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Malignant ovarian germ-cell tumours

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Malignant ovarian germ-cell tumours account for about 5% of all ovarian malignancies and typically present in the teenage years. They are almost always unilateral and are exquisitely chemosensitive. As such, the surgical approach in young women with such tumours confined to a single ovary should aim to preserve fertility. In early disease, a unilateral salpingo-oophorectomy with careful surgical staging is of great importance in selecting appropriate adjuvant therapy. In advanced disease, the role of aggressive cytoreducation is not well defined, and removal of both ovaries does not confer improvement in outcome. Bleomycin, etoposide and cisplatin combination chemotherapy is regarded as the gold standard for adjuvant therapy. Studies evaluating ovarian and reproductive capacity after conservative surgery and chemotherapy for malignant ovarian germ-cell tumours have consistently demonstrated excellent prognosis, with the return of normal menstrual function and fertility rates in these women with no increase in the risk of teratogenicity.

Introduction

Malignant ovarian germ-cell tumours (MOGCTs) account for 5% of all ovarian malignancies in Western countries. Because they principally occur during adolescence and early adulthood, decisions on

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treatment used to present a challenge to the gynaecologic oncologist. Before the mid-1960s, virtually all women with advanced non-dysgerminomatous disease died. Even for the exquisitely radiosensitive dysgerminomas, women who had been radiated were left with their fertility destroyed. Subsequent introduction of effective combination chemotherapy brought about dramatic improvements in the prognosis for MOGCTs. These have largely been extrapolated from the experience of treating the more common testicular germ-cell tumours. Many randomised-controlled trials for testicular germ-cell tumours have provided a strong evidence base for treatment decision making in MOGCT.[1,2] In the evolution of care, combination chemotherapy using vincristine, dactinomycin, and cyclophosphamide (VAC regimen) was able to achieve cure rates of 86% in women with International Federation of Gynecology and Obstetrics (FIGO) Stage I non-dysgerminomatous disease,[3] but women with metastatic disease then had a 50–70% mortality.[3,4] The introduction of cisplatin-containing drug regimens, notably the BEP (bleomycin, etoposide and cisplatin) regimen from the 1980s onward, resulted in a 5-year survival rate of up to 100% for dysgerminomas and 85% for non-dysgerminomatous MOGCT.[5–7] Prompt initiation of appropriate chemotherapy is the critical factor for young women with an advanced MOGCT.[8] Contemporary principles of surgery for MOGCTs dictate that fertility preservation is appropriate even in the face of extensive metastatic disease if fertility is desired, because there is no evidence that removing an uninvolved ovary enhances survival. Hence, the focus of subsequent studies on MOGCT has shifted to long-term sequelae for these young women, with particular emphasis on ovarian and reproductive function, including teratogenicity, especially after combination chemotherapy.

Classification of malignant ovarian germ-cell tumours

The current 2003 World Health Organization classification of system for MOGCT is as shown in Table 1.[9]

Table 1
Histological classification of malignant ovarian germ-cell tumours.[9]

Primitive germ-cell tumours
Dysgerminoma
Yolk-sac tumour
Polyvesicular vitelline tumour
Glandular variant
Hepatoid variant
Embryonal carcinoma
Polyembryoma
Non-gestational choriocarcinoma
Mixed germ-cell tumour, specify components
Biphasic or triphasic teratoma
Immature teratoma
Mature teratoma
Solid
Cystic, dermoid cyst
Fetiform teratoma, homunculus
Monodermal teratoma and somatic-type tumours associated with biphasic or triphasic teratoma
Thyroid tumour group
Carcinoid group
Neuroectodermal tumour group
Carcinoma group
Melanocytic group
Sarcoma group
Sebaceous tumour group
Pituitary-type tumour group
Retinal anlage tumour group
Others