

CASE REPORT

Serous Papillary Cystadenofibroma of Ovary with Extremely Elevated CA 125—A Masquerader of Malignancy: A Case Report

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ABSTRACT

Background: Benign serous papillary cystadenofibroma (SPCAF) of the ovary is a very rare subtype of serous tumors of the ovary. Commonly occurs between 15 and 65 years of age. They are usually asymptomatic or diagnosed as an incidental finding. Routine imaging features masquerade malignancy and lead to extensive surgeries. Intraoperative frozen section helps in avoiding radical surgeries.

Case description: A 28-year-old multiparous, sterilized woman presented with complaints of lower abdominal pain, but no menstrual complaints. Abdominal examination revealed 14 × 14 cm solid with cystic lesion occupying lower abdomen. Ultrasound (US) and contrast enhanced computed tomography (CECT) of the abdomen showed a complex left ovarian cyst with torsion. CA 125 was extremely elevated 3646 U/mL. Other tumor markers were normal. A staging laparotomy was done. Intra-operative findings showed the left ovary replaced by 14 × 14 cm cyst, which underwent torsion 2 times. The ovarian cyst was untwisted and a left salpingo-oophorectomy was done. The cut section of the specimen showed papillary projections of 2 × 2 cm over the inner wall of the cyst. The frozen section histopathology report was benign SPCAF. Hence radical surgery was avoided. The final histopathology report revealed benign SPCAF of the left ovary.

Conclusion: This case is presented in view of its rarity and atypical presentation. The presentation was atypical, since CA 125 was extremely elevated and imaging features suggested a malignant neoplasm. But intraoperative frozen section histopathology turned out to be a benign tumor thus avoiding radical surgery in a young patient.

Clinical significance: Diagnosis of benign SPCAF ovary is difficult as imaging features masquerade malignancy, hence intraoperative frozen section plays a crucial role in definitive diagnosis and helps in avoiding extensive surgeries.

Keywords: Case report, CA 125 antigen, Cystectomy, Frozen section, Torsion of Adnexa.

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BACKGROUND

Surface epithelial tumors account for 60–70% of ovarian tumors, of which serous tumors comprise 46%.¹ Benign serous papillary cystadenofibroma (SPCAF) is a rare subtype of benign serous cystadenofibroma. Benign SPCAF is a slow-growing benign epithelial neoplasm of the ovary with unknown etiology. It has a wide age distribution between 15 and 65 years of age. Diagnosis of SPCAF is difficult as basic investigations like ultrasonography (USG) and computed tomography (CT) abdomen masquerade malignant ovarian neoplasm. Hence, the intraoperative frozen section plays a crucial role in definitive diagnosis and thus helps in avoiding radical surgeries (Fig. 1).

CASE DESCRIPTION

A 28-year-old multiparous, sterilized woman presented with complaints of lower abdominal pain for 4 days. There were no menstrual complaints, no history of loss of appetite or loss of weight. She was a known case of type 2 diabetes mellitus on oral hypoglycemic drugs for the past 6 years. There was no history of gynecological malignancies in the family. On examination patient was clinically stable and the BMI was 40. Abdominal examination revealed a solid cystic mass of 14 × 14 cm occupying the right iliac, right lumbar, supra pubic, and epigastric regions and it was tender. A bimanual examination confirmed the same mass. Basic investigations were normal. Ultrasonography showed normal uterus

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and right ovary, Left side complex ovarian cyst of 14 × 12 × 9 cm, with torsion. CA 125 was extremely elevated-3646 U/mL, other tumor markers were within normal limits. In view of the complex ovarian cyst and elevated CA 125, contrast enhanced computed tomography (CECT) abdomen was done which showed features suggestive of a malignant ovarian cyst arising from the left ovary with torsion.

In view of a complex ovarian cyst with torsion and extremely elevated CA 125, ovarian malignancy was suspected hence staging laparotomy was done. In view of the patient's young age, proceeded with an intraoperative frozen section.

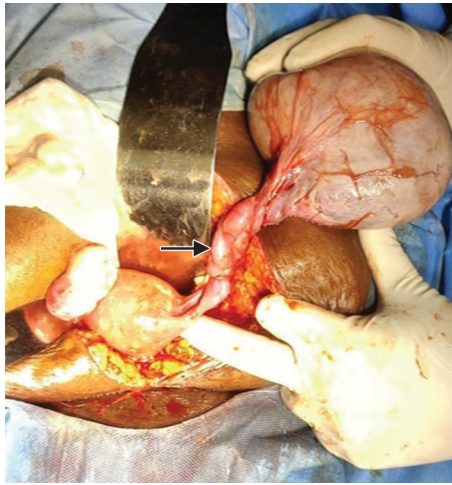


Fig. 1: Left ovarian complex cyst with torsion

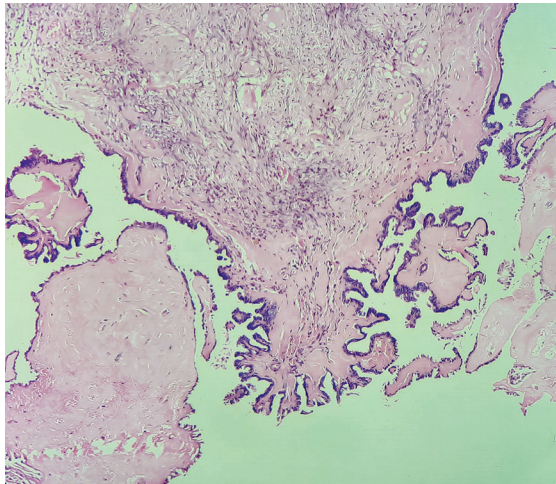


Fig. 2: Microscopic appearance of the cyst

Intraoperative—there was no evidence of ascites. Peritoneal washings were taken for cytology. The left ovary was replaced by a cyst of 14 × 14 cm, which underwent torsion twice. The surface of the cyst was smooth and there were no papillary projections. The uterus, right ovary and right fallopian tube were normal. Left ovarian cyst was untwisted and a left salpingo oophorectomy was done (Fig. 2).

The cut section showed a thick cyst wall, and smooth and regular well-defined margins. Papillary projections of 2 × 2 cm were present in the inner cyst wall. No septations noted. A left ovarian cyst was sent for a frozen section and the report turned out to be “Benign serous cystadenofibroma”. Since it was a benign tumor, salpingo oophorectomy was sufficient hence avoiding extensive surgery. The postoperative period was uneventful. Histopathological examination showed: Gross examination uniloculated cystic mass measuring 14 × 14 cm with a smooth inner wall and focal areas of papillary excrescences measuring 3.5 × 1.5 × 1 cm, wall thickness ranging from 0.3 to 0.5 cm. Microscopy section showed left ovary with fibrous cyst lined by low cuboidal to columnar cell epithelium with papillae lined by single-layered columnar epithelium. No epithelial atypia, or

cellular stratification or invasion seen. No atypical cells were seen in peritoneal fluid. The fallopian tube shows normal histology. The final histopathological report was “Benign SPCAF” the patient is under regular follow-up. CA 125 levels returned to normal and the patient is asymptomatic with no recurrence of abdominal mass or abdominal pain.

DISCUSSION

Benign SPCAF is a rare subtype of serous epithelial tumors. It is slow-growing with unknown etiology. Mostly occurs between 15 and 65 years of age.² It contains both epithelial and fibrous stromal components. It contains cystic as well as solid components. Mostly unilateral, but sometimes can affect both ovaries. Mostly presents as a single mass within the ovary or can occur as multiple masses.

Usually, it is asymptomatic, diagnosed as an incidental finding, but it may present with abdominal pain or vaginal bleeding as well. Complications like rupture of a cystic portion of the mass or torsion of the affected ovary can occur.

Routine imaging features of this tumor mimic malignancy. The presence of fibrous components gives a characteristic low-intensity appearance on magnetic resonance imaging (MRI) thus helping in differentiating benign from malignant neoplasm.³ Macroscopically they appear cystic with thin walls, may lack solid areas, and show papillary projections sometimes. They may grow up to 20 cm in diameter, encapsulated, multi-loculated with short broad papillary projections. Treatment is complete surgical removal of the tumor. It has an excellent prognosis. As gross examination and ultrasonographic features masquerade malignancy, the intraoperative frozen section helps in avoiding unnecessary radical surgeries.

Serum concentrations of CA 125 are rarely elevated beyond 1000 U/mL in benign conditions of the ovary.⁴ In this case CA 125 levels were extremely elevated to 3646 U/mL with imaging and macroscopic appearance mimicking malignancy. Staging laparotomy was planned with suspicion of malignancy. In view of the young age of patient proceeded with the intraoperative frozen section and the histopathology report turned out to be benign SPCAF. Hence avoided radical surgery.

A similar case was reported in the literature, a case of benign ovarian cyst with extremely elevated CA 125 by Tolman et al.⁴ He reported a case of benign serous cystadenofibroma with a highly elevated CA 125 value of 2897 U/mL. Extremely elevated CA 125 in our case could be due to stretching of the peritoneum by an ovarian cyst. Another factor contributing to elevated CA 125 in this could be torsion of the ovarian cyst. There are case reports in the literature showing elevated CA 125 in benign ovarian cysts which underwent torsion.⁵ Clinicians should be aware that a false positive increase of CA 125 is possible in benign lesions of the ovary.

Intraoperative frozen section plays an important role in avoiding radical surgery. We would like to emphasize the need for a frozen section facility in every tertiary care health facility.

Clinical Significance

This case of benign SPCAF is reported in view of its rarity and its atypical presentation in terms of extremely elevated CA 125, gross and imaging features masquerading as malignant neoplasm. Intraoperative frozen section played a crucial role and extensive surgery was avoided in this case.

Clinicians should be aware of false positive incidents of tumor markers and need to plan treatment accordingly. It is possible to avoid radical surgery for benign lesions if every tertiary care center has the facility of intra-operative frozen section.

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