

Mature Cystic Teratoma in a Five-year old Female: Dilemmas in Management

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The patient is a five-year old female who complained of abdominal pain. A ping pong ball-sized mass on left lower quadrant of the abdomen was palpated six (6) months ago, which rapidly grew in size and caused abdominal pain. Physical examination showed a 10 cm x 10 cm firm, non-tender, smooth hypogastric mass with limited mobility. Primary impression was a mature cystic teratoma and a differential diagnosis of a malignant ovarian mass was considered because of the elevated tumor marker CA-125 and papillary excrescences on abdominal ultrasound. A multidisciplinary team composed of pediatric anesthesiologist, pediatric hematology-oncology expert, pediatric surgeon and the obstetrician gynecologist was formed to do thorough pre- and post-op planning. The patient underwent exploratory laparotomy, peritoneal fluid cytology, left oophorectomy, and frozen section under general anesthesia. Frozen section revealed teratoma with mature components in three sections. Final histopathologic result was mature cystic teratoma, 13.5 centimeters in greatest tumor dimension. She was discharged on the tenth hospital day. Dilemmas in management were discussed.

Key words: mature cystic teratoma, pediatric age

Introduction

An ovarian mass in a five-year old is uncommon. The incidence of ovarian cysts in prepubertal children is actually lower than in neonates and perimenarchal/ menarcheal girls because gonadotropin stimulation of the ovary decreases after the neonatal period and generally remains low until puberty

A review of the Department of Laboratories' records from 2019-2022 showed 32-cases of ovarian masses in patients less than 18 years old. All were admitted at the Philippine General Hospital and 9 patients' histopathologic results were signed out as malignant.¹

Dilemma in management of this patient arose. What is the best plan of management? What if it

turned out to be malignant? Who will do the surgery? Will it be the pediatric surgeon or the gynecologic surgeon? Do we need to do a total hysterectomy, bilateral salpingo-oophorectomy, peritoneal fluid cytology, bilateral lymph node dissection in case the frozen section turned out to be malignant? The patient is pre-pubertal and extirpation of her uterus and ovaries would mean loss of her normal development. How do we explain this to the parents? What are the possible short and long-term complications? These questions are all addressed in this case.

The Case

This is a case of a 5-year-old female, who came with a chief complaint of abdominal pain. Patient's height and weight were appropriate for age, and she

came conscious, and oriented to time and space. Vital signs were stable. Both breasts and pubic hair areas were at Tanner stage 1. The abdomen was globular, and slightly distended, but soft. A hypogastric mass which measured approximately 10 cm x 10 cm, with limited mobility, non-tender and doughy, was palpated. She had no bipedal edema.

Transabdominal ultrasound revealed an anteverted uterus with smooth contour and homogeneous echopattern measuring 5.5 cm x 2.1 cm x 1.6 cm. The cervix measured 1.8 cm x 0.9 cm x 0.5 cm with homogeneous stroma and distinct endocervical canal. The endometrium was uniform and hyperechogenic measuring 0.2 cm. At the right adnexal area, occupying the right lower quadrant up to the subhepatic area was a multilocular-solid mass measuring 10.3 cm x 9.5 cm x 6.3 cm (volume 322.4 cc) containing mixed echo fluid and echogenic foci casting acoustic shadows. At the inferior pole of the mass, there was a locule containing echogenic stipplings. There were also papillary excrescences measuring 1.2 cm x 1.4 cm x 1.0 cm (volume 0.9 cc), 1.7 cm x 1.2 cm x 0.6 cm (0.6 cc) and 1.1 cm x 0.8 cm x 0.8 cm (volume 0.4 cc) at the superior pole of the mass. The septum and capsule both measured 0.5 cm. The left ovary was not visualized. The liver parenchyma was homogeneous. The bilateral renal calyces were not dilated. Power doppler of the right adnexal mass showed absent vascularity (Color Score = 1). Sonographic impression was a right adnexal mass, probably a dermoid cyst by subjective assessment with 6.9% risk of malignancy based on Simple Rules RISK calculation (cut off is 10%; more than 10% means there is a likelihood that the mass is malignant).

Tumor markers showed normal Alpha fetoprotein 0.42 iu/ml and Beta HCG (< 1.20 miu/ml), and slightly elevated CA -125 (45.2 u/ml) levels. Patient had normal complete blood count profile with a hemoglobin of 134 mg/dl.

The patient was referred to the division of Gynecologic Oncology who advised referral to the Pediatric Surgery Division. The Gynecologic Oncology Division had no surgical instruments for the pediatric age group, and are not trained in the surgery of children. The patient was presented in the pre-operative conference of the Pediatric Surgery Division. General Pediatrics assessed the patient as Moderate risk (ASA II, Class II, Risk score 10) in

developing intra- and post-operative complications for the contemplated procedure. They also referred the patient to the Pediatric Hematology-Oncology Division due to the consideration of malignancy.

A repeat whole abdomen CT scan was done, revealing an abdominopelvic mass whose dimensions remained unchanged since February 2020. CT scan findings was highly suggestive of ovarian teratoma, with minimal pelvic ascites.

On the 7th hospital day, she underwent exploratory laparotomy. Intraoperatively, the liver and diaphragm were smooth, there was minimal peritoneal fluid, the ovarian mass did not have any point of rupture, it was not adherent to any adjacent structure (Figure 1). It was decided upon intraoperatively, that the surgery will be performed by the Obstetrician-Gynecologist assisted by the Pediatric Surgery fellow. Careful dissection was done to prevent tumor rupture. The right ovary was normal. A conservative procedure was performed - left oophorectomy - because grossly, the mass looked benign and a frozen section was to be performed.



Figure 1. Gross finding in the case of a five-year old female with an ovarian mass.

Grossly, the ovarian mass had a tan-red to cream-red, smooth glistening external surface and measured 13.5cm x 5.5cm x 7.0cm. Cut section revealed variegated, solid-cystic surfaces. The solid components had cream-yellow to cream-white, soft solid cut surfaces with firm cartilaginous and hard bony components. The solid areas were covered by cream-white rough surfaces with hair and waxy material. The multiloculated cystic component

was lined by cream-red, smooth, glistening surface containing light, yellow, serosanguinous, hazy fluid. (Figure 2).



Figure 2. Cut section of the left ovarian mass in a five-year old girl. There was note of sebum and hair. The ovarian capsule was thin and no areas of necrosis was noted. The papillary excrescences noted on ultrasound was absent.

Peritoneal washing was done for peritoneal fluid cytology. The procedure done was left oophorectomy and frozen section under general anesthesia. The gynecologist performed the procedure assisted by the pediatric surgeon. Frozen section showed the following: teratoma with mature components seen in 3 sections. A formalin-fixed paraffin embedded tissue processing for further evaluation. Estimated blood loss was 100 cc, allowable blood loss for this patient is 341 cc. Postoperatively, CBC showed a hemoglobin of 123 g/li, from an initial of 134 g/li.

The patient was discharged well and ambulatory. Final histopathologic diagnosis was mature cystic teratoma 13.5 cm in greatest tumor dimension. (Peritoneal Cavity), Peritoneal Fluid Cytology, Few reactive mesothelial cells and inflammatory cells, negative for tumor.

Discussion

A mature cystic teratoma may arise from one or three germ cell layers – ectoderm, mesoderm and endoderm. In females, these mature cystic teratoma are found in the ovaries. These well-differentiated teratomas are generally called mature cystic teratoma. These account for 10-20% of all ovarian neoplasms. These encapsulated masses with mature tissue are composed of well-differentiated derivatives from at least two of the three germ layers.²

Ovarian mature cystic teratomas are the most common histologic type of childhood germ cell tumors. However, in the literature, the appropriate approach to treatment in the pediatric age group remains unclear. In a study which reviewed the outcome of 58 patients aged 9-15 years old, 55 patients had mature cystic teratoma, 3 were immature teratoma or the malignant type. Twenty eight (28) girls had exploratory laparotomy and twenty three (23) patients had laparoscopy. As to the question of ovarian preservation, in earlier studies (1999-2003), only 11.11% underwent ovarian preservation. Much later (2004-2016), ovarian preservation rates improved to 40.54%.³

This was one of the dilemmas in the management of the index patient. Upon exploration, grossly, the mass seemed benign. Thus, oophorectomy with frozen section was done. Since a frozen section was planned, if the specimen turned out to be malignant, a salpingo-oophorectomy with bilateral lymph node dissection, infracolic omentectomy, and peritoneal fluid cytology was deemed as the best management for the patient. Preservation of the contralateral ovary, fallopian tube and uterus will be the best option so that the patient could still have her full reproductive potential.

Another dilemma in the management was category of the pediatric clearance which was moderate *risk for an adverse event*. Can the patient tolerate the procedure? How much blood loss could the patient tolerate? To compute for the estimated blood volume and allowable blood loss:

Estimated blood volume = weight x factor for age (75)
= 18.2 kg x 75
= 1,365 ml.

75 is the constant for computing the estimated blood volume for patients 3-6 yrs old.²

$$\text{Allowable blood loss} = \frac{\text{actual Hematocrit} - \text{lowest acceptable Hematocrit}}{\text{actual Hematocrit}} \times \text{Estimated blood vol. (EBV)}$$

$$\text{In this patient: } \frac{(0.4 - 0.3)}{0.4} \times 1,365 \text{ ml} = 341.25 \text{ ml}$$

Thus, the allowable blood loss for this patient is 341.25 ml

For children aged 3-6 years old, 70-75 ml is the factor used for children of this age. The computed EBV is 1,274 for 70 ml and 1,365 for 75 ml. For a female child, the lowest factor is usually used.⁴ The actual blood loss was 150 ml.

The patient was able to tolerate the procedure well and the post-operative course was uneventful. A good clinical practice notable in this case is the multidisciplinary approach to management: team consultation composed of the pediatric anesthesiologist, pediatric hematology-oncology expert, pediatric surgeon and the obstetrician gynecologist was carried out. All aspects of the case were analyzed to ensure the best procedure and outcome for the patient.

Reviewing the results of the laboratories where the CA-125 level was slightly elevated, and a pelvic ultrasound which revealed the presence of papillary excrescences, plus the history of rapid increase in tumor size, there was a possibility that the mass was malignant. Prior to the surgery, the parents of the patient were thoroughly counselled on the possibility of the mass being malignant. Since the patient has not reached menarche, the surgical team with the parents' consent, agreed that the most conservative procedure, unilateral oophorectomy, would be the procedure of choice. If on frozen section, the ovarian mass turned out to be malignant, the surgical procedure would be a unilateral salpingo-oophorectomy, peritoneal fluid cytology, and bilateral lymph node dissection. The uterus and the contralateral ovary will be preserved so that the child can reach her full reproductive potential.

Since mature cystic teratomas can be bilateral in 8-14% of patients, the parents of the index case were made aware that another mature cystic teratoma could develop in the future in the contralateral ovary, and there is likewise a possibility of recurrence

in 2-10 years.⁵ Thus, regular check-ups with the gynecologist was advised.

In the PGH Ovarian Tumor Registry for pediatric patients, records from 2019-2022 showed the following census: in 2019, ten pediatric patients were operated on, 7 patients for year 2020, 11 patients for year 2021, and 4 for January to June 2022. Of these ovarian tumor cases, nine (9) pediatric patients were diagnosed as having malignant ovarian tumors.

Table 1. Years the pediatric patients were admitted and the histopathologic diagnosis. Note that for January-June 2022 – No malignant ovarian tumor patients were registered.

Year	Histopathologic Diagnosis
2019	Immature Teratoma – 1 Germ Cell Tumor -1
2020	Yolk Sac Tumor – 1 Poorly Differentiated Carcinoma – 1 Germ Cell Tumor with Yolk Sac Tumor Component - 1
2021	Mixed Germ Cell Tumor with Yolk Sac Component – 1 Yolk Sac Tumor – 1 Immature Teratoma – 1 High Grade Adenocarcinoma of Mullerian Differentiation with Immunohistochemical Stains Consistent with Mucinous Carcinoma -1

There is also the possibility of short and long term complications, such as post-op infection, adhesions, and malignant transformation.⁶ Malignant transformation occurs in 1% of patients, but is more commonly seen in adults between 30-70 years of age. The mature cystic teratoma could transform into squamous cell carcinoma (most common), transitional cell carcinoma, malignant melanoma, choriocarcinoma, carcinoids, sarcoma and adenocarcinoma.⁷

Conclusion

The best approach to managing a pediatric patient with a large abdominopelvic tumor is a multidisciplinary team approach. The team should be composed of the obstetrician-gynecologist, pediatric

hematologist-oncologist, general pediatrician, pediatric surgeon and a pediatric anesthesiologist. Another important aspect is the establishment of trust and rapport between the doctors and the patients' parents. This comes with the responsibility to do thorough preoperative and post-operative counselling and a show of support for the patient and her family. Counseling should cover dilemmas in management, and the possibility of short and long-term complications such as infection, recurrence, and malignant transformation.

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