CASE REPORT

Malignant Ovarian Tumors in Adolescents: A Case Series

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Abstract

Objective: To analyze the case series of malignant ovarian tumor in adolescents (13–21 years) with various presentations, histopathological types and management.

Design: Case series.

Settings: Department of Obstetrics and Gynaecology, Sri Ramachandra Medical College and Research Institute (SRMC), Chennai, Tamil Nadu, India.

Period: January 2015 to January 2016.

Materials and methods: Adolescent female patients with malignant ovarian tumor were included. Clinical history, examination findings, radiological findings, surgical management, histopathology types were analyzed.

Results: During the study period of 1 year, 10 girls with malignant ovarian tumors were studied. Majority of patients were 19–21 years of age. The main presenting complaints were pain abdomen in 50% of patients, followed by abdominal distension in 30% and menstrual irregularities in 20%. Of the 10 cases, 8 were of non-epithelial origin and 2 were epithelial. Fertility-conserving treatment was performed in all patients but for a single patient with endodermal sinus tumor (EST).

Conclusion: Though benign ovarian lesions are very common, malignant ovarian tumor represent 1.5% of all tumors in adolescents. Clinical examination, radiological and biochemical tests together help us in arriving at an accurate diagnosis. The main attention is towards the timely diagnosis and fertility-conserving procedures in these young patients.

Keywords: Abdominal pain, Adolescent, Fertility preservation, Ovarian tumors.

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Introduction

Ovarian tumors are relatively rare in adolescents. Adolescents with pelvic mass may be asymptomatic or may have chronic or acute symptoms.[1] The pressure of enlarging ovarian mass can cause bowel-related symptoms such as constipation, vague abdominal discomfort, urinary frequency or even ureteric obstruction.

A ruptured ovarian cyst is a classic diagnosis when an adolescent presents with pelvic pain. Alternatively ovarian masses can cause severe acute or intermittent symptoms, caused by torsion, intraperitoneal rupture or bleed into ovarian tissue.[2] This condition can represent true surgical emergency and diagnosis can be challenging.

Laboratory studies should include pregnancy test (to exclude unmarried pregnancy), CBC (to exclude infective/inflammatory conditions) and tumor markers (α fetoprotein, β hCG, LDH, CEA, CA 125, CA-19.9 for preoperative diagnosis and follow-up).[2]

Ultrasound is a valuable tool for diagnosing pelvic masses in adolescents. Transabdominal ultrasound is preferred as transvaginal USG may not be possible in adolescents. In patients for whom a diagnosis is not reached by ultrasound, other cross sectional imaging tests (CT or MRI) can give additional information for arriving at a diagnosis.

If ovarian tumor is diagnosed in an adolescent female, conservative surgery should be the first choice to preserve reproductive function.[3]

Materials and Methods

This is a descriptive study conducted in Obstetrics and Gynaecology Department, SRMC. Consecutive female patients with ovarian tumors were included. Clinical history, examination findings, surgical management and histopathological types were analyzed (Tables 1 and 2).

The following data were collected: age of patient, clinical presentation, tumor size, unilateral or bilateral involvement, type of surgery, presence or absence of extra ovarian spread, histopathology and chemotherapy.

Results

During this period, 10 girls between the age group 13–21 years with malignant ovarian tumor were studied (Table 1). Pain abdomen (50%) and abdominal distension (30%) and menstrual irregularities (20%) were the commonest clinical presentation in these patients (Table 1). All these patients were diagnosed to have malignant ovarian tumors of which eight were non-epithelial tumors and the rest were epithelial tumors.

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Imaging revealed predominantly cystic tumors in seven patients and predominantly solid tumors in three patients. Tumor markers were done for all these patients such as CA-125, β hCG, α fetoprotein, LDH, CEA, CA 19.9 (Table 3).

Of the 10 patients, the first patient (age 21 years) underwent unilateral laparoscopic ovarian cystectomy. Histopathology report revealed endodermal sinus cell tumor. The second patient (age 20 years) underwent diagnostic laparoscopy. Frozen section revealed dysgerminoma, hence proceeded with staging laparotomy with unilateral ovariotomy, omental biopsy and underwent postoperative chemotherapy with injection etoposide and injection cisplatin. The third patient (age 19 years) underwent unilateral oophorectomy with partial salpingectomy. Histopathology of this patient revealed immature ovarian teratoma and underwent postoperative chemotherapy with injection bleomycin, injection carboplatin and injection etoposide. The fourth patient (age 21 years) underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy with total omentectomy with lymph node dissection (Fig. 1). Histopathology revealed granulosa cell tumor. Patient underwent postoperative chemotherapy with injection paclitaxel, injection cisplatin. The fifth patient (age 20 years) underwent unilateral ovarian cystectomy, histopathology revealed sero mucinous tumor. Patient underwent postoperative chemotherapy. The sixth patient (age 21 years) underwent unilateral oophorectomy with omental biopsy. Histopathology showed mixed malignant germ cell tumor. Patient underwent postoperative chemotherapy (Figs 2 and 3). The seventh patient (age 21 years) underwent left salpingo-oophorectomy. Histopathology showed gonodoblastoma. Patient underwent postoperative chemotherapy (Figs 4 and 5). The eighth patient (age 21 years) underwent unilateral ovariotomy with pelvic lymph node dissection with omentectomy. Histopathology revealed borderline serous ovarian tumor. Postoperatively patient was lost to follow-up (Figs 6 and 7). The last two patients (age 13 and 15 years) were diagnosed to have germ cell tumor (embryonal carcinoma) by tissue biopsy and are undergoing neoadjuvant chemotherapy with injection bleomycin, injection carboplatin and injection etoposide (Fig. 8).

Table 1: Distribution of HPE and imaging findings of cases

	Number
Histopathological type	
Non-epithelial	
Germ cell tumor	
Dysgerminoma	1
Gonadoblastoma	1
Mixed malignant germ cell tumor	1
Immature teratoma	1
Granulosa cell tumor	1
Endodermal sinus tumor	1
Embryonal carcinoma	2
Epithelial	
Borderline serous tumor	1
Borderline seromucinous tumor	1
Imaging findings	
Predominantly cystic	7
Predominantly solid	3

Discussion

Ovarian tumors in adolescents, as in other age groups, are diagnosed when a patient presents with symptoms or at that time of general physical examination or incidental discovery during radiological imaging. Ovarian tumors are slow in progression and are diagnosed at late stage. However, timely diagnosis and well organized adolescent gynecological services can improve the prognosis. Accurate diagnosis at an early stage and preserving the reproductive ability of the patient are of foremost importance in adolescents.

Table 2: Distribution of presenting age and type of surgery

	Number
Age at presentation	
12–15 years	2
16–18 years	0
19–21 years	8
Surgical outcome	
Unilateral cystectomy	2
Unilateral oophorectomy	1
TAH with BSO	1
Unilateral ovariotomy	2
Unilateral salpingo-oophorectomy	1
Unilateral oophorectomy with partial salpingectomy	1

Table 3: Tumor markers

Type	*Tumor markers*					
Non-epithelial	*AFP*	*CA-125*	*β hCG*	*CEA*	*LDH*	*CA-19.9*
Germ cell tumor						
Dysgerminoma	N	N	N	N	⇑	N
Gonadoblastoma	N	N	N	N	⇑	N
Mixed malignant germ cell tumor	⇑	⇑	N	N	⇑	N
Immature teratoma	N	⇑	⇑	N	⇑	N
Endodermal sinus tumor	⇑	⇑	N	N	⇑	N
Embryonal carcinoma	N	N	⇑	–	–	–
Granulosa cell tumor	N	⇑	N	N	–	–
Epithelial						
Borderline serous tumor	N	⇑	N	–	–	–
Borderline seromucinous tumor	N	⇑	N	⇑	⇑	⇑

N, normal; –, not done; ⇑, increased

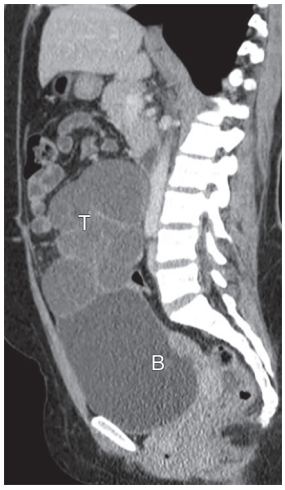


Fig. 1: Sagittal CECT showing multiloculated cystic lesion (T) superior to the urinary bladder (B). The lesion is displaced superiorly by the distended urinary bladder. The lesion was found to be granulosa cell tumor on histopathology

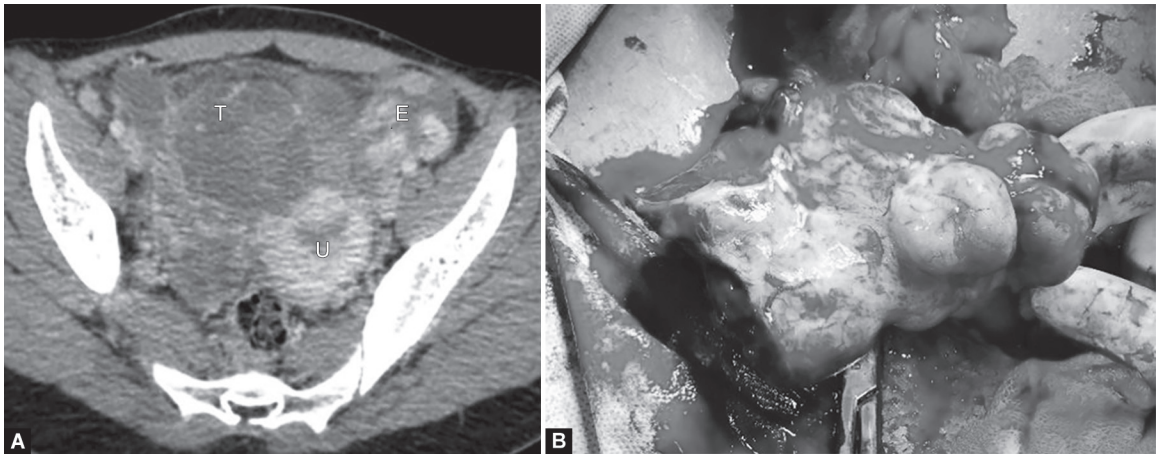
Ovarian malignancies contribute to about 1.5% of all adolescent tumors.[3] However recent studies report that in adolescents who undergo surgery for persistent ovarian neoplasms, the incidence of malignancy ranges from 4% to 9%.[4]

Gynecological tumors are rare in adolescents, but still they should be considered when an adolescent presents with mass abdomen. The commonest presenting symptoms are abdominal pain, distended abdomen, abnormal menstruation, mass abdomen, bowel or urinary symptoms.

The strategy for treating adolescent ovarian tumors is broadly similar to that for adult ovarian tumor but for the difference that importance is given for preserving anatomy as much as possible.[3] In all our cases, careful tissue handling, hemostasis preservation, steps to prevent adhesions was performed so that fertility was preserved. Frozen section was performed in all eight patients, when the result of frozen section was confirmed the above mentioned surgeries were performed.

The chances of a pelvic mass being functional are more in adolescent post menarche.[2]

Neoplasms of germ cell origin contribute to about 20–25% of all benign and malignant neoplasms. In patients less than 20 years



Figs 2A and B: (A) Axial CECT showing multiloculated cystic lesion (T) with enhancing mural component (E) nearly occupying the entire pelvis and displacing the uterus (U) to the left. The lesion was found to be mixed malignant germ cell tumor on histopathology; (B) Intraoperative photograph showing lobulated mass which turned out to be mixed malignant germ cell tumor

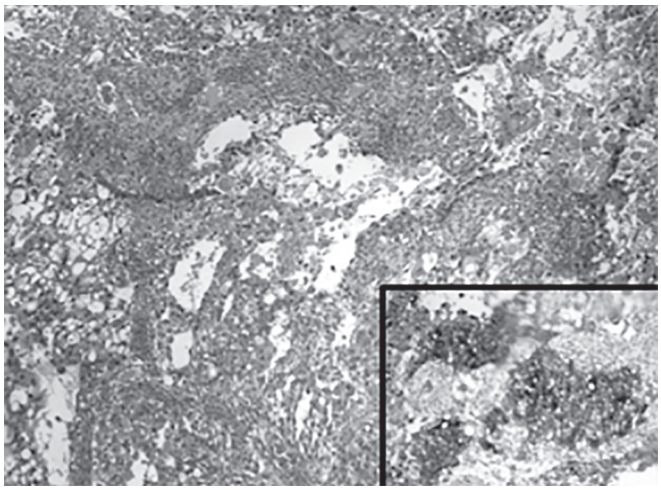


Fig. 3: HPE showing mixed malignant germ cell tumor (yolk sac tumor (80%), dysgerminoma (20%) (inset showing positive for AFP and CD 117) *100×, H&E

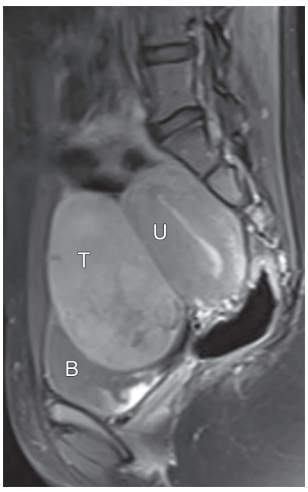


Fig. 4: Sagittal CE MR showing solid enhancing pelvic lesion (T) postero-superior to the urinary bladder (B) and anterior to the uterus (U). The lesion was found to be gonadoblastoma on histopathology

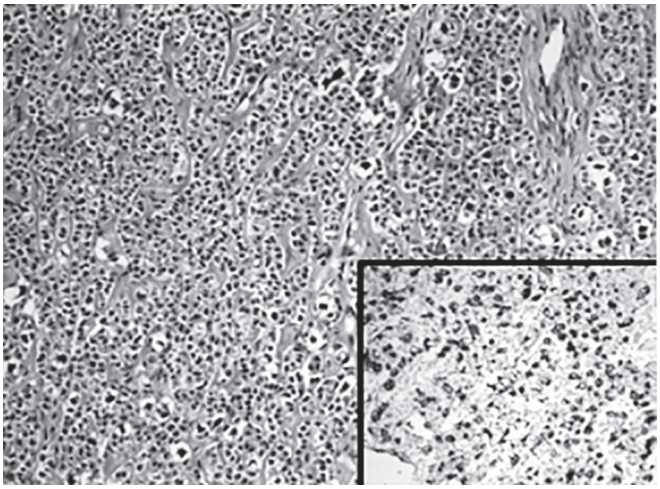


Fig. 5: Gonadoblastoma with focal granulosa cell components in left ovary. *100×, H&E. Inset shows positive for CD117

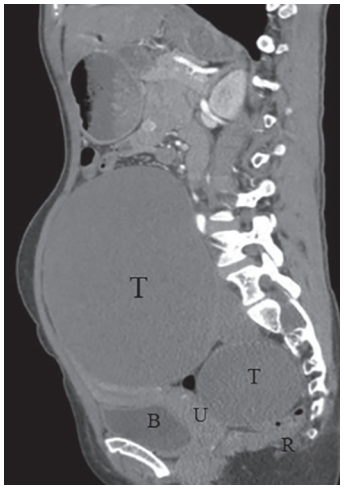


Fig. 6: Sagittal CECT showing large abdominopelvic multiloculated cystic lesion (T). The lesion is compressing the uterus (U) antero-inferiorly. The rectum (R) is compressed posteroinferiorly by the lesion. The lesion was found to be borderline serous tumor on histopathology

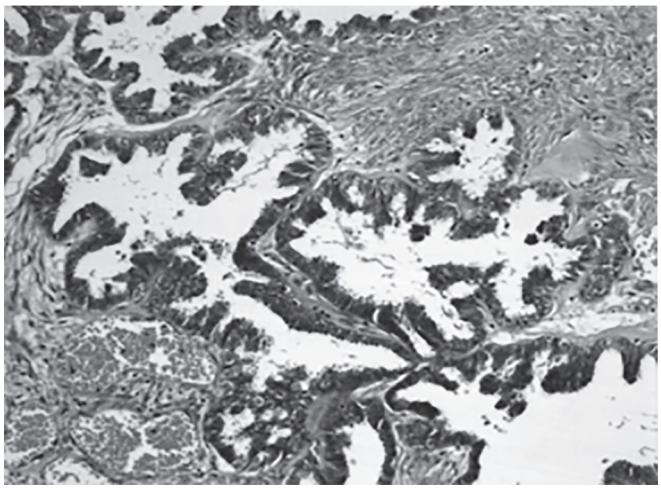


Fig. 7: Borderline serous tumor of right ovary showing complex papillary architecture *100×, H&E

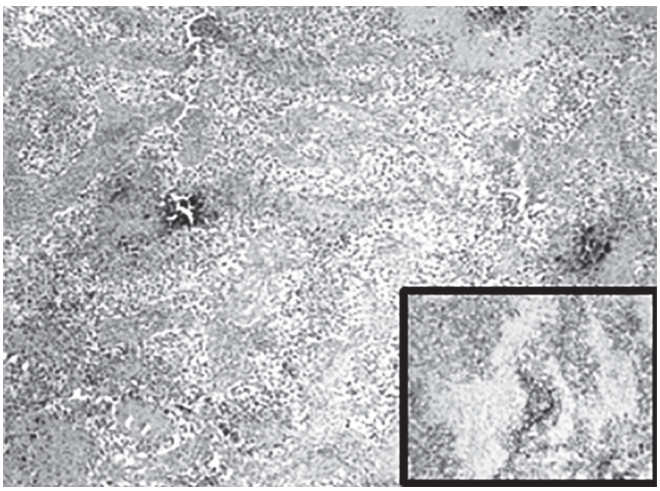


Fig. 8: HPE showing large pleomorphic cells prominent nucleoli suggestive of embryonal carcinoma (inset showing cytokeratin positivity) *100×, H&E

of age, germ cell origin tumors contribute to almost 70% of ovarian tumors and 1/3 of these are malignant.[2] Germ cell malignant ovarian tumors exhibit rapid growth unlike epithelial tumors which are slow growing. Germ cell malignant tumors are characterized by subacute pain related to capsular distension by the tumor and hemorrhage, necrosis within the tumor.[3] Sometimes, they may present acutely, mimicking acute appendicitis or ureteric colic. If the tumor is not diagnosed early, then patients present late with abdominal distension either due to large size of tumor or secondary to ascites. Tumors which are either predominantly solid or with combined solid and cystic features on USG are more in favor of malignant neoplasm.[5]

Dysgerminoma is the most common germ cell tumor, and it accounts for 5–10% of all ovarian tumors in the first two decades of life. They rarely occur after 50 years.[2] The primary treatment is surgery. Chemotherapy is given to patients with metastatic disease.[2] Even though dysgerminoma is very sensitive to radiation therapy, it is not used as it leads to fertility loss.

Immature teratoma is the second most common germ cell malignancy, accounting for about 10–20% of all ovarian malignancy in women during their first two decades. These tumors may occur in combination with other germ cell tumors as well. For adolescents in whom lesion is confined to one ovary, unilateral oophorectomy and surgical staging is done. Patients with IA grade one tumors require no adjuvant therapy and their prognosis is excellent. Patients with stage IA grades two or three, require adjuvant chemo. Patients with ascites will require chemotherapy regardless of tumor grade. The standard approach is bleomycin, etoposide, cisplatin (BEP) regimen.

Mixed germ cell malignancy contains two or more germ cell tumor elements. They mostly consist of dysgerminoma in combination with endodermal sinus tumor (EST), immature teratoma, embryonal carcinoma and choriocarcinoma. The primary tumor size and relative size of the malignant component are the most important prognostic features. The survival rate is 100% for stage IA tumor which are smaller than 10 cm. Tumor markers such as AFP, hCG may or may not be secreted depending on the constituents of the tumor.[2]

Granulosa cell tumors are low grade malignancies that secrete estrogens. It is seen in women of all age groups and they are rarely bilateral. Pre pubertal incidence is 5% and as they secrete estrogen, patients present with sexual pseudo precocity. The treatment for children with stage IA tumors is unilateral salpingo-oophorectomy. There is no evidence to support radiation therapy. BEP is given for stage III or IV or recurrent disease. When compared with adult counterpart, the behavior of juvenile granulosa cell tumor is less aggressive. Majority of the tumors (about 90%) are diagnosed at stage I and associated with good prognosis.[2]

Borderline ovarian tumor mostly occurs in young patients and are FIGO stage I. Ovarian epithelial cancers are so rare in the first two decades, that only less than 1% of such cancers are seen in this age group. The main treatment for these tumors is surgical resection. Adjuvant chemo-radiotherapy has no proven role. The borderline ovarian tumors may be serous, mucinous or other less common histological types.[6]

The five year survival rate for women with stage I borderline tumor is about 95–97%. In case of advanced borderline ovarian tumor the survival rate is 85.5%.

BRCA 1 and BRCA 2 mutations are lower in early borderline ovarian tumor (4.3% of patients) when compared with early stage ovarian cancer (24.2% of patients). Tumor marker CA-125 is of little role in follow-up as it is negative in 76% of recurrent borderline ovarian tumors.[6]

Regular follow-up is necessary for detecting recurrence. The recurrence rate in patients treated for borderline ovarian tumor is 11%. The absolute rate for malignant transformation of borderline ovarian tumor is about 2–4%.[6]

Endodermal sinus tumor is the most malignant ovarian germ cell tumor. Tumor originates from yolk sac hence it is called as yolk sac tumor. They are usually unilateral but rarely bilateral (5% of cases). AFP is the tumor marker which is elevated in EST. There is the good correlation between serum AFP levels and extent of disease. The treatment for EST is unilateral salpingo-oophorectomy and a frozen section for diagnosis. Hysterectomy and contralateral salpingo-oophorectomy does not alter outcome, hence fertility can be preserved. Adjuvant or therapeutic chemotherapy is required for all patients with EST.[2] Survival rate for stage I and II is 60–100% whereas stage III and IV is 50–75%.[7] The progress of the tumor and the development of recurrences can be monitored by serial estimations of serum α-fetoprotein levels.[8]

Embryonal carcinoma is a highly malignant extremely rare germ cell tumor. They may elaborate estrogens and hence the patient can present with precocious puberty. AFP and hCG may be elevated.[7] The treatment of embryonal carcinoma is unilateral oophorectomy with chemotherapy (BEP regimen). The survival rate improved to 60–70% after introduction of chemotherapy and early diagnosis.

Conclusion

This hospital based case series showed that ovarian tumors were quiet common in adolescent girls. Surgical intervention should be directed towards the preservation of ovarian tissue and uterus. However we need bigger studies to show how much fertility preservation is possible. Timely diagnosis will improve survival and preserve future fertility in these young patients.

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