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Management of ovarian tumors and adnexal torsion in children; how to save?

Çocuklarda over tümörlerinin ve adneks torsiyonlarının sağaltımı; nasıl kurtarabiliriz?

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Abstract

Introduction: Although rare, torsion and malignancy of the uterine adnexa, are a common concern of differential diagnosis while dealing with abdominal pain in girls.

Methods: Forty-seven patients who were operated for adnexal torsion or ovarian tumor between March 2004 and December 2010 are enrolled in the study.

Results: Twenty one patients had torsion of the uterine adnexa, 18 had ovarian tumors and 8 had an ovarian tumor with torsion. The rate of radical surgery was 47% in torsion which was elevated to 72% in tumor and 100% in tumors with torsion. In ovarian torsion, the rate of radical surgery and adnexal sparing were not different between early versus late presenting cases.

Discussion and Conclusion: Despite encouraging data in the literature favor ovarian sparing, radical surgery still seems to remain high in girls with adnexal torsion and tumors. Ovarian sparing seems possible in tumors but further studies are needed to determine the best management strategy.

Keywords: Child; ovary; torsion; tumor.

Öz

Amaç: Nadir görülmekle birlikte, uterin adnekslerin malignite ve torsiyonları, karın ağrısı ile başvuran kız çocuklarının değerlendirilmesinde önemli bir sorun oluşturmaktadır.

Gereç ve Yöntem: Kliniğimizde, Mart 2004 ile Aralık 2010 tarihleri arasında adneks torsiyonu veya tümörü tanısı almış 47 hasta çalışmaya dahil edildi.

Bulgular: Yirmi bir hasta uterin adnekslerin torsiyonu, 18 hasta over tümörü ve 8 hastanın ise tümör ile birlikte torsiyon nedeniyle ameliyat edilmişti. Radikal cerrahi oranı torsiyonlarda %47, tümörlerde %72 iken, hem torsiyon hemde tümör varlığında %100'e ulaşmaktaydı. Over torsiyonlarında radikal cerrahi ve over kurtarılma oranları açısından, erken ve geç başvuran vakalar arasında fark yoktu.

Sonuç: Overlerin korunabileceğini destekleyen verilerin giderek artmasına rağmen, kız çocuklarında adneks torsiyonu ve tümörlerinin tedavisinde radikal cerrahi oranları halen yüksek gözükmektedir. Over tümörlerinde koruyucu cerrahi mümkün gözükmekle beraber en uygun yolunu bulmak için daha ileri çalışmalara ihtiyaç vardır.

Anahtar Sözcükler: Çocuk; over; torsiyon; tümör.

Torsion of the uterine adnexa and ovarian tumors are uncommon in children.[1–3] Although their frequency is low, these conditions may be misdiagnosed in daily practice and should be considered during the evaluation of abdominal pain as most patients admit with non-specific complaints such as abdominal pain, abdominal discomfort or urinary symptoms. Some of these conditions are managed in emergency conditions and the surgeon may sometimes have to make clinical decision without sufficient clinical data. The concern to miss a malignancy and to avoid morbidity or complication should be dissolved without compromising fertility of a child with a life expectancy of at least 50 years. Another issue is, as some of the ovarian tumors may cause adnexal torsion, the engorged and twisted adnexa may resemble a solid mass, which complicates the decision of extent of surgical approach as well (radical or conservative).[2] Although ovarian sparing surgery is encouraged in recent decades, reflection to the daily practice is poor as radical surgery rates did not decrease significantly and clinical practice varies widely.[2,4,5] The aim of this study is to evaluate adnexal torsion and ovarian tumors in children and find out if the rate of adnexal loss can be reduced avoiding over- or under-treatment without compromising clinical outcome.

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Table 1. Salvage rates in early vs. late cases of adnexal torsion

Intervention interval	Radical	Conservative	Control US	Normal sized ovary	Follicular activity	Salvage rate
>36 hours	9 (53%)	8 (47%)	7	2	3	71%
<36 hours	8 (67%)	4 (33%)	3	1	2	100%

Table 2. Sensitivity and specifity of gray scale and Doppler US

	Gray scale US	Doppler US
Sensitivity	55.17% [%95 CI (0.3755–0.7159)]	58.33% [%95 CI (0.3195–0.8067)]
Specificity	93.94% [%95 CI (0.8543–0.9762)]	91.67% [%95 CI (0.6461–0.9851)]

CI: Confidence interval.

Materials and Method

With institutional review board approval (#30/09/2010-2-21), the charts of the female patients younger than 18 years old who underwent surgery for adnexal conditions between March 2004 and December 2010, were retrospectively reviewed. Forty-seven patients with adnexal torsion or ovarian tumor whose operative charts and follow up data are available, are enrolled in the study. Infants younger than 2 months old are excluded as all of them are regarded as intrauterin torsion which is beyond the scope of this paper. Clinical, radiological and laboratory findings were evaluated.

Descriptive statistics were given as mean±standard deviation. Categorical variables were given as number and percentage. Age of patients was compared with the Mann-Whitney U test. Surgery types,the presence of torsion and malignancy were compared with Chi-square and Fisher's exact test. ROC analysis was performed to determine the tumor size with maximal sensitivity and specifity to detect malignancy. Statistical significance was accepted as $p<0.05$. Statistical analyses were performed using jamovi (Computer Software Version 0.9 2018 www.jamovi.org) ROC analysis with was performed using MedCalc Statistical Software version 15.8 (Medcalc Software, Ostend, Belgium (https://www.medcalc.org;2015).

Results

Among the 47 patients, adnexal torsion was encountered in 21 patients, tumor in 18 and both conditions in 8.

Adnexal torsion was identified in 29 patients in whom 24 had received urgent operation. The right side was affected in 22 (%76) patients while left side in 7 (%24). One patient (3%) had isolated tubal torsion and had undergone salpingectomy. No synchronous or asynchronous bilateral torsion or recurrent torsion was encountered during the study period. Median age at operation was 10 years (range 3-16 years). Mean follow-up was 5,5 months (1-56 months) .

Among the patients with torsion, histological examination revealed 7 mature and 1 immature teratomas (grade 3, stage 1). Urgent operation is performed in 5 of these patients which were all mature teratomas while other 3 were operated electively. When simple cysts are excluded, an underlying tumor was found in 8 (28%) patients with adnexal torsion, of which 1 (3%) was malignant (7 mature, 1 immature teratoma). In another point of view, a total of 13 teratomas (2 immature, 11 mature) were identified in our series of which 8 presented with torsion (62%).

Of 29 patients, 17 (59%) patients underwent radical surgery (oophorectomy, salpingectomy or salpingo-oophorectomy). Indications for radical surgery mostly depended on the clinical picture and appearance of the adnexa during surgery. Radical surgery was decided in 7 as the torsed mass resembled a tumor and in 11 due to ischemic appearance that had no chance to survive. No tumor was identified in 2 of these 7 patients with suspected tumor while 3 tumors are identified in those 11 ovaries excised for ischemic appearance. The overall justification error rate was 28%.

Ovarian sparing surgery was performed in 12 patients (41%). In 10 of these patients ultrasonographic examination could be obtained during follow up. Follicular activity has been demonstrated in 5 of these patients and normal sized ovaries in 3. Overall salvage rate of conservative surgery was 80%. The time lag between onset of symptoms and surgery is defined as "intervention interval". Patients are classified as "early" and "late" when the intervention interval was lower or higher than 36 hours, respectively. Although salvage rate seems to be higher in early cases (100% vs. 71%), radical surgery was performed more frequently in early cases (67% vs. 53%) (Table 1).

Mean age of patients with torsion were not different from the patients without torsion (9.55 ±3.57 years vs. 9.41±4.42 years,respectively) (p=0.956).

All the patients had undergone gray scale ultrasound (US) examination but only 14 had Doppler US before the operation. Although specificity for both modalities was high, sensitivity was significantly low. Both sensitivity and specificity of both modality was similar (Table 2).

In total, 27 ovaries of 26 patients had tumor and 8 patients had co-existing torsion. Although the preoperative diagnosis was tumor, an urgent operation has been undertaken in

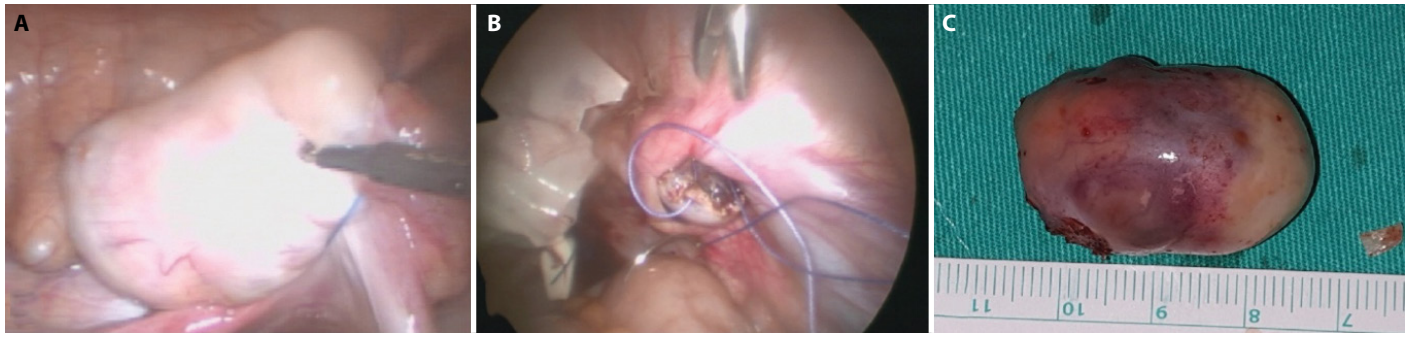

Figure 1. Mature teratoma of the right ovary **(A)**. Ovary spared and fixed to the lateral pelvic wall **(B)**. Teratoma excised and extracted intact **(C)**.

7 patients to exclude an acute condition as all of them admitted with abdominal pain. In 5 of these 7 patients, adnexal torsion was encountered and radical surgery was performed for ischemic appearance in 2 and for tumor-like appearance in 3. Histological examination revealed mature teratoma in all 5 patients who underwent urgent and radical surgery. Cystadenoma was the final diagnosis in those 2 who had ovarian sparing surgery. Three patients with tumor and torsion were operated electively and 2 patients were found to have mature teratoma while the other one had immature teratoma. These patients were operated electively as their clinical picture was predominantly indicating a malignancy rather than an acute condition.

Most common tumor was mature teratoma (n=11), followed by cystadenoma (n=5), immature teratoma (n=2), yolk sac tumor (n=2), non-Hodgkin lymphoma (n=2), dysgerminoma (n=2), mixed germ cell tumor (n=1) and cavernous hemangioma (n=1). Nine of the 26 patients had a malignant tumor (35%).

Mean size of the malignant lesions were 17±7.18 cm (median =20, range 2-25 cm).

Mean size of the benign lesions were 10.1+7.84 cm (median=8, range 1-40 cm).

Mean age of patients was not different between malignant and non-malignant cases (8.72±3.33 vs. 9.68±4.00 years, respectively) (p=0.424).

Regardless of size, none of the pure cystic tumors were malignant.

Using the ROC analysis, it was found that a cut-off value of 10 cm has 88.9% sensitivity and 67.7% specificity (AUC: 0.753, p<0.0157) for detecting malignancy.

Conservative surgery was performed in 4 (18%) patients while radical surgery was performed in 22 (82%). Histological studies revealed cystadenoma in 3 and mature teratoma in one of the patients who underwent conservative surgery (Figure 1). No relapse was encountered during 6 months follow up in these patients. In four patients that ovaries were spared, follow up US revealed follicular activity in 3 and the ovarian size was normal in 1 patient.

The involved ovary was right in 15 cases (56%) and left in 10 (37%). In one case of Non-Hodgkin lymphoma (NHL) (7%), both ovaries were involved and the patient had deceased 9 months after diagnosis despite aggressive chemotherapy. No other death was observed during a median of 6.6 months (0.1-5 years) follow up period.

All ovarian malignancies in our series had elevated one or more of the tumor markers (AFP, β-hCG or LDH).

AFP levels was found high in all patients with immature teratoma and yolk sac tumor while β-hCG was high in all patients with dysgerminoma and mixed germ cell tumors (Table 3). Mild elevation of CA-125 was found in one patient with dysgerminoma and in one patient with yolk sac tumor which was not considered a useful marker for ovarian tumors in children. Isolated elevations in LDH were encountered in 2 patients with ovarian NHL, which may be considered as an indicator of extra-adnexal origin if other markers are negative.

In our series, 8 patients with torsion also had a tumor and 5 of them had undergone urgent operation. One patient who was operated electively, found to have immature teratoma. The rest 7 patients had mature teratoma. All of these 8 patients received radical surgery (OO or SPO). In our series, teratoma was the only underlying neoplastic condition patients with adnexal torsion, with only one being malignant which was cured with surgery alone. In other words, 64% (7/11) of the mature teratomas and 50% (1/2) of the immature teratomas had co-existing torsion.

Presenting with painless abdominal mass was associated with malignancy when compared with other symptoms (6/11 (54.5%) versus 3/36 (8.3%), respectively) (p<0.001). Presenting with painless abdominal pain had 2.2 (1.05-3.88, 95 %CI) times relative risk for having malignancy.

Radical surgery rates were 47% (10/21) in patients with torsion, 72% (13/18) with tumor and 100% (8/8) with torsion and tumor; and was found significantly different among groups (p=0.019).

Discussion

Although relatively rare in children, torsion and tumors of the uterine adnexa are a major cause of ovarian loss and must be considered in girls presenting with common complaints such as abdominal pain and distension. Some patients can have both tumor and torsion which makes the management

process more complicated.[3] Despite increasing evidence encouraging ovarian sparing surgery, the frequency of radical surgery still remains high.[5,6] Although some authors reported better results with earlier intervention in ovarian torsion, we did not find a significant difference and think that conservative surgery is still possible in late cases as suggested by many others.[4,7] Although there is no clear evidence on the long-term side effects of unilateral oophorectomy, some authors related it with early menopause and its' consequences.[8,9] Theoretically, reduced number of ovary and thus oocyte reserve, may have a negative impact on fertility. Although theoretical complications of ovarian sparing surgery are reported as thromboembolic events, peritonitis due to infarcted adnexa and the possibility of missing a malignancy, many reports did not encounter any thromboembolic event or peritonitis related with ovarian sparing surgery.[6,10,11] As ovarian malignancy is rare in children and survival is not compromised with ovarian sparing surgery and even after deviations from the surgical guidelines, fear of missing a malignancy is likely to be managed with close surveillance.[8,12,13] Some authors evaluate the effect of surgical specialty on the rate of ovarian sparing surgery and found that conservative surgery is performed more frequently by the pediatric gynecologists than pediatric surgeons although the rate of malignant lesions was higher in patients managed by pediatric surgeons.[14]

As significant rate of mature teratoma is reported to present with complications as in our series, surgical excision is recommended when detected, even if asymptomatic.[12,15,16]

We think that the negative tumor markers should encourage ovarian sparing surgery as the likelihood of malignancy is significantly low.[17] Although malignanant potential of heterogenous lesions is hard to define, pure cystic lesions should always be managed conservatively as cystadenomas are the most neoplastic conditions in such cases.[13]

Although not adequate, the ovarian sparing rate is higher in ovarian torsions than ovarian tumors and it was not possible if the patient had a torsed tumor. The indications for radical surgery in our study were, malignant appearance, the tumor-like appearance of the torsed mass or ischemic appearance of the torsed adnexa. When the intraoperative diagnosis is compared with histopathological diagnosis, in 5 of 7 patients with tumor suspicion and 3 of the 11 patients who were excised for ischemic appearance, a tumor is found. This may mean a 28% justification error rate. The question is, if this justification error would be solved, could the ovaries still be spared with clear surgical margin without detrimental effect on long-term survival. Another issue is the unclear behavior of the adnexa after torsion/detorsion process; will it end up with necrosis and cause future problems, will it survive or will it rupture and cause dissemination of a co-existing tumor.

As reported by others,the sensitivity of ultrasonography (Gray Scale or Doppler) for detecting ovarin torsion was found low.[10]

In conclusion, ovarian tumors and torsion can be encountered at any age and should be considered in the differential diagnosis of girls presenting with abdominopelvic symptoms. Tumor markers (AFP, β-hCG, LDH) should be obtained if possible for any case when an adnexal condition is possible. In cases of elevated levels of AFP, β-hCG, and maybe LDH, a malignancy is highly possible and ovarian sparing surgery should be undertaken very cautiously if deemed necessary (i.e. solitary ovary or bilateral cases etc.) without hesitation to convert to open surgery and obtaining frozen sections.

In an unprepared scenario, such as the patient underwent an operation without tumor markers obtained and found to have adnexal torsion with/without the possibility of a tumor, we recommend simply untwisting without disturbing the torsed mass (biopsy etc.) as most of them are benign. Tumor markers are obtained during or immediately after the operation and a second look operation may be indicated in accordance with tumor markers and close surveillance (clinical and radiological).

As no malignancy is encountered in pure cystic lesions, ovarian sparing surgery is strongly recommended. Significant number of teratomas (benign or malign) present with torsion, they should be excised as soon as possible when detected, independent of their size and whether they are symptomatic or not. Small sample size and retrospective design are the major limitations of our study. Further studies are needed to increase the ovarian salvage rates in adnexal torsion and especially ovarian tumors in children.

Conflict of interest: There are no relevant conflicts of interest to disclose.

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