Mixed ovarian germ cell tumor in a child: A case report of a rare association

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1. Introduction

Ovarian neoplasms are relatively rare in the paediatric population. Their estimated annual incidence does not exceed 2.2 cases per 100,000 girls and only about one-fourth of all ovarian tumors in females younger than 16 years are described to be malignant (27%) [1]. Moreover, ovarian cancer represents 1.1% of all malignant childhood tumors [2]. Histopathologically and in contrast with ovarian cancer distribution in adults, malignant germ cell tumors (GCTs) are more common than epithelial and sex cord stromal neoplasms [3]. According to the literature, their overall prognosis is reported to be excellent [3]. Despite advances in understanding the etiopathogenesis of malignant ovarian GCTs, their etiology is still not well understood [4]. Here, we report the findings of a mixed ovarian GCT in a 13-year-old girl treated in our primary academic care center. This case report has been reported in line with the SCARE Criteria [5].

2. Case presentation

A 13-year-old girl with no significant pathological history was referred by family physician to our department of Paediatric Surgery for swelling associated with a right abdominal flank pain. The patient had no family, drug history or any past surgery nor a particular genetic predisposition to any diseases. Clinical examination found an apyretic child and in a good general condition. On abdominal examination, a right flank tenderness with a solid mass measuring 12 cm was characterized. Additionally, an abdominal ultrasound was performed and showed a poorly limited right para-uterine mass with 100*52 mm in size. Magnetic resonance imaging (MRI) revealed a right ovarian tumor mass measuring 97*47 mm with irregular seams, on iso-signal T1, peripheral hypersignal and central hyposignal T2 in relation with a central necrosis (Fig. 1). Moreover, serological alpha-fetoprotein (aFP) level was elevated (490.68 ng/ml) in contrast with a normal level of Beta-human chorionic gonadotropin (β-hCG) (7.77 mU/ml). All other serum parameters were normal. No diagnostic challenges in our patient...
could be reported. An exploratory laparotomy by a pfannenstiel incision was performed. Intraoperative findings showed a huge mass originating from the right ovary. A right adenexectomy, biopsies of the uterus and a peritoneal fluid sample were carried out (Fig. 2). The whole surgical intervention was performed by a senior paediatric surgeon with the aid of an assistant surgeon and two resident doctors. The postoperative time was uneventful with no hemorrhagic or infective complications. The diagnosis of a mixed ovarian germ cell tumor comprising dysgerminoma, as predominant component, and yolk sac tumor as minor component, was made based on the histological appearance, elevated serological level of αFP and immunohistochemical results (Fig. 3). Uterine biopsies and peritoneal fluid sample were found clean from any malignancy. The patient received adjuvant chemotherapy using a combination of vinblastine, bleomycin and cisplatin 21 days apart after the normalisation of αFP in serum. An MRI control was performed and showed no recurrence or tumor residus after six months of follow up (Fig. 4). Also, no negative incidents in terms of adherence and tolerability were observed. Taken together, the evolution of the disease in our patient, our management, and follow-up are summerized in the timeline of Table 1.

3. Discussion

In children and adolescents, GCTs represent the most common histological type of ovarian tumors and are benign in the majority of cases \[6,7\]. According to the World Health Organisation (WHO), ovarian GCTs are classified into many histological subtypes including dysgerminoma, yolk sac tumors, embryonal carcinoma, polyembryoma, choriocarcinoma, teratomas and mixed GCTs \[8\]. The term mixed GCTs

![Fig. 1.](image1.png)  
**Fig. 1.** a. Coronal MRI view showing a right ovarian tumor with irregular seams. Peripheral hypersignal and central hyposignal T2 are in relation with a central necrosis. b. Sagittal MRI view showing bladder compression by the tumor. c. Transverse MRI views showing bladder compression by the tumor. 

![Fig. 2.](image2.png)  
**Fig. 2.** Surgical specimen of right adnexectomy with a peritoneal nodule.
is applied to neoplasm containing a combination of malignant germ cell elements. Dysgerminoma followed by yolk sac tumor, which secretes aFP, are the most common subtypes of the ovarian GCTs and particularly concern patients in their second or third decades of life [8]. In our patient, the pathological examination of the resected tumor revealed a combination of these two types.

The clinical presentation of patients with ovarian GCTs can be similar in both benign and malignant forms; the most common signs are the presence of a mass of the lower abdomen and abdominal pain which is usually chronic [9]. This neoplasm is generally asymptomatic or minimally symptomatic as long as it does not reach a considerable size and without compression of the adjacent organs. Vaginal bleeding, constipation or amenorrhea can also be reveling signs but less frequently. In other cases, a complication such as the rupture of the

Fig. 3. a. The predominant component (dysgerminoma) b. The minor component (Yolk sac tumor) c. Diffuse cytoplasmic expression of CD117 by tumor cells of the dysgerminoma component d. Cytoplasmic expression of Cytokeratine by tumor cells of yolk sac tumor component.

Fig. 4. a. transverse and b. Sagittal MRI control after 6 months not showing any recurrence or tumor residue.
neoplasm can contain calcifications, hemorrhagic foci or necrotic areas. Doppler ultrasound with an intensive flow signal [11]. In addition, the signal voids. The tumor releases vascular properties through striking contrast enhancement and multiple components with areas of necrosis and hemorrhage [16]. The MRI voluminous mass with heterogeneous composition: cystic and solid area is seen. The MRI appearance of the tumor can reveal predominancy of the solid component contrasting with some areas of cysts and hemorrhage, and hypervascular properties through striking contrast enhancement and multiple signal voids. The tumor releases α-FP in serum and its levels can be used for diagnostic and follow-up after surgery [17].

The initial surgical procedure is crucial as it is the first therapeutic option consisting mainly of ovariectomy or ovarosalpingectomy which allows the diagnosis of the disease as well as its extension. Depending on the malignant behavior of the neoplasm and the tumor size, surgery can be performed by a transverse infraumbilical or a Pfannenstiel incision or through a midline approach. During the operation, the following staging principles should be considered according to the Children’s Oncology Group for pediatric ovarian germ cell neoplasms:

1. Intact ovarian removal without rupture of the tumor capsule. A salpingectomy must be performed if the fallopian tube is adherent.
2. Examination of the contralateral ovary with biopsy if a suspicious aspect is seen.
3. Inspection of the peritoneum, the liver and the omentum and resection of any abnormal tissue.
4. Inspection of aorto-caval and iliac lymph nodes and biopsy of suspicious ones.
5. Sampling of ascitic fluid for cytological examination. If ascites is absent, a washing is required [18,19].

Biopsy by laparoscopic approach is the best option in case of involvement of neighboring structures or if there is an evidence of a bilateral form. When the initial surgical approach consists of a biopsy or an excision with micro/macrosopic residues or in patients with metastatic disease, a neoadjuvant chemotherapy is recommended. A post-chemotherapy evaluation show generally a surgically resectable tumor. If the tumor is bilateral, the majority of authors recommend ovarian preservation on the least involved side. Mutilating excisions and bilateral ovariectomies are accepted only when chemotherapy is not effective [20]. Despite the marked favorable outcomes after our management of this patient, our case reporting is limited by the single institution experience. Additional survival studies with prospective enrollment are awaited to confirm these findings.

### 4. Conclusion

Ovarian tumors and especially mixed ovarian germ cell tumors are uncommon in children. Their management must be multidisciplinary between pediatricians, pediatric surgeons and pathologists. Surgery is the standard of care, which should be conservative to preserve reproductive function. A laparotomy approach still the decision of choice if malignancy is suspected, surgical staging is required, and in case of large tumors.

During her last visit and follow-up, our patient achieved a complete remission and her family was satisfied with our management strategy.

### Table 1

| A timeline showing the evolution of the disease in our patient, its medical management and following up. |
|----------------------------------------------------------|----------------------------------------------------------|----------------------------------------------------------|----------------------------------------------------------|
| One year before the consultation | One week before the consultation | Medical consultation | Postsurgical management | After six months of follow-up |
| Paroxysmal abdominal pain | Worsening of abdominal pain | • Apyrexia and good general condition | Adjuvant chemotherapy | • No recurrence or tumor residus. |
| | | • Abdominal mass | | • Normalisation of tumor markers. |
| | | • Abdominal ultrasound and MRI. | | |
| | | • Tumor markers | | |
| | | • Surgery | | |
| | | • Histological diagnosis. | | |

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