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Recent Advances in Granulosa Cell Tumor Ovary: A Review

Granulosa cell tumors constitute less than 5 % of all ovarian tumors. Unlike epithelial ovarian tumors, they occur in a younger age group, are usually detected in an early stage and often have features of hyperestrogenism. The presenting symptoms are usually nonspecific with abdominal pain or distension. They follow an indolent course and are characterized by a long natural history. Mutation of FOXL2 (402C->G) seen in 97 % of adult GCT may be pathognomonic for adult GCT. Only stage of the disease has been consistently shown in various studies to affect survival of patients with GCT. The initial management of patients, for whom fertility is not an issue, is total abdominal hysterectomy, bilateral salpingo-oophorectomy and removal of all gross disease. Nodal dissection is not a significant factor for survival and is not recommended in surgical staging of GCT. Fertility preserving surgery with unilateral salpingo-oophorectomy is feasible in young patients with stage Ia GCT. Patients with early stage disease (stage I and II) have a very good prognosis with 5 year DFS and OS of 89 % and 99 % respectively and these groups of patients usually don't require any postoperative treatment. Patients with stage Ic disease associated with poor prognostic factors like large tumor size or high mitotic index and stage II, have a higher chance of relapse, and may benefit with postoperative treatment but role of chemotherapy is still debatable. In advanced stage disease (stage III and IV) the 5 year DFS and OS disease was 72 % and 80 % respectively hence the option of postoperative treatment with 6 cycles of BEP should be considered in this group. Recently paclitaxel is being investigated as an effective tool in GCT. The efficacy of radiation in GCT is not well defined but in optimally debulked cases postoperative radiation is a viable option. Due to high chance of recurrence even years after apparent clinical cure of the primary tumor, lifelong follow up with clinical examination and tumor markers like inhibin B is recommended. About 25 % GCT develop recurrence and the median time to recur is usually 4–5 years. Most recurrences are intraperitoneal and usually a complete debulking of the disease is feasible even in the recurrent setting. Postoperative chemotherapy (platinum based) is usually given after surgery more so in cases with widespread disease or after suboptimal cytoreduction. Recurrent chemoresistant, progressive non-responding GCT or patients with high surgical risk are ideal candidates for targeted therapy.