Cholangiocarcinoma masquerading as an ovarian tumour

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CASE PRESENTATION

A 76-year-old woman presented to the emergency department complaining of nausea, vomiting, diarrhea, right-sided abdominal pain and early satiety. Her physical examination was within normal limits, with the exception of nodularity in the cul-de-sac and right parametrium on pelvirectal examination. Liver enzyme levels were elevated: aspartate aminotransferase 150 U/L; alanine aminotransferase 369 U/L; gamma-glutamyl transferase 953 U/L; alkaline phosphatase 370 U/L; and a direct bilirubin level of 12 $\mu\text{mol/L}$. A computed tomography scan of the abdomen and pelvis showed minimal dilation of the biliary system, with no obvious mass lesions or obstruction (Figure 1). However, multiple soft tissue nodules within the omentum and a prominent cystic lesion within the right adnexa were apparent. Pelvic ultrasound revealed a multiseptated right adnexal mass 6.2 cm in size, which raised concern for an ovarian cystic neoplasm. A subsequent carbohydrate antigen (CA) 125 level of 129 U/mL was measured. The patient underwent a total abdominal hysterectomy and bilateral salpingo-oophorectomy with pelvic lymph node dissection, omentectomy, appendectomy, and resection of the retroperitoneal mass in the posterior cul-de-sac and left uterosacral ligament. A frozen section of the right adnexal mass intraoperatively showed a mucinous cystadenoma with no evidence of malignancy (Figure 2). However, final pathology results yielded a well-differentiated mucinous adenocarcinoma consistent with metastasis from a primary cholangiocarcinoma. Metastatic cholangiocarcinoma was also identified in the cervix, uterine serosa, left fallopian tube, retroperitoneal posterior cul-de-sac lesion, left uterosacral lesion and the omentum. Both the ovarian neoplasm and omental deposits stained negative for CA 125. A serum CA 19.9 level of 30,000 U/mL was measured, necessitating an endoscopic retrograde cholangiopancreatography (ERCP) for further evaluation. ERCP demonstrated a complete obstruction/stricture 3 cm in size in the proximal common hepatic duct up to the hilum. A biliary stent was successfully placed above the stricture with adequate drainage of bile.

Figure 1 Computed tomography scan of the abdomen and pelvis showing a prominent cystic lesion within the right adnexa (arrow)

DISCUSSION

Cholangiocarcinoma (CC) is a rare malignancy of the biliary epithelium within the intrahepatic or extrahepatic bile ducts. It carries a poor prognosis and surgical management is the only curative treatment currently available. However, surgery is often extensive and is associated with significant morbidity and mortality (1). CC with metastasis to the ovary can present a diagnostic challenge because it has the propensity to mimic primary mucinous neoplasms (2,3). Intraoperative frozen sectioning provides a limited sample and can cause significant error, as in the present case. Therefore, multiple sampling, histological staining, clinical findings and gross features are extremely important in the diagnosis of metastatic CC in which an ovarian mass is the initial discovery. The literature regarding ovarian metastasis from the biliary system is limited. An autopsy study from Japan (3) suggested that CC is likely under-reported. However, due to differing incidences of primary tumours around the world, it is difficult to infer whether this is also the case in North America (3). To the best of our knowledge, the present case is the first to be reported in Canada.

Figure 2 A Hematoxylin and eosin staining (2.5 $\times$ power) of the corpora albicans of the ovary showed a mucinous cyst neoplasm. B Immunohistochemical staining of ovarian sample positive for cytokeratin 7. C Immunohistochemical staining of ovarian sample negative for Wilms tumour 1. D Immunohistochemical staining of ovarian sample positive for carbohydrate antigen 19.9. E Immunohistochemical staining of ovarian sample positive for cytokeratin 20

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