

## Case Report

# Coexistence of borderline ovarian epithelial tumor, primary pelvic hydatid cyst, and lymphoepithelioma-like gastric carcinoma

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## Abstract

**Objectives:** Borderline ovarian tumors (BOTs) represent a heterogeneous group of ovarian epithelial neoplasms. Despite a favorable prognosis, 10–20% of BOTs exhibit progressively worsening clinic. Primary involvement of pelvic organs with echinococcus is very rare. Lymphoepithelioma-like gastric carcinoma is a rare neoplasm of the stomach.

**Case Report:** A 58-year-old woman referred with abdominal swelling and gastric complaints. Imaging studies revealed a huge cystic mass with multiple septations and solid component, another cystic mass with an appearance of cyst hydatid in the pelvis, and thickening of the small curvature of stomach. Gastroscopy revealed an ulcer with a suspicious malignant appearance, and histology of the endoscopic specimen showed severe chronic inflammation and lymphocytic infiltration. No other involvement of hydatid cyst was detected. In the exploration, there was a 25 cm cystic lesion with solid components arising from right ovary, another 6 cm cyst over the former, 7 cm cystic lesion arising from left ovary, and 10 cm mass near the small curvature of the stomach. Excision of the masses; total gastrectomy with esophagojejunal anastomosis; total abdominal hysterectomy; bilateral salpingo-oophorectomy; omentectomy; appendectomy; splenectomy; and pelvic, paraaortic, and coeliac lymphadenectomy were performed. Final pathology revealed lymphoepithelioma-like gastric carcinoma, bilateral serous BOT, and hydatid cyst.

**Discussion:** Hydatid cyst should always be considered in the differential diagnosis of abdominopelvic masses in endemic regions of the world. Preoperative diagnosis of primary pelvic hydatid disease is difficult and awareness of its possibility is very important especially in patients residing in or coming from endemic areas.

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**Keywords:** Borderline ovarian tumor; Explorative laparotomy; Lymphoepithelioma-like gastric carcinoma; Primary pelvic hydatid cyst

## Introduction

Epithelial ovarian tumors of borderline malignancy are tumors with histologic features and biologic behavior between benign and frankly malignant epithelial ovarian neoplasms. By International Federation of Gynecology and Obstetrics stage,

10-year relative survival of ovarian tumors of borderline malignancy is: Stage 1, 99%; Stage 2, 98%; Stage 3, 96%; and Stage 4, 77% [1]. Factors most commonly associated with poor outcome are advanced stage at presentation, presence of surgical residual, and presence of invasive peritoneal implants. Stage, histologic type, and age have prognostic significance [2]. In general, this tumor type has a favorable prognosis. Nevertheless, 10–20% of borderline ovarian tumors (BOTs) exhibit a progressively worsening clinical course, with widespread peritoneal implants and death of the patient within 5 years [3].

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Hydatid disease, or echinococcosis, is a parasitic infection caused by echinococcus larvae. *Echinococcus granulosus* is the most common type and endemic in the Mediterranean, Middle East, Eastern Europe, and South America. Echinococcus cysts are found mostly in the liver (60%) and lung (15%), but they can be located in any part of the body. Pelvic echinococcosis is rare with an incidence between 0.2% and 0.9% [4,5]. Nearly 80% of all pelvic masses involve the genital area, the ovary being the most frequent location, followed by the uterus. These cases are usually secondary to the accidental rupture of a cyst in other areas of the body [4]. However, primary pelvic hydatid cysts have also been reported [6,7].

Lymphoepithelioma-like gastric carcinoma is a rare neoplasm of the stomach with a better prognosis than conventional adenocarcinoma. It constitutes a 3.8% of gastric carcinomas [8]. Most lymphoepithelioma-like gastric carcinomas are associated with Epstein-Barr virus (EBV) infection, whereas a subset is associated with microsatellite instability [9].

In the present case, simultaneous coexistence of two different malignancies and primary pelvic echinococcus in the same patient are reported.

### Case report

A 58-year-old, Gravida 6, Para 6 woman was referred with abdominal swelling, gastric complaints, and pelvic pain. Abdominal ultrasonography revealed a huge mass within the umbilical region 80 × 63 mm in size, and another mass 25 cm in size arising from the right pelvis, additionally another 8 cm mass over the former. Computerized tomography suspected pelvic hydatid disease with no involvement of the liver and also revealed thickening in the small curvature of the stomach. Gastroscopy revealed an ulcer with a suspicious malignant appearance. Histology of the endoscopic specimen showed severe chronic inflammation and lymphocytic infiltration with *Helicobacter pylori* organisms. No other involvement of hydatid cyst was detected. In laboratory tests, tumor marker levels were normal.

On exploratory laparotomy, a 25 cm cystic lesion with solid components arising from right ovary, another 6 cm cyst over the former, 7 cm cystic lesion arising from left ovary, and 10 cm mass near the small curvature of the stomach were found. Frozen sections from each of the pelvic masses showed bilateral serous tumor of borderline malignancy and scolexes consistent with hydatid disease. Frozen section of the mass near the small curvature of the stomach showed malignant lymphoepithelioma-like gastric tumor. General surgeons attended the operation. Excision of the masses; total gastrectomy with esophagojejunal anastomosis; total abdominal hysterectomy; bilateral salpingo-oophorectomy; omentectomy; appendectomy; splenectomy; and pelvic, paraaortic, and celiac lymphadenectomy were performed.

Final pathology revealed lymphoepithelioma-like gastric carcinoma, bilateral serous tumor of borderline malignancy, and pelvic hydatid cyst (Figs. 1–3). Only one celiac lymph node and six lymph nodes selected from omentum showed

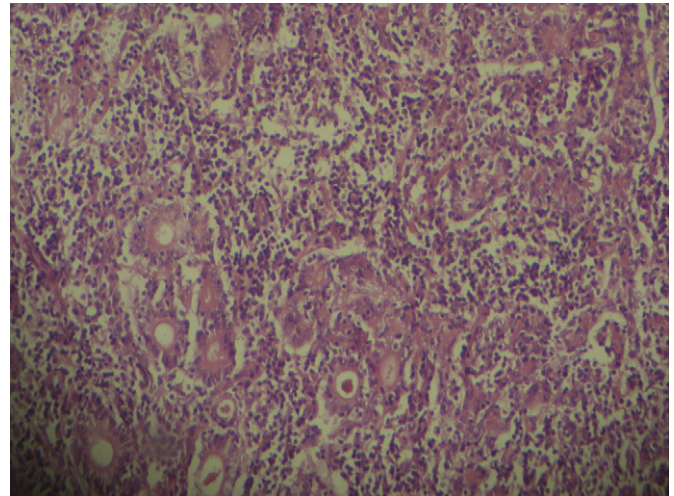


Fig. 1. Lymphoepithelioma-like gastric carcinoma (hematoxylin and eosin, 100×).

lymphoepithelioma-like gastric carcinoma metastases. Bilateral ovarian tumors showed no capsular invasion and no serous ovarian tumor metastases were detected in pelvic and para-aortic lymph nodes. Both tubes and paratubal regions did not show tumoral infiltration.

Formalin-fixed and paraffin-embedded tissue section of the gastric tumor was analyzed by quantitative real-time polymerase chain reaction for the detection of EBV. No EBV DNA was detected.

After certain diagnoses, the patient was conducted to general surgery, medical oncology, and infectious diseases departments.

### Discussion

The coexistence of epithelial BOT, primary pelvic hydatid cyst, and lymphoepithelioma-like gastric carcinoma in the same patient is exceedingly rare.

Hydatid disease located in the pelvis is very rare. In most cases, coexistent cysts are detected elsewhere, usually in the

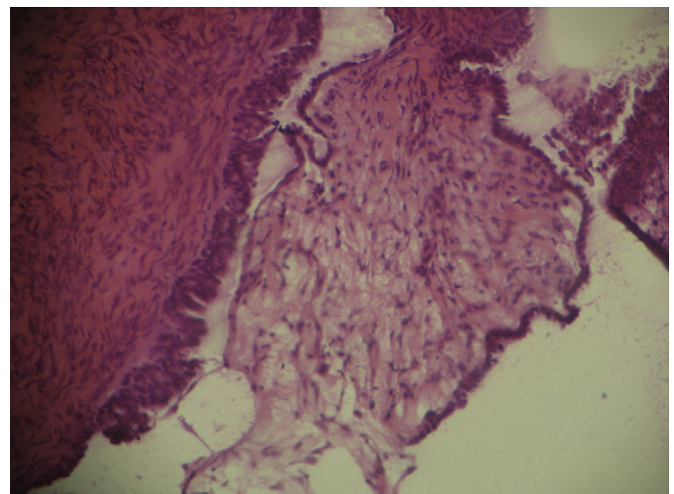


Fig. 2. Borderline ovarian tumor (hematoxylin and eosin, 40×).

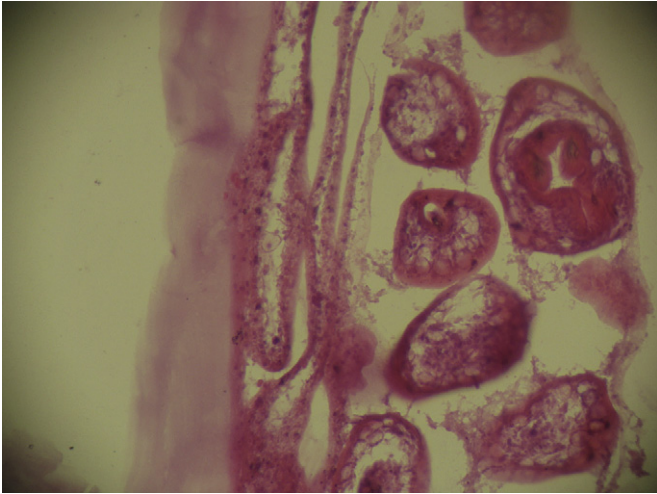


Fig. 3. Echinococcosis with cuticle material on the left side and daughter vesicles on the right side (hematoxylin and eosin, 20 $\times$ ).

liver. Only a few cases of primary pelvic hydatid cyst have been reported. Primary pelvic echinococcosis may simulate malignancies [4,5,10], multicystic ovary [11], and may be disseminated as ovarian carcinomatosis [12]. Ladeb et al [13] reported that only 2% of cystic hydatid disease cases were localized in the pelvis. Safioleas et al [14] reported an isolated hydatid disease in the pelvis attached to the urinary bladder. Varedi et al [15] described a case of primary pelvic hydatid cyst presenting with weight loss and abdominal pain.

Serodiagnostic assays, ultrasonography, and computed tomography can be helpful for preoperative diagnosis, but definite diagnosis is confirmed by histological examination. Although ultrasonography is usually the initial diagnostic tool, computed tomography is much more efficient in demonstrating calcification and daughter cysts and is therefore regarded more sensitive and accurate than ultrasonography for the differential diagnosis [7]. Especially, in patients with coexistence of different pelvic and/or abdominal masses, as seen in the present case, importance of tomography is manifested. Surgical intervention is the treatment of pelvic hydatid cyst. Laparotomy should be the choice to avoid intraoperative rupture of the cyst. It is very important to use a scolical agent in the operating field, such as a sodium chloride solution or a povidone-iodine solution. The treatment for recurrence and complications is very difficult and definition of the best option needs multidisciplinary approach; therefore, medical therapy should be applied postoperatively in the patients with multiple and/or large cysts. There are many studies showing that pre- and postoperative use of mebendazole from the beginning of the '80s, and albendazole in the following years decreased recurrence rates from 80% to 10% [16–18]. The case presented here was given albendazole (per oral 400 mg/d) therapy for 6 months after discharging from the hospital. Abdominal computerized tomography at the postoperative 6<sup>th</sup> month did not show any recurrence.

Lymphoepithelioma-like carcinomas are defined as tumors with histologic similarity to nasopharyngeal carcinoma. Lymphoepithelioma-like carcinoma of the stomach is a rare

type of gastric carcinoma that was first described by Watanabe et al [19] in 1976 as gastric carcinoma with lymphoid stroma. It constitutes 3.8% of gastric carcinomas [8]. More than 80% of lymphoepithelioma-like gastric carcinomas have been found to be related to EBV infection [20]. However, the association of EBV with some epithelial neoplasms has been reported to depend on ethnic and/or regional background [21,22]. Specifically, the association of EBV with lymphoepithelioma-like carcinoma of the salivary gland and lung is restricted to Asian patients, whereas the association of EBV with gastric and thymic lymphoepithelioma-like carcinoma is independent of race [23]. The present case was one of an EBV-negative lymphoepithelioma-like gastric carcinoma. To the best of our knowledge, this is the first report of a simultaneous coexistence of two different malignancies and primary pelvic echinococcus in the same patient.

In conclusion, the present report demonstrates simultaneous coexistence of two different malignancies and primary pelvic echinococcus in the same patient. Hydatid disease can affect any organ in the body and a high suspicion of this disease is justified in any cystic neoplasm of any organ, especially in endemic regions. Preoperative diagnosis of primary pelvic hydatid disease is difficult and awareness of its possibility is very important especially in patients residing in or coming from endemic areas.

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