

Management of Familial Ovarian Teratoma: The Need for Guidance

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Abstract

Keywords

- ▶ dermoid cyst
- ▶ familial ovarian dermoid
- ▶ mature cystic teratoma
- ▶ cystic teratoma
- ▶ ovarian teratoma

Ovarian teratomas in prepubertal females are uncommon, but familial ones are exceedingly rare. We report an ovarian teratoma in an 8-year-old girl, her mother, and her maternal grandmother. The risk of a metachronous tumor and subsequent complications (such as torsion) in the contralateral ovary remain unclear. There is no clear guidance on follow-up management of patient and family members in the literature. We have reviewed the literature and discuss the challenges for the pediatric surgeon arising from such cases.

New Insights and the Importance for the Pediatric Surgeon

Familial ovarian dermoids are extremely rare, but can have significant implications for all female family members. There is a significant risk for development of an ovarian dermoid, which may result in torsion and infarction of the ovary. There is a clear lack of guidance in the literature as how to manage and follow up these patients and their families. We present a case of a familial ovarian dermoid occurring through three generations. In this article, we have reviewed the literature and discuss management options. We encourage all pediatric surgeons to share similar experiences to establish guidance for future cases.

Introduction

Ovarian teratomas are derived from primordial ovarian germ cells and can be divided into three categories: mature (cystic or solid, benign), monodermal (benign), and immature (malignant). Mature ovarian teratomas are also known as dermoid cysts or ovarian dermoids, because of the great predominance of skin elements on histopathology. These tumors are slow growing and benign in the vast majority of cases. However, significant complications such as torsion, rupture, or ovarian hemorrhage may occur, especially if the tumor remains unidentified for a long time. The incidence of torsion is reported at approximately 16%, and rupture at 1 to 4%.¹

Lower abdominal pain is the most common complaint at presentation (~44% cases). Although such tumors constitute the most common ovarian tumor in prepubertal females, their incidence in children is reported as less than 0.1/100 000.

Familial ovarian teratomas are exceedingly rare. We report an ovarian teratoma in an 8-year-old girl, her mother, and her maternal grandmother, and discuss the difficulties in management of such cases.

Case Report

An 8-year-old girl presented to a tertiary pediatric surgery unit with a 1-week history of vague abdominal pain. She was

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Fig. 1 The CT scan demonstrates a complex ovarian mass in the center of the image. The mass has solid and cystic components and includes calcifications (arrow), all of which are typical of an ovarian teratoma.

systemically well. On clinical examination, she was found to have a palpable suprapubic mass. Abdominal ultrasound (US) was suggestive of a mass arising from the right ovary, and a computed tomographic (CT) scan was obtained to clarify the nature of the lesion. The CT scan showed a 6 cm complex ovarian mass, highly suggestive of an ovarian teratoma (**Fig. 1**).

Her full blood count was normal and the tumor markers α -fetoprotein (AFP) and β -human chorionic gonadotropin (β -hCG) were negative. Of relevance, 3 months prior to this presentation, she had been admitted to her district general hospital with abdominal pain when her abdominal US scan was normal.

At laparotomy, the right ovary and ovarian tube were found to be twisted and infarcted and were removed. Histopathology confirmed immature teratoma. The girl made a subsequent uneventful recovery.

Of particular interest was the girl's family history: her mother had required bilateral ovary-sparing resection of ovarian teratomas at the age of 21 years, and her grandmother had undergone oophorectomy for the same reason aged 25 years. We hence sought advice on the follow-up management for both the girl and her sisters—only to find that to date there is no established guidance.

Discussion

Familial ovarian teratomas are extremely rare, and there are no figures available on the exact incidence. The literature to date is limited to a handful of case reports that describe ovarian teratomas occurring through several generations (**Table 1**). A common genetic basis has been suggested numerous times, but remains to be elucidated. As

Table 1 Familial ovarian teratoma: summary of reported occurrences across several generations

Paper	Year	Relation	Age
Plattner and Oxorn ¹¹	1973	Mother 2 daughters	22 23, 23
Hecht et al ¹²	1976	Grandmother Granddaughter	N/A 21
Brenner and Wallach ²	1983	3 generations	7, 34, 45
Gustavson and Rune ¹³	1988	Mother 2 daughters	26 24, 22
Kim et al ⁹	1993	Mother 2 daughters	42 26, 20
Nezhat et al ¹⁴	2010	Mother 3 daughters	26 27, 22, 22
Braungart et al (this report)	2016	Grandmother Mother Daughter	25 21 8

demonstrated in our case, such tumors can have significant implications for all female family members. The risk of a metachronous tumor and subsequent complications (malignant degeneration, rupture, or torsion) remain to be established. This leaves the pediatric surgeon with a significant challenge both in terms of patient management and counseling and management of family members.

We performed a literature review of the Medline and PubMed database from 1971 to 2015, aiming to establish guidance. However, even when including literature on management of isolated, nonfamilial ovarian teratomas, the evidence is sparse.

To date, only six further similar cases have been reported in the literature (→Table 1). Only one of these reports—apart from our case—involves an ovarian teratoma in three consecutive generations.² All these previous reports have mainly focused on the theories of origin for ovarian teratomas, rather than the patient and family management.

A Canadian study retrospectively reviewed the follow-up management of children with nonfamilial ovarian teratomas following resection in their institution.³ The study found a recurrence rate of up to 15%. Whether these recurrences were true metachronous tumors, or recurrence of an incompletely excised tumor is difficult to establish. Nevertheless, this is significantly higher than previously published for adult populations, where the risk of recurrence ranges from 0 to 4%. Follow-up varied depending on the operating surgeon—ranging from one single follow-up appointment without any imaging to annual follow-up with imaging for at least 5 years postoperatively. Imaging modality at follow-up in these cases was by US scanning. The paper demonstrates clearly that the management of such cases is very subjective, with no guidance on follow-up.

US is an excellent modality to screen for ovarian pathology and is often used to confirm the presence of a mass.⁴ It is comparably cheap and easy to obtain. However, the appearance of mature ovarian teratoma on US is frequently nonspecific; certain pathology such as blood clots within hemorrhagic cysts can mimic mature ovarian teratoma. Furthermore, use of US to distinguish mature from immature ovarian teratoma is problematic, because it often fails to identify the solid components that are characteristic for immature teratoma.⁴ Therefore, further imaging in the form of either magnetic resonance imaging (MRI) or CT scanning is usually obtained to characterize the nature of an ovarian mass (as it happened in our case).

Both CT and MRI have an excellent sensitivity and specificity for detecting mature ovarian teratoma, mainly through detection of calcifications and fat within the lesion. Fat in the cystic lumen is the most specific finding in mature ovarian teratoma and is easy to detect both on MR and CT.^{5,6} Advantages of CT are mainly practical: it is faster and often easier to obtain than an MRI scan. Furthermore, CT seems to have a slightly higher sensitivity to detect the Rokitansky's protuberance compared to MRI.⁷ On the other hand, CT of course exposes the patient to ionizing radiation, which MRI does not. The girl in our case underwent a CT scan because she

presented out of hours, and further imaging was needed in a timely fashion prior to laparotomy to further characterize the mass detected on US.

In adult patients with nonfamilial teratomas, bilaterality is reported in 10 to 25% cases whereas in familial ones the reported risk is up to 46%.^{8,9} Rogers et al suggested follow-up with annual US scans in nonfamilial teratomas.³ However, Brown in 1979 described a case of identical twins with ovarian teratomas, where one twin required removal of seven mature ovarian teratomas in three separate operations.¹⁰ Our patient had a completely normal US 3 months prior to the subsequent presentation with a large infarcted ovarian teratoma, and therefore annual US scans for follow-up do not seem to be frequent enough. But for how long should follow-up be continued? And should other female family members be screened, too? We were unable to establish evidence-based guidance.

Our patient is being followed up with 3 monthly US scans and reviewed annually. Her younger sisters have had a single (normal) US scan.

Many open questions remain. It is to be hoped that further research into the etiology and genetics of familial ovarian teratoma will provide a sound basis for follow-up. We hope that this report will encourage other pediatric surgeons to share their experience, to establish better guidance for the future.

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