20 percent of Sertoli-Leydig cell tumors. These Sertoli-Leydig cell tumors with heterologous elements show mostly intermediate differentiation, characterized by immature Sertoli cells arranged in islands or in cords. In addition, the lining mucinous epithelium of glands or cysts of these tumors is usually of the gastric-type or intestinal-type, unlike our case which was of the endocervical type. The possibility of a gonadal teratoma with predominant mucinous elements could be easily ruled out by the finding of foci of sex-cord tumor components. A gonadal teratoma never contains neoplastic sex-cord elements by definition. Because our case showed a characteristic sex-cord element and did not have any connection between the stromal tumor and adjacent mucinous cystadenoma, it is reasonable to interpret the mucinous cystadenoma as having arisen de novo.

In summary, our case may be the first report of a mucinous cystadenoma coexisting with an ovarian stromal tumor with minor sex-cord elements of indiscriminate type in both Korean and English literatures.

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