

Cystic Teratomas in Adolescent Girls

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Abstract

Diagnosis and Management of ovarian masses in adolescent is a challenging problem faced by gynaecologist. Amongst germ cell tumours mature teratoma is the most common. These masses arise from totipotent cells in the ovary and are benign in nature. Abdominal pain and palpable abdominopelvic mass is the commonest clinical presentations. Complications of cystic teratomas are torsion, hemorrhage, rupture, infection and malignant transformation. Rokitansky nodule, dermoid mesh and tip of iceberg sign are USG signs of mature cystic teratomas. ovarian cystectomy has proved to be safe method. Surgical treatment for teratomas by laparoscopy or laparotomy is discussed. Early diagnosis and preservation of fertility should be the aim. Counselling to adolescent girls and her parents is vital.

Keywords: Mature Cystic Teratoma; Immature Cystic Teratoma; Dermoid, Adolescent; Ovarian cystectomy

Introduction

The study of ovarian tumors in the young and adolescent girls is a story of "Triumph and Tragedy". Triumph because today, fertility prevention modalities are evolved. The diagnosis and management of ovarian masses in adolescents is a challenging problem faced by gynaecologists. Pelvic tumor in childhood and adolescence are devastating to these young patients and their families. Ovarian masses in adolescents are uncommon and the actual incidence is difficult

to estimate. An approximate incidence has been estimated as 0.0026%, and malignant ovarian tumours comprise about 1% of all childhood cancers Ovarian tumours occurring in adolescent age group have diverse presentations ranging from asymptomatic masses to acute abdominal pain.¹ Pathology of ovarian neoplasm is one of the most complex areas of gynaecology, because the ovary gives rise to a great range and variety of tumors than does any other organ. Germ cell tumors occur prior to puberty or in elderly adult life.

The Germ Cell Tumors Include Many Types

- Teratomas- Mature Cystic Teratoma and Immature Teratoma
- Dysgerminomas
- Endodermal sinus tumors
- Non gestational choriocarcinomas

Among germ cell tumours mature teratoma is the most common ovarian germ cell tumor and is composed of well- differentiated adult tissue type and little tendency to malignant change. The immature teratoma is composed of variety of tissues, including immature mesenchymal or fetal neural elements.² most frequent neoplastic tumor of children and adolescents is mature cystic teratoma and is accounts for more than one half of ovarian neoplasms in women less than 20 years of age.³ These masses arise from totipotent cells in the ovary which develop into fully differentiated ectodermal, mesodermal, and endodermal tissue and are benign

in nature.⁴ Mostly they are asymptomatic and are discovered incidentally on exam or imaging.⁵

MCTs are indolent tumors with an estimated growth rate of 1.8 mm per year. It is unclear what signals these lesions to grow. The sebaceous gland component of these tumors are stimulated by increasing levels of estrogen and progesterone which can be explained by increase in size seen in MCTs after puberty and the arrested growth after menopause.⁶ In approximately 10–20% of cases they are bilateral.⁷

Origin of Mature Teratoma

Theorised to develop from genetic material contained within a single oocyte. The oocytes capable of parthenogenesis result from an arrest of oocyte development following meiosis. As a result, almost all mature cystic teratomas have a 46, XX karyotype. If a germ cell tumor undergoes parthenogenesis, embryonal tissues form teratomas either mature or immature.⁸

Pathological Features

Macroscopic Appearance

At gross inspection, an MCT is a well circumscribed cystic mass with a smooth surface that is surrounded by a capsule of variable thickness and is filled with sebaceous material. Most MCTs are unilocular (88%), but some are multilocular.⁹

Microscopic Appearance

Histologically, the neoplasm often has a focal internal protuberance, known as a Rokitansky nodule, that may contain hair, bone, teeth, muscle, or cartilage, and the cyst wall is lined by squamous, respiratory, or gastrointestinal epithelium.^{9,10}

Clinical Presentations

- Abdominal pain is the most common presenting symptom of ovarian tumors (57%)
- Palpable abdominal or pelvic mass (46%).
- Nausea
- Vomiting
- Poor appetite
- Weight loss
- Constipation

- Urinary frequency

They may be Asymptomatic, with the Tumor being Detected Incidentally

- Tenderness may be a symptom of
- Torsion (3%-16%)
- Hemorrhage (3%-5%)
- Rupture of ovarian tumors (1%-4%)
- Malignant Transformation (1%-2%)
- Infection (1%)¹¹

Rarely, Paraneoplastic Syndromes, Including

- Autoimmune hemolytic anemia
- Anti-N-methyl-D-aspartate (NMDA) receptor encephalitis^{12,13}

Investigations

USG Signs of MCT

- *Rokitansky Nodule/Dermoid Plug:* Which is a cyst containing highly echogenic nodule inside
- *Dermoid Mesh:* Which represents floating hair inside the cyst
- *Tip of Iceberg sign:* sign resulting from acoustic shadowing of the sebum on hair content of the cyst;
- *Fat-fluid level.*

Ref: Kathleen E. O'Neill, Amber R. Cooper, J Pediatr Adolesc Gynecol. 2011 June ; 24(3): 176–180.

The probability of malignancy in dermoid cyst is determined mainly by the ultrasound which characteristically appears as cystic adnexal mass or dermoid plug which protrudes into the cystic lumen and showing posterior acoustic shadow.¹⁴ In about 70% of MCTs acoustic shadowing occurs secondary to calcifications or a mixture of hair and adipose tissue in the tubercle. Other ultrasonographic findings include a diffusely or partially echogenic mass which represents sebaceous material, multiple thin echogenic bands attributable to hair, fat-fluid levels, floating debris, either alone or a combination of these features.¹⁵ Although ultrasonography is diagnostic in most cases, CT or MR imaging should be considered as of great importance cases as these imaging modalities are much more sensitive in relation with fat identification. At CT, fat attenuation in

a cyst, with or without calcification in the cyst wall or Rokitansky nodule, is the characteristic finding. A fat-fluid level or a floating mass of hair is occasionally seen.¹⁶ At MRI, fat is seen in the mass, which is hyper intense on T1-weighted images and shows signal loss on frequency-selective fat-saturated T1-weighted images.¹⁷ A small percentage of cases (15%) demonstrate a minimal amount of fat in the cyst wall or Rokitansky nodule and show signal drop with gradient-echo opposed-phase T1-weighted MR sequences.¹⁸ MR imaging is the preferred imaging modality in some cases owing to the excellent soft-tissue contrast it provides.¹⁹

Looking forward into sensitivity of ultrasound for detection of MCT, annual imaging in prepubertal and young adolescents followed by annual pelvic examinations in older adolescents is appropriate.²⁰

Tumor Markers

The serum AFP level is elevated in patients with GCTs such as yolk sac tumors, immature teratomas, embryonal carcinomas, and mixed GCTs with yolk sac elements.²⁰

Treatment

The conflict between surgical vs. conservative management in adolescent ovarian masses is challenging. In the past, oophorectomy was treatment of choice in children with MCTs due to large cyst size, concern for occult germ cell malignancy, presence of torsion, decreased viability of remaining ovarian tissue, and risk of spillage and/or incomplete removal. However with the advancement of surgical techniques, the use of ovarian cystectomy has proved method for tumor removal in the pediatric and adult populations as oophorectomy.^{21,22} Cystectomy should be the first line treatment in lesions that are preoperatively consistent with teratoma.²³ Some surgeons propose that intraoperative biopsy of the contralateral and seemingly unaffected ovary should be performed due to the increased risk of bilateral MCTs. Yet, ovarian wedge biopsy is with the complications like the possible hemorrhage, infection, and adhesion formation. Safe alternative to wedge biopsy is preoperative ultrasound in combination with careful inspection of the contralateral ovary at the time of surgery.²⁴

Selection of the Best Choice of Surgical Treatment in Dermoid Cyst: Laparoscopy or laparotomy? Many trials have concluded laparoscopy as a best treatment in surgical management of dermoid

cyst. The gold standard surgery in dermoid cyst management is the Laparoscopy.

Following are the Advantages of the Laparoscopic Approach to Dermoid Cyst Treatment

- Less bleeding
- Less postoperative pain
- Less need for postoperative analgesic
- Shorter hospital stay
- Less adhesion formation
- Better cosmetic results
- Less cost (overall)
- Better magnification

Disadvantages of the Laparoscopic Approach in Dermoid Cyst Compared to Laparotomy Include

- longer duration of operation
- higher rate of spillage
- higher rate of recurrence
- higher cost (individually)
- increased risk of surgery unique to laparoscopy^{25,26}

When Should Laparotomy be Considered in Dermoid Cyst Management

- Large mass controversy exists in the precise tumour size for which laparotomy is the recommended treatment. Some authors have recommended 10 centimetres as the cut-off size for laparotomy. There is evidence of solid components in the tumour mass and a suspicion of malignancy.
- Bilateral cysts²⁷

Cystectomy is considered as dermoid cyst surgery of choice instead of oophorectomy. It has been indicated that about 35 primordial follicles are found in each 1 mm² of ovarian surface in women.

The Techniques Listed Below are Suggested for Maximum Preservation of Ovarian Tissue

- A combination of hydro-dissection and blunt dissection by laparoscopy instead of the traditional stripping technique.
- *The mesial side incision of the ovary:* The mesial side of ovary is the point of connection of fimbria to ovary.

Following Dermoid Cyst Operation Indices Used to Clarify Remaining Ovarian Tissue and its Function are Listed Below:

- Visualization of remaining ovarian cortex by sonography reported as tissue volume
- Measurement of FSH
- Evaluation of basal antral follicle number
- Measurement of ovarian size
- Doppler velocimetry of peak systolic velocity, and measurement of anti-mullerian hormone (AMH).^{28,29}

In 40% - 50% of laparoscopic cystectomy cases, Spillage of cyst content is reported which is highest spillage rate in this setting. Lower spillage rates in laparoscopic oophorectomy and even less spillage in laparotomy have been reported (10%-15%). Chemical peritonitis and adhesion formation might be occurring due to the Spillage of dermoid cyst.^{30,31}

Complications of Dermoid Cyst

1. *Torsion:* Dermoids are vulnerable for torsion (15%) because of their long pedicle and (heavy) weight (due to fat).
2. *Rupture:* Spontaneous rupture is rare. Their thick cyst wall resists rupture compared with other ovarian neoplasms. But if it does rupture, it leads to a very serious chemical peritonitis. Peritonitis is attributed to sebum and hair contents of the cyst. Chronic leakage of teratoma contents leads to granulomatous peritonitis that may be interpreted as widespread malignancy.
3. *Recurrence 3-4%:* Recurrence is more common in cases of laparoscopic treatment, probably due to unrecognized small cysts. Other predictive factors of recurrence are: young age (less than 30), large cyst size (8 cm or larger) and bilateral cyst.
4. *Malignancy:* Rare 1.7% usually squamous cell carcinoma, rarely malignant thyroid tumors. Risk factors of malignant transformation in dermoid cyst are summarized in the following 4 considerations: age of over 45 years, cyst size of more than 10 cm, rapid growth and abnormal sonographic and Doppler findings including increased vascularity, hetero echo pattern, papillary projections and septation.³²

Immature Teratoma

Immature teratoma typically affects a younger

age group. The younger the patient, the more possibility is that the teratoma will be the immature germ cell type. Immature teratoma frequently occurs between the ages of 10 and 20 years, with a median age of 17 years. This represents 10%-20% of all ovarian malignancies in patients younger than 20 years.³³ Due to its more aggressive behavior and a worse prognosis than does mature teratoma it must be treated as malignancy, although it is not truly malignant.

In 33%-65% of patients with immature teratoma, elevated serum AFP levels have been reported.³⁴ At present treatment of choice for immature teratoma is surgery with unilateral salpingoophorectomy and staging procedure.

Immature teratoma is frequently unilateral, large, and predominantly solid. Histologically, immature teratoma contains a variable amount of primitive immature embryonal tissues derived from three germ cell layers admixed with mature tissues. The histologic grade is determined by the quantity of the immature neuroepithelial tissue.

At CT and MR imaging, the tumor typically appears as a large heterogeneous mass with predominantly solid or mixed solid and cystic lesions. Discrete calcifications throughout the tumor and punctate foci of fat in the solid components are usually found. Hemorrhage is rarely seen. Identification of a solid portion with numerous cystic areas is of great importance as it help to differentiate between immature teratoma and MCT.³⁵

Discussion

Ovary gives rise to wide variety of tumour masses ranging over a wide spectrum of pathology from benign to highly aggressive malignant tumors. The incidence of malignant of tumours in adolescents is higher than in adults. Therefore, the detection of these lesions is worrisome to patients, her families, and gynaecologists. Many germ cell tumors are of mixed type, with various patterns or permutations of Dysgerminoma, Teratoma, Choriocarcinoma and Yolk sac found within a single tumor. Early diagnosis is possible by imaging techniques. Ultrasound is an excellent, non-invasive, inexpensive and useful diagnostic tool. Dermoid cyst similar to those found in the ovary are sometimes found retroperitoneally especially in the presacral area. It is remarkable that tissues which develop appear to be those normally found in the

cranial end of a foetus hair, skin, teeth, cartilage (of trachea) and nervous tissues. Comprises 10–25% of all ovarian neoplasms. They are bilateral in 10% cases, usually unilocular with smooth surface, contains hair and sebaceous material, lined in part by squamous epithelium. Sometimes the sebaceous material collects together in the form of pellet.

Forty per cent of dermoid cysts are associated with mucinous cystadenoma. The nomenclature dermoid cyst is a misnomer as it contains tissues from all the three germ cell layers that is ectoderm, endoderm and mesoderm, but there is preponderance of ectodermal tissue.

In the inner surface there is one area of solid projection called Rokitansky's protuberance which is covered by skin, sebaceous glands and at times teeth and bones. Mesodermal teratomas are rarely present. They are composed either solely or predominantly of one highly specialised tissue type like thyroid tissue- struma ovarii which may be associated with hyperthyroidism. Struma ovarii may be either benign or malignant. Ten per cent of dermoid cysts are diagnosed during pregnancy.

Bilateral dermoid cyst in a young adolescent, nulliparous woman is the most challenging situation because the considerable amount of ovarian stroma need to be preserved for her menstrual functioning and future fertility.

In adults, the incidence of recurrent MCT following cystectomy is 3–4%. In younger patients there is 2–3% incidence of subsequent development of germ cell tumors with multiple or bilateral MCT. Malignant transformation in a mature teratoma is a very rare complication. Approximately 80–85% of malignant transformations have been reported as squamous cell carcinomas.

For fertility preservation laproscopic approach should be adopted where possible. The surgical approach depends upon the laproscopic skills of particular surgeon. Conservative surgery is paramount in development of normal puberty and in optimizing future fertility for benign ovarian disorder in young women. When dealing with MCTs, cystectomy is almost always preferred over the oophorectomy. The thinned cortical tissue which remains after cystectomy contains numerous viable follicles which can play an emence role for the future hormonal production and oocytes for reproduction. It is important that the amount of ovarian tissue remaining can dramatically have a good impact on the future ferility.

Recent Advances

Are we increasing the chances of recurrence or incomplete resection by choosing laparoscopy
When trying to prevent adhesion formation ?

Are we taking risk of subsequent morbidity by avoiding surgery in smaller suspected teratomas?

Could low dose oral contraceptive pills diminish sebaceous gland stimulation and subsequent growth?

Much of these questions warrant further studies. In reality, the risks of expectant management in women be so small that we should consider follow-up without intervention to preserve ovarian function and future fertility more often.

Conclusion

Early correct diagnosis and preservation of fertility should be the aim. Intervention should be pursued if the patient becomes symptomatic or considered if the individual lesions grow to a diameter of greater than 5 cm. When there is need of surgery the ideal approach combines laparoscopy and cystectomy and leaves as much viable ovarian tissue in place in order to preserve the patient's reproductive functioning. It is high time to address the issue in our community as patients not only belong to a tender age group, there are important questions related to their future fertility and quality of life as well. Support and sensitivity for the frightened girl and her family is essential as is high quality decision making by the gynaecologist. Though rare, these tumors require careful treatment. Tragedy has to be avoided. Gynaecologist need to be updated on the value of early diagnosis and conservative fertility sparing treatment. Counsell ing to adolescent girl and her parents is very important. Life long follow up has to be impressed on families.

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