

Ovarian Tumors in the Pediatric Age Group: 37 Cases Treated over An 8-year Period

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Background: Ovarian tumors have generally been considered rare in the pediatric age group. We reported our experience dealing with pediatric ovarian tumors during an 8-year period.

Methods: Between January 1998 and December 2006, 37 girls with ovarian tumors were treated at the Department of Pediatric Surgery, Chang Gung Children's Medical Center. Modes of clinical presentation, pathology diagnosis, methods of treatment and clinical outcome were retrospectively analyzed.

Results: Twenty-nine of the 37 patients were symptomatic with abdominal pain, abdominal distention or the presence of a palpable mass, reduction in appetite or nausea and vomiting and precocious puberty. Another 8 patients were diagnosed prenatally. Thirty patients had benign disease and 7 had malignant tumors. The malignant lesions included 5 germ cell tumors (2 yolk sac tumors, 2 immature teratomas, 1 dysgerminoma), and 2 sex cord stromal tumors. Operations performed were salpingo-oophorectomy (n = 22), oophorectomy (n = 8), cystectomy (n = 3), aspiration (n = 2) and biopsy only (n = 2). A laparoscopic approach was performed in 10 cases. Patients with stage II yolk sac tumors (n = 2) or grade III immature teratomas (n = 2) had elevated alpha-fetoprotein levels, and the patient with dysgerminoma was diagnosed as stage II b. All underwent salpingo-oophorectomy and received chemotherapy following their initial operation and remained free of disease at 8 months to 6 years of follow-up.

Conclusions: In our studies, most ovarian tumors were benign. Epithelial cysts and teratomas were the most common benign lesions, and germ cell tumors were the most common malignancy. A laparoscopic approach was feasible in most cases. With accurate staging, complete resection, and chemotherapy for malignant tumors, patients are expected to have excellent survival rates.

(*Chang Gung Med J* 2010;33:152-6)

Key words: ovarian tumors, pediatric age group

Ovarian tumors are relatively uncommon in children. It has been estimated that such lesions make up 1.5% of all childhood malignancies.⁽¹⁾ Ovarian tumors represent a range of pathologies

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Received: Aug. 11, 2008; Accepted: May 4, 2009

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from highly aggressive malignant tumors to benign cysts. These lesions also may have multiple presentations.⁽²⁾ In this report, we reviewed our experience with these tumors in order to evaluate their clinical presentation, pathology, treatment, and outcome.

METHODS

During the years 1998 to 2006, 37 girls with ovarian tumors were treated at the Department of Pediatric Surgery, Chang Gung Children's Medical Center. The mean age of the patients at the time of diagnosis was 9.8 years, with a range from 2 days to 17 years. All the patients underwent laparoscopy or open laparotomy to confirm the diagnosis.

The clinical presentation, pathology diagnosis, methods of treatment, and clinical outcome were retrospectively evaluated. The length of follow-up ranged from 3 months to 7.5 years. Girls who had malignant tumors were followed up at the oncology department with a range of 8 months to 7.2 years.

RESULTS

In most cases, the presenting signs and symp-

toms included lower abdominal pain, an abdominal palpable mass, abdominal distention, or a combination of these findings.

The clinical presentation of these patients is summarized in Table 1. In the case of two girls, acute abdominal pain and tenderness were noted in the right lower quadrant, resulting in an emergency operation for the presumed diagnosis of appendicitis.

Of the 37 ovarian tumors in this series, 30 patients had benign disease and 7 had malignant tumors (Fig. 1). Of those with benign disease, 9 had

Table 1. Clinical Presentations of the 37 Patients

Clinical presentation	No. of patients (%)
Intermittent abdominal pain	18 (48.6)
Severe abdominal pain	6 (16.2)
Palpable mass, abdominal distention	10 (27.0)
Nausea, vomiting, reduced appetite	6 (16.2)
Precocious puberty	2 (5.4)
Prenatal diagnosis	8 (21.6)

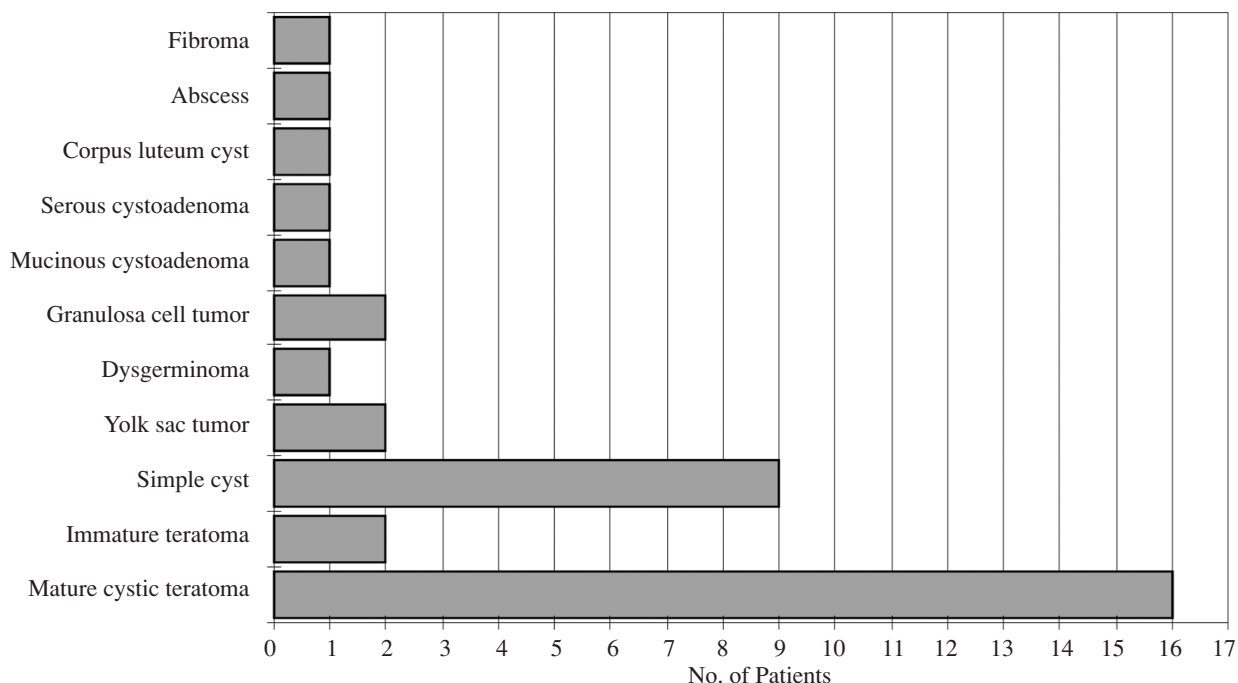


Fig. 1 Summary of the pathologic findings in the 37 patients.

simple or epithelial cysts, 16 mature cystic teratomas, 1 mucinous cystadenoma, 1 papillary serous cystadenoma, 1 corpus luteal cyst with rupture, 1 abscess and 1 fibroma. The malignant lesions included 5 germ cell tumors (2 yolk sac tumors, 2 immature teratomas, 1 dysgerminoma), and 2 sex cord stromal tumors (juvenile granulosa cell tumor).

Ultrasound or computer tomography (CT) scans revealed cystic lesions in 11 patients and all were benign. Combined solid and cystic lesions added some components of calcification in 17 patients (Fig. 2); 15 were benign mature cystic teratomas and 2 were malignant immature teratomas. There were predominantly solid lesions with or without hypervascular enhancement in 6 patients; 5 were malignant and 1 was benign.

Operations performed were salpingo-oophorectomy (n = 22), oophorectomy (n = 9), cystectomy (n = 3), aspiration (n = 2) and biopsy only (n = 2). Twelve patients had an incidental appendectomy. A laparoscopic approach was performed in 10 cases. Patients with stage II yolk sac tumors (n = 2) and grade III immature teratomas (n = 2) had elevated alpha-fetoprotein (AFP) levels (from 1323 ng/ml to 11450 ng/ml), and the patient with dysgerminoma was diagnosed as stage II b. All underwent salpingo-oophorectomy and received chemotherapy (cisplatin, etoposide, bleomycin and vinblastine) following their initial operation. All remained free of disease at 8 months to 6 years of follow-up.

DISCUSSION

Ovarian tumors are rarely diagnosed in the pedi-

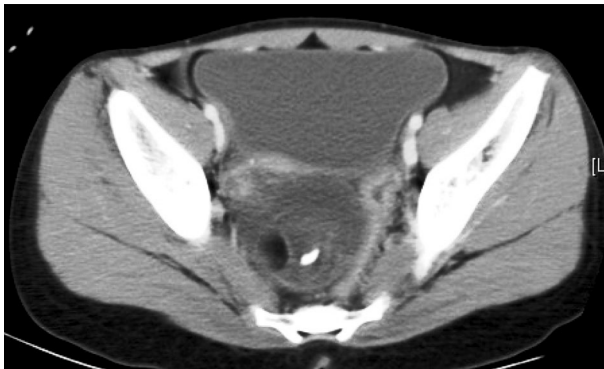


Fig. 2 Enhanced computed tomography scan revealing partly solid and partly cystic components with calcification within the tumor occupying the pelvic region.

atric population, and represent approximately 1.5% of childhood malignancies.⁽¹⁾ The symptoms are often insidious and commonly the tumor is quite large by the time the diagnosis is finally determined. At times, an ovarian tumor may be discovered when a patient undergoes an operation for symptoms consistent with appendicitis. Such symptoms are more likely to be associated with a simple ovarian cyst, cystic teratoma torsion, or corpus luteal cyst rupture. Ultrasonography may be helpful in differentiating non-operative ovarian pathology from appendicitis and other acute surgical conditions.^(2,3) Only two girls had this mode of presentation in our series.

Ovarian tumor cell lines develop embryologically from cells derived from stromal elements of the urogenital ridge, the germinal epithelium covering the urogenital ridge, and germ cells that arise from the yolk sac.⁽⁴⁾ Cells from each of these lines may become subsequently transformed and develop into an ovarian neoplasm. In adult women, the vast majority of ovarian tumors are derived from the epithelial line, and adenocarcinomas predominate. In contrast, the germ cells are the most common cells of origin for ovarian neoplasms in the pediatric population.^(5,6) In our series, 80% of the ovarian tumors were germ cell tumors. Epithelial tumors were rare, occurring in only two patients in our series, both teenagers.

Teratomas are the most common germ cell tumors observed in most published series.^(7,8) This subgroup of tumors may be further divided into mature teratomas, which are benign, or immature teratomas, which may be either malignant or benign. Most benign teratomas are composed of mature cells, but 20-25% also contain immature elements, most often neuroepithelium.⁽⁹⁾ According to Norris' grading system, grade III immature teratomas are defined as the presence of numerous neuroepithelial elements occupying four or more low-magnification fields (40X).⁽⁸⁾ In our series, eighteen were teratomas. However, both malignant and benign teratomas can appear identical in ultrasound or CT findings, and, therefore, AFP is required to help distinguish these lesions preoperatively. Patients who had mature cystic teratomas with torsion accepted laparoscopy or open salpingo-oophorectomy. Two patients with grade III immature teratomas had elevated AFP and received salpingo-oophorectomy and multi-agent chemotherapy: cisplatin, etoposide, bleomycin and

vinblastine. All remained free of disease at follow-up.

Other malignant germ cell tumors include dysgerminomas, yolk sac tumors, and choriocarcinomas.⁽¹⁰⁾ There were two patients with yolk sac tumors and one patient with dysgerminoma IIb in our series. On imaging studies, these tumors appear predominantly solid. Treatment for all childhood ovarian malignancies involves salpingo-oophorectomy.⁽¹¹⁾ Complete staging with omentectomy, lymph node sampling, and peritoneal washing is recommended. For patients with malignant germ cell tumors, cisplatin-based chemotherapy is very effective. The 100% survival rate in our series may be attributed to the multimodality care and treatment.

Although the true incidence of ovarian cysts in fetuses is unknown, they have been seen in as many as 3% to 7% of routine obstetric ultrasound scans.^(12,13) Most of these cysts resolve, which may explain why only 8 patients had operations in our series. Occasionally, these cysts are further complicated by intracystic hemorrhages, ovarian torsion, or rarely, by a mass effect and respiratory distress or hydronephrosis. In general, operations have been advocated only for those cysts that increase in size or persist for more than 4 months after birth.⁽¹²⁾ Although we only had 8 patients that required an operation in this series, the high rate of ovarian torsion suggests an early operation. Laparoscopy to evaluate for torsion with oophorectomy and scope-guided cyst aspiration to improve ovarian preservation were carried out in 6 of our series.

In conclusion, ovarian lesions in children include a broad array of pathologic diagnoses that have variable and nonspecific clinical presentations. Most ovarian tumors are benign.⁽¹⁴⁾ Epithelial cysts and teratomas are the most common benign lesions and germ cell tumors are the most common malignancy.⁽¹⁴⁾ A laparoscopic approach is feasible in most cases. With accurate staging, complete resection, and

chemotherapy for malignant tumors, patients are expected to have excellent survival rates.

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兒童時期的卵巢腫瘤——八年期間的三十七病例回溯及探討

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背景： 分析卵巢腫瘤的病例在兒童時期都是相當罕見。我們藉由過去八年的經驗，試圖去尋找一個最佳的診斷及治療策略。

方法： 我們整理了長庚兒童醫學中心外科部 1998 年一月至 2006 年十二月間，總共有 37 個女童被診斷有卵巢腫瘤，我們採用病歷回溯方式分析這些病童的臨床表現特徵。

結果： 分析這些病童的臨床表徵，有 29 例最主要的表現是腹痛，腹脹或是摸到腹部腫塊，食慾減退或是產生噁心或是嘔吐症狀，或是性早熟。而另 8 例是出生前就已經被偵測到腫瘤。這 37 個卵巢腫瘤病例中，接受手術治療後，病理報告其中有 30 位是屬良性的卵巢，而另外 7 個病例是屬於惡性的卵巢腫瘤。屬於惡性卵巢腫瘤的 7 例中，有 5 例是胚胎細胞瘤 (Germal cell tumors)；其 2 例是卵黃囊瘤 (yolk sac tumor)，2 例是不成熟型畸胎瘤 (immature teratoma)，1 例是惡性胚胎瘤 (dysgerminoma)，而另 2 例是性索間質腫瘤 (sex cord stromal tumors)。手術施行的方法分別是輸卵管卵巢切除術 (n = 22)，卵巢切除術 (n = 8)，囊腫切除術 (n = 3)，細針抽吸術 (n = 2) 以及單獨切片而已 (n = 2)。有 10 個病人是接受了腹腔內視鏡的手術。在胚胎細胞瘤中的 5 位病童，分別是卵黃囊瘤 stage II (n = 2) 或不成熟型畸胎瘤 grade III (n = 2)，她們都有升高的胎兒蛋白 (Alpha-fetoprotein, AFP)，而另一位病童是惡性胚胎瘤 stage IIb。所有診斷為惡性卵巢瘤的病童都接受了輸卵管卵巢切除術以及在手術後接受化學治療，然後在隨後追蹤的八個月到六年間完全康復。

結論： 在我們的病例中，其實大部分的兒童時期卵巢腫瘤都是良性的，表皮囊腫或是畸胎瘤是最常見的。腹腔內視鏡的手術對大部分病例是合適的選擇。而卵巢腫瘤中胚胎細胞瘤是最常見的惡性腫瘤；針對較嚴重的病人當給予合適的腫瘤分級、完整的切除，以及妥適的術後化學治療，仍然可以預期會有不錯的預後及存活。

(長庚醫誌 2010;33:152-6)

關鍵詞： 卵巢腫瘤，兒童時期