Original Research Article

Our experience with adnexal masses in the pediatric age group and review of literature

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Adnexal masses; Pediatric; Benign; Malignant; Torsion

Abstract
Background and objectives: Adnexal masses are rare in the pediatric age group. We present our experience with 20 patients with adnexal masses.

Design and setting: This retrospective observational analysis was performed on 20 children with adnexal masses who were treated at our institute between May 2011 and November 2015.

Patients and methods: Fifteen pediatric patients who were admitted between May 2011 and November 2015 were reviewed and retrospectively analyzed based on their age at the time of admission, their presenting complaints, clinical and radiological findings, tumor markers, management and follow-up.

Results: The patients’ age at the time of admission ranged between 3 days and 12 years. Abdominal pain and lump were the most common presenting complaints.

Four patients (20%) had antenatally diagnosed cystic ovarian lesions. On postnatal scan, 2 patients had a simple cyst measuring less than 6 cm, which resolved on follow-up ultrasound at 3 months. One neonate had a simple cyst, larger than 6 cm on postnatal scan, which was managed by marsupialization. One antenatally diagnosed patient had a dermoid cyst that required oophorectomy.

Ten patients (50%) had dermoid cyst and underwent complete surgical excision of the mass. Based on histopathologic results, two of these patients had immature teratoma and required adjuvant chemotherapy (Bleomycin, Etoposide, and Cisplatin). The serum AFP levels of these patients were carefully monitored.

One patient with bilateral ovarian cysts was diagnosed with Van Wyk–Grumbach syndrome, which resolved significantly after a 3-month regimen of thyroxin supplementation.

Five patients presented with torsion and required emergency surgery—three had mature teratoma, one had an immature teratoma and one had large simple cysts.

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1. Introduction

Ovarian masses are most commonly observed in adults; they rarely occur in children. The majority of the ovarian masses encountered in children or patients of premenarchal age are non-neoplastic lesions. The clinical signs and symptoms of ovarian masses are usually non-specific. Early gynecological management is necessary to preserve fertility. Gynecological malignant conditions constitute approximately 3% of all types of cancer in children. Ovarian tumors in children account for only 1% of childhood malignancies. However, the true incidence of malignant ovarian tumors in the pediatric population is unknown [1].

2. Materials and methods

The records of 20 girls under the age of 12 years with adnexal masses who were treated at a tertiary referral center between May 2011 and November 2015 were reviewed and retrospectively analyzed based on their age at the time of admission, presenting complaints, clinical and radiological findings, tumor markers, management and follow-up.

All of the patients underwent pelvic ultrasound. CT scan was performed when necessary. Tumor markers—serum alpha fetoprotein (AFP), beta-human chorionic gonadotropin (β-HCG), and cancer antigen 125 (CA-125), were tested in ten patients. A thyroid function test was conducted in one patient because of associated precocious puberty. Complex lesions were surgically excised. A standard Pfannenstiel incision was made to remove the lesions. One patient was operated on laparoscopically. The patients with immature teratoma underwent complete surgical excision of tumor followed by chemotherapy; Bleomycin, Etoposide and Cisplatin (BEP regimen) were administered, and the patients’ serum AFP levels were monitored. The diagnoses of all patients were histopathologically confirmed. Simple cysts measuring less than 6 cm were managed conservatively using ultrasound scans performed monthly for 3 months.

3. Results

The patients’ age at the time of admission ranged between 3 days and 12 years. Four neonates were antenatally diagnosed, as confirmed by postnatal ultrasound scans. Abdominal lump (Fig. 1) (n = 12) and pain (n = 11) were the most common presenting complaints. Lesions were unilateral in 14 patients and bilateral in 1 patient. Tumor markers were sent in 10 patients, and the results were within normal limits.

Four patients (20%) had antenatally diagnosed unilateral cystic ovarian lesions. On postnatal scan, 2 patients had a simple cyst measuring less than 6 cm, which was conservatively managed. These lesions resolved on follow-up ultrasound at 3 months. In one patient, a cyst measuring more than 6 cm was marsupialized. One antenatally diagnosed patient had a dermoid cyst and required oophorectomy.

Ten patients (50%) underwent complete surgical excision of the adnexal mass (Figs. 2–5). Oophorectomy was performed in eight patients; based on histopathologic results, these patients had mature teratoma, and they required no additional treatment. Two patients were managed by salpingo-oophorectomy; histopathology revealed immature teratoma. The patients received adjuvant chemotherapy; Bleomycin, Etoposide and Cisplatin (BEP regimen) were administered and the patients’ serum AFP levels were monitored. One patient had a large paraovarian cyst, which was laparoscopically excised (Figs. 6 and 7).

One patient (5%) presented with bilateral multicystic ovarian masses and precocious puberty and had severe hypothyroidism on hormonal evaluation. She was diagnosed with Van Wyk—Grumbach syndrome and was administered thyroxin supplementation. The masses resolved significantly after 3 months.

Five patients (25%) presented with acute abdomen and were diagnosed as torsion on USG and CT requiring surgery.

Fig. 1 Clinical photograph of an adnexal mass presenting as abdominal lump.
emergency surgery. All of the patients had adnexal masses measuring greater than 8 cm, in addition to gangrene observed in the ipsilateral ovary. Three patients were managed by salpingo-oophorectomy and two required oophorectomy. Histopathologic results revealed teratoma in three patients, immature teratoma in one patient and simple cysts in one patient. The patient with immature teratoma received adjuvant chemotherapy (BEP regimen) and the patient’s serum AFP levels were monitored.

All of the patients had no complaints on follow-up (Table 1).

4. Discussion

Adnexal masses may originate from the ovaries, fallopian tubes and other pelvic organs. The types of adnexal masses include tumors, inflammatory or functional cysts. Approximately one third of adnexal masses are ovarian tumors.

Malignant ovarian tumors are rare, particularly in patients under 5 years of age. Solid ovarian tumors are uncommon in the pediatric population; however, ovarian tumors, when present, constitute a major source of anxiety for the patients and their family. Ovarian tumors must be considered in the differential diagnosis of young girls with abdominal pain, mass or other non-specific symptoms. In a large study, Templeman et al stated that malignant ovarian tumors in children and adolescents are rare, accounting for 0.9% of all malignancies in this age group [2]. Hassan et al confirmed that during the first two decades of life, ovarian tumors represent the most frequent tumors found in the female genital tract [3].

Fig. 2 CT image showing large left adnexal mass.

Fig. 3 Intraoperative image of a left immature teratoma.

Fig. 4 Excised specimen of the left immature teratoma.

Fig. 5 Excised specimen of the left mature teratoma.
palpable abdominal mass was the most frequent physical finding.

Primary ovarian cysts and tumors are uncommon in children. Two-thirds of malignant tumors in children are germ cell tumors. The majority of the malignant germ cell tumors are dysgerminoma, in contrast to adult ovarian tumors, where 90% are of epithelial cell origin and approximately 10% are non-epithelial. In our study, the most common malignant tumor was immature teratoma, and none of the patients had dysgerminoma.

Although the true incidence of ovarian cysts in the fetus is unknown, they have been reported in 3%–7% of routine obstetric ultrasound analyses [4,5]. Most of these cysts were resolved, which may explain why no antenatally detected ovarian cyst in our series required surgery.

Follicular cysts are commonly detected incidentally on antenatal ultrasound examination [6]. The etiology is unclear, but they most likely arise from ovarian stimulation by maternal and fetal gonadotropin [7]. The majority of fetal ovarian cysts are unilateral, although both ovaries may be involved.

Follicular ovarian cysts in fetuses and neonates are common and increase in frequency with advancing gestational age and some maternal complications, such as diabetes mellitus, preeclampsia, and rhesus isoimmunization [7,8]. In one autopsy series of 332 ovaries from stillbirths and neonatal deaths, one or more follicular cysts lined by granulosa epithelium and measuring greater than 1 mm in diameter were detected in 113 infants [8]. Among live births, the best estimate of the incidence of clinically significant ovarian cysts is 1 in 2500 [9].

Occasionally, these cysts are further complicated by intracystic hemorrhages, ovarian torsion, or rarely, by a mass effect and respiratory distress or hydronephrosis. Spontaneous regression usually occurs by 4–6 months of age [10].

The differential diagnosis of a fetal cystic intrainfundibular mass includes genitourinary tract disorders (e.g., reproductive tract anomalies, urinary tract obstruction, urachal cyst), gastrointestinal tract disorders (e.g., mesenteric or omental cyst, volvulus, colonic atresia, intestinal duplication), or miscellaneous disorders (e.g., choledochal, splenic, or pancreatic cyst; lymphangioma). Spontaneous regression of both simple and complex cysts often occurs either antenatally or postpartum by 6 months of age; therefore, the management is usually expectant. In one review of 66 published cases of simple cysts, 50 percent resolved by 1 month of age, 75 percent by 2 months, and 90 percent by 3 months [9]. The rate of malignancy is so low that it need not be considered in making therapeutic decisions. Ultrasound examination should be performed every 3–4 weeks antenatally.

The standard management of neonatal cysts consists of serial ultrasound examinations at birth and every 4–6 weeks thereafter until the cyst resolves, enlarges, has persisted for 4–6 months, or becomes symptomatic. The aspiration of simple cysts measuring ≥ 4–5 cm is advised [7]. Surgical intervention is reserved for complex and symptomatic cysts and for cysts that increase in size and persist for more than 4–6 months [11–13]. The transumbilical approach has been reported to be both a feasible and safe approach for a broad spectrum of surgical
TABLE 1  Table summarizing the presentation and management of 20 patients with adnexal masses.

<table>
<thead>
<tr>
<th>Sub-type/size</th>
<th>Simple cyst</th>
<th>Teratoma</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;6cm</td>
<td>Lump</td>
<td>Torsion</td>
<td>Abdominal lump and pain</td>
</tr>
<tr>
<td>Conservative</td>
<td>2</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>Marsupilisation</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Excision</td>
<td>1</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Oophorectomy</td>
<td>8</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Salpingo-oophorectomy</td>
<td>1</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td>2</td>
<td>8</td>
<td>2</td>
</tr>
</tbody>
</table>

procedures, including ovarian tumors, in neonates and infants. The cosmetic results have been reported to be excellent [14]. The transumbilical approach seems to be an attractive alternative for managing ovarian cysts in children in the absence of appropriate settings for laparoscopy [15].

Teratomas are the most common germ cell tumors observed in the majority of published series [16,17]. This subgroup of tumors may be further divided into mature teratomas, which are benign or immature teratomas, which may be either malignant or benign.

Teratomas are composed of recognizable tissues of ectodermal, mesodermal and endodermal origin, in any combination. Immature teratomas are common germ cell tumors comprising two or more germ cell layers (ecto-, meso- or endoderm) derived from a pluripotent malignant precursor cell. Mature teratomas account for approximately 15% of all ovarian tumors [18] whereas immature tumors are rare, representing less than 1% of ovarian tumors [19]. Mature teratomas are classified as cystic, solid or mixed. Immature teratomas show only solid mass. In mature teratomas, the most commonly mature ectodermal elements such as skin, hair, sweat and sebaceous glands are noted, whereas in immature teratomas, tissues with partial somatic differentiation identical to that of fetal tissues are found. In this study, 7 patients had teratoma; of these, 5 patients (87.5%) had mature teratoma and two patients (13.33%) had immature teratoma.

However, both malignant and benign teratomas can appear to be identical by ultrasound or CT findings. Germ cell tumors are serologically evaluated using tumor markers. In immature teratoma cases, AFP is widely used. It has been suggested that the AFP level in immature teratoma is not correlated to either stage or grade of the tumor. Thus, AFP plays a limited role in the evaluation and management of germ cell tumors [20–23]. In this study, all of the patients underwent surgical resection. Two patients were histopathologically diagnosed with immature teratoma, and they received chemotherapy.

Van Wyk—Grumbach syndrome is a rare condition characterized by breast development, uterine bleeding and multicystic ovaries in the presence of long-standing primary hypothyroidism. The syndrome is characterized by juvenile hypothyroidism, delayed bone age, and isosexual precocious puberty with reversal to a prepubertal state following thyroid hormone replacement therapy [24]. The gonadotropin releasing hormone (GnRH)-dependent activation of the hypothalamic-pituitary-gonadal axis leads to central precocious puberty (CPP). The extrapituitary secretion of gonadotropins or secretion of gonadal steroids independent of pulsatile GnRH stimulation may lead to pseudoprecocious puberty or GnRH-independent sexual precocity [25].

Hypothyroidism should be considered, especially when young girls present with bilateral multicystic ovarian mass and vaginal bleeding accompanied by additional clinical presentations, such as cold intolerance, constipation, delayed bone age. Thyroxin replacement therapy should lead to complete resolution of symptoms and promote normal physical and mental development.

Paravarian cyst is a benign condition that is uncommon in children, is incidentally diagnosed, and occurs in the mesosalpinx between its two leaves. The cyst is believed to originate from mesothelium or the remnant of Mullerian duct and Wolffian duct [26]. Complications such as torsion, hemorrhage, rupture, and neoplastic transformation may occur. The preoperative diagnosis is difficult because of its close proximity to the ovary. Laparoscopy is a diagnostic as well as therapeutic approach.

5. Conclusion

Ovarian lesions in children include a broad array of pathologic diagnoses that have variable clinical presentations. Most ovarian tumors are benign. Epithelial cysts and teratomas are the most common benign lesions and germ cell tumors are most commonly malignant. A laparoscopic approach has been reported to be attractive in the majority of the studies. With accurate staging, complete resection and chemotherapy for malignant tumors, patients have demonstrated excellent survival rates. In patients with bilateral ovarian lesion, endocrine causes should be ruled out.

Ethical clearance

Not applicable as this is a retrospective study. All patients in this study were managed according to the existing protocols. No new investigations and intervention was done on the patient.

Source of funding

Nil.
Conflict of interest

Nil.

References