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Paediatric Gynaecologic Neoplasms

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Introduction

A variety of gynaecologic neoplasms occur in the pediatric population. Differentiating between benign and malignant lesions and reaching the correct diagnosis in a timely manner is crucial to preserve fertility, which is a priority in this population. Awareness of the clinical, laboratory and pertinent imaging characteristics of such lesions will facilitate their recognition in clinical practice.

Ovarian neoplasms

Ovarian tumors are uncommon in childhood having an incidence of 2.6 /100,000 girls per year. Majority of these are benign; less than one third are malignant [1,2]. Presentation is usually as a palpable mass or abdominal pain. Precocious puberty or virilisation may occur with hormone-secreting neoplasms [3]. Diagnosis is often delayed and can be difficult owing to non-specific symptoms at presentation and diverse features on imaging. However, familiarity with the demographics, laboratory results and imaging features allows narrowing the deferential diagnosis and may allow a fertility-sparing ovarian surgery. Checking tumor markers may not be only useful for diagnosis but also for treatment evaluation and recurrence detection. Elevated serum levels of AFP or β-hCG are highly indicative of malignancy; however, they are elevated with only 50% of malignant neoplasms [4,5]. On imaging, a mass size ≥ 10 cm, presence of solid components and tumoral enhancement are highly suggestive of malignancy, while a simple cyst is highly predictive of benignity [6]. Usually, management of ovarian tumors is by resection in addition to uni- or bilateral salpingo-oophorectomy, and prognosis is generally good [7]. Ovarian neoplasms are classified into three main groups; germ cell tumors, sex cord- stromal tumors, and epithelial tumors as described below.

Gem cell tumors (GCTs) are the most common ovarian neoplasms in children. Common tumors in this class include teratomas (mature and immature), dysgerminoma and yolk sac tumors [8]. Mature cystic teratoma (MCT) is a benign cystic lesion, and is the commonest (55-70%) pediatric ovarian neoplasm. Torsion may occur in one third of the cases and present with acute abdomen [9]. On Ultrasonography (US), MCT show variable appearance depending on its contents which could be a mix of fluid, sebum, fat, hair and/or calcifications. Most commonly, they manifest as a unilocular cyst with a hyperechoic tubercle originating from the wall, the so-called Rokitansky nodule. This contains hair follicles and fragments of teeth or bone which may cause posterior acoustic shadowing preventing assessment of the bulk of the tumor posteriorly on US [9]. On CT and MRI, large amounts of fat or coarse calcifications within the lesion are diagnostic features of a teratoma [10]. Immature teratomas (IT), on the contrary, demonstrate a clinically malignant behavior and are much less common than mature teratomas. Degree of malignancy is determined based on the level of neuroectodermal component differentiation in tissue samples [11]. Teratomas can also occur extragonadal; the important one which needs a particular mention includes sacrococcygeal teratoma (SCT). SCT is the most common teratoma in neonates, and often detected on prenatal US. Most SCTs are benign at birth; however, malignant transformation can occur. Associated anorectal and genitourinary malformations may also occur. MRI is the best modality for depicting this lesion and evaluating surrounding structures for invasion. After resection of SCT, surveillance for recurrence is recommended for 3 years utilizing a combination of physical exam, AFP levels and periodic pelvic imaging [12- 14]. Dysgerminoma is the most common malignant ovarian germ cell tumor (MOGCT). On MRI, it appears as a solid lobulated mass with T2 hypointense fibrovascular septa that enhance avidly on gadolinium injection [3]. Yolk sac tumor is another MOGCT which is known for AFP secretion. However, its imaging finding is non-specific [15]. Prognosis for GCTs, even malignant types, is generally excellent [16].

Sex cord stromal tumors (SCSTs) are a class of tumors known for hormonal secretion resulting in hormonal events. Although fibromas are the most common SCST in adult women; Juvenile granulosa cell tumor is the most common in pediatrics. This tumor is classically associated with isosexual pseudo-precocious puberty in 80% of cases due to estradiol secretion, which can be used as a tumor marker in addition to inhibin [17]. On imaging, tumor may show a characteristic sponge-like appearance on T2WI on MRI, attributed to innumerable cystic lesions intermixed within a solid mass. Extended follow up periods are needed for this tumor, since it has a tendency for late recurrence even 10 years after diagnosis [18,19]. Sertoli-Leydig cell tumors are very rare in children and show a low malignant potential. Classically, patients present with virilization and hirsutism due to androgen secretion by the tumor, however, the tumor is only functional in only one third of the cases. Color Doppler US and/or MRI are helpful tools for detecting small virilizing tumors which are often difficult to detect even with transvaginal US [19,20].

Epithelial tumors are classified into 3 categories: benign, malignant and borderline malignant. Carcinomas are extremely uncommon in pre-menarchal girls. Serous and mucinous cystadenomas are the most common lesions. On imaging, serous cystadenoma are uni- or multilocular, with homogenous contents and thin septations. While mucinous cystadenomas are typically multilocular with heterogeneous

components [21]. Other miscellaneous ovarian tumors include gonadoblastoma which is a rare neoplasm composed of a mixture of germ cells and sex cord-stromal derivatives, and arises almost exclusively in dysgenetic gonads of patients having Y chromosome mosaicism; Prophylactic bilateral gonadectomy is recommended in those patients [22]. Small cell carcinoma of the hypercalcemic type is a rare and aggressive malignancy of uncertain histogenesis with a high tendency forextraovarian spread and metastases which are seen in about 50% of the cases. Classically described hypercalcemic manifestations (e.g. polydipsia, polyuria, etc.) occur in two third of these patients [23].

Uterovaginal neoplasms

Uterovaginal tumors are rare in the paediatric population, but they are more likely to be malignant than in adults. Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in childhood and it most commonly arises in the pelvis being the most common paediatric uterovaginal malignancy. It often presents with a haemorrhagic perineal mass or a grape-like lesion protruding from the vagina [24,25]. US reveal a hypo echoic mass. At MRI, it has uniform hypo intensity on T1WI but heterogeneous hyper intensity on T2WI and contrast-enhanced T1W fat-saturated images. CT chest and bone marrow biopsy should be part of the staging work up in these cases, since most common sites for metastases are lungs and bone morrow. Chemotherapy is the first-line treatment [26].

Few gynaecologic neoplasms are associated with some hereditary tumor syndromes, such as endometrial and ovarian cancer in Lynch syndrome. Awareness of this relationship can be the key to raise the suspicion about certain syndrome-related neoplasms when imaging reveals a non-specific gynaecologic mass in a patient known to harbour one of these syndromes. Vice versa is also true [27]. In summary, a variety of gynaecologic tumors occur in the paediatric population. Maximizing survival while preserving fertility is precedence in such young patients. Imaging plays an important role in diagnosis and management planning. US remain the imaging modality of choice for initial evaluation. MRI is of great benefit for further lesion characterization and as problem solving. Although imaging findings may overlap in some lesions, differentiating imaging features of separate lesion groups in combination with the clinical picture and tumor marker profile assures delivering the optimal care.

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