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CASE REPORT

Ovarian Malignant Mixed Germ Cell Tumour Presenting as Advanced Disease in an Adolescent Girl

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ABSTRACT

Ovarian malignant germ cell tumours are uncommon and predominantly affect girls and young women. Dysgerminomas (equivalent to testicular seminomas) and non-dysgerminomatous tumours are the 2 most common histological types; the mixed type is even rarer and carries a worse prognosis. This report is of a 15-year-old girl with malignant mixed germ cell tumour who presented with advanced disease. She underwent tumour debulking followed by intensive chemotherapy, and is subsequently in remission.

Key Words: *Dysgerminoma; Endodermal sinus tumor; Neoplasms, germ cell and embryonal; Ovary*

INTRODUCTION

Malignant germ cell tumours comprise less than 5% of all ovarian neoplasms.[1] The incidence of malignant ovarian germ cell tumours ranges from 1% to 6% and 8% to 19% in Caucasian and Asian populations, respectively.[2] There are 2 major histological groups: dysgerminomas (equivalent to testicular seminomas) and non-dysgerminomatous tumours (of which yolk sac tumour is the commonest). Pure dysgerminoma and yolk sac tumours constitute 50% and 20% of all malignant germ cell tumours, respectively. The remaining, less common, entities include embryonal carcinoma, immature teratoma, choriocarcinoma, polyembryomas, and mixed germ cell tumours. This report is of an adolescent girl with malignant mixed germ cell tumour who presented with advanced disease.

CASE REPORT

A 15-year-old previously healthy girl presented to the Prince of Wales Hospital, Hong Kong, in 2002 with insidious onset of malaise and abdominal distension. Physical examination revealed a large abdominal mass. Computed tomography (CT) of the abdomen and pelvis showed a large predominantly cystic mass occupying the central abdomen and pelvis. Irregular septations and ill-defined enhancing nodular components were seen along the enhancing capsule of the mass (Figure 1). The lesion caused obstructive hydronephrosis bilaterally (Figure 2). Extensive metastatic peritoneal nodules, ascites (Figure 1), and liver metastases were also evident (Figure 2). Laboratory investigation revealed elevated serum α-foetoprotein (AFP) level. Both imaging features and blood biochemistry suggested aggressive ovarian germ cell tumour.

In view of the extensive disease, debulking of the primary tumour was first attempted with right salpingo-oophorectomy. Histology confirmed the lesion to be malignant mixed germ cell tumour with predominantly endodermal sinus (yolk sac) tumour, and a smaller portion of dysgerminoma.

The patient was given 6 cycles of intensive chemotherapy, consisting of etoposide, carboplatin, and bleomycin, and radiotherapy. A second laparotomy for omentectomy and removal of the peritoneal nodules was performed. After completion of the treatment regimen, follow-up CT showed complete resolution of the peritoneal and liver metastases (Figure 3). The patient is currently in remission with preserved ovarian function, and has achieved disease-free survival of 5 years.

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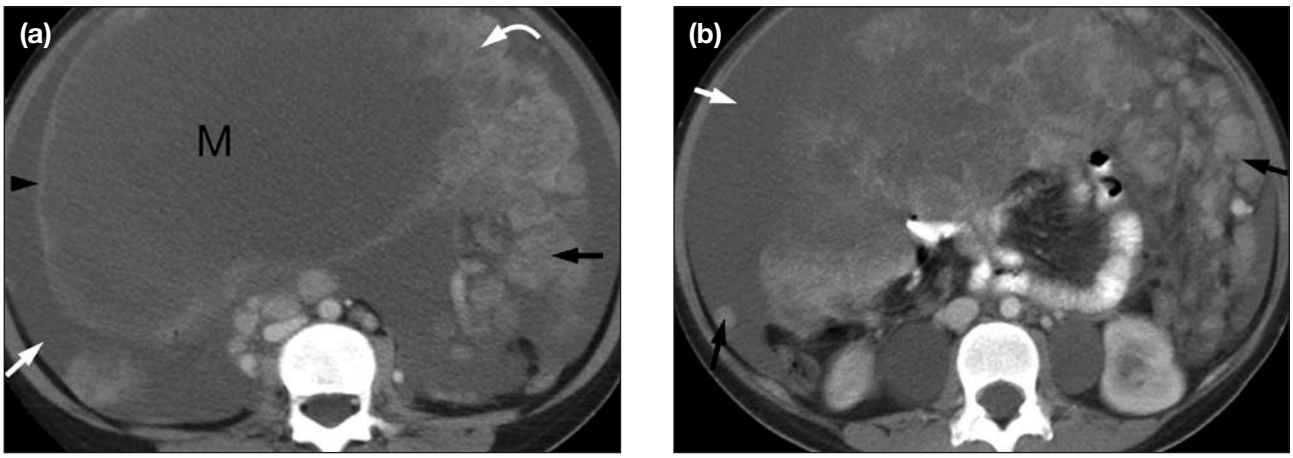

Figure 1. Contrast-enhanced axial computed tomography images of the lower abdomen showing (a) a large predominantly cystic ovarian mass (M) with peripheral enhancing capsule (arrowhead) and solid enhancing components (curved arrow); and (b) ascites (white arrow) and multiple omental metastases (black arrows).

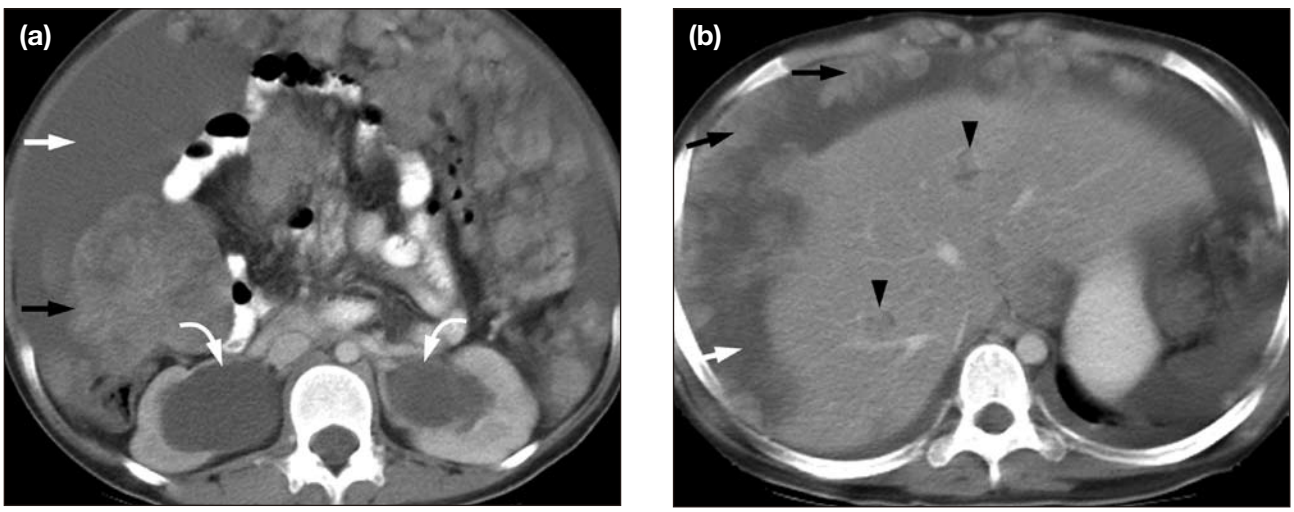

Figure 2. Contrast-enhanced axial computed tomography images of the upper abdomen showing (a) bilateral obstructive hydronephrosis (curved arrows); and (b) ill-defined metastatic lesions in the liver (black arrow heads), multiple metastatic omental soft tissue nodules (black arrows), and ascites (white arrow).

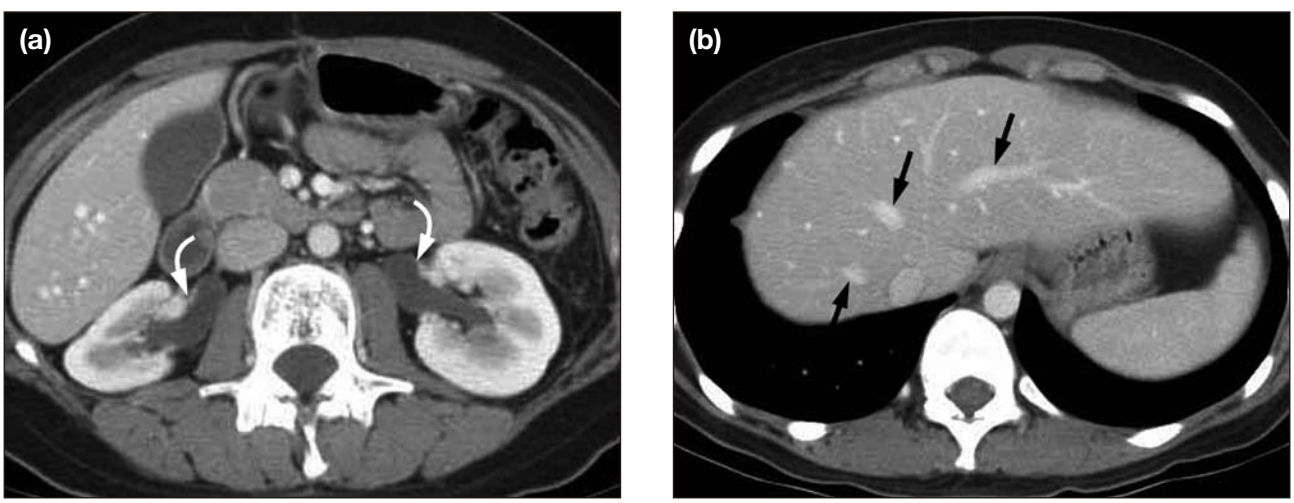

Figure 3. Contrast-enhanced axial computed tomography image of (a) the upper abdomen showing mild residual bilateral hydronephrosis (curved arrows), while the peritoneal metastases and malignant ascites have completely resolved; and (b) the liver showing resolved liver lesions at the level of the hepatic veins (black arrows).

DISCUSSION

Mixed germ cell tumour of the ovary is rare. However, a combination of endodermal sinus and dysgerminoma (similar to this patient) accounts for one-third of reported cases.[3] The endodermal sinus component is aggressive and carries a poor prognosis. The reported mortality rate is >90% within 2 years of diagnosis. At laparotomy, spread beyond the ovary is seen in approximately 30% of patients, and usually consists of metastases to the liver and peritoneum and, less frequently, to the lymph nodes, lung, and diaphragm.[3]

Although the radiological features do not distinguish the different histological subtypes of germ cell tumour, the advanced stage of disease in this patient suggested an aggressive subtype with a poor prognosis.

Ultrasound is the primary investigation for an abdominal mass. Ultrasound usually shows a large pelvic mass that extends into the abdomen and has both solid and cystic components. Concurrent dermoid cysts of the ovary, the presence of ascites, and urinary tract obstruction have been reported on ultrasound and CT.[4,5] CT is the investigation of choice to characterise the complex nature of the primary tumour at the ovary, and the cystic components of the mass are more visible on CT. CT is also helpful for staging advanced disease.[4] In stage I disease, where the tumour is localised to the ovary, the non-specific imaging appearance and symptoms may lead to misdiagnosis of other more benign conditions such as tubo-ovarian abscess, appendiceal abscess, or benign cystic teratoma of the ovary. Due to the rapid growth of the tumour, a delay in diagnosis may lead to advanced disease and a poor prognosis. Malignant germ cell tumours should therefore always be considered when adolescent girls or young women present with a complex or predominantly cystic pelvic mass. Serum AFP and human chorionic gonadotrophin are useful tumour markers during the diagnostic stage. In advanced disease, CT helps to evaluate liver and peritoneal metastases, as for this patient. CT also plays a valuable role during follow-up for assessing the response to chemotherapy.

Treatment of malignant germ cell tumour of the ovary consists of salpingo-oophorectomy with adjunctive chemotherapy. Chemotherapeutic regimens have evolved to combination therapy with overall disease-free survival rates of >95%.[6] For this patient, despite the advanced stage of the disease, an aggressive treatment protocol including surgical removal of the primary tumour and metastases, combination chemotherapy, and radiotherapy have successfully achieved disease remission.

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