IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 14, Issue 5 Ver. V (May. 2015), PP 39-41
www.iosrjournals.org

Ovarian Mucinous Borderline Tumor In Perimenarchal Girls- A Dilemma To The Treating Doctor.

Dr. Poornima Vijayan[1]MBBS, MD
[1]Assistant Professor, Dept. of Pathology, MES Medical College, Malappuram district, Kerala, India.

Abstract: *Ovarian epithelial tumors are common in adult women but infrequent in children and rare before puberty. These tumors constitute a dilemma to the clinician because no general consensus exists regarding their treatment protocol in adolescent girls. In young patients with ovarian cysts requiring removal, a conservative approach of ovarian cystectomy is advocated to enable retention of functioning ovarian tissue for endogenous hormone production and future conception. However, incidence of recurrences are high in such cases. Borderline tumors in adolescents are usually associated with a favourable prognosis. We report two cases of mucinous borderline tumor in adolescent girls aged 15 and 14 years respectively, one of whom presented with a recurrent ovarian mass previously treated twice with cystectomy. Both of them showed evidence of huge multiloculated cystic masses on ultrasound examination and both were treated with cystectomy. Because the management of young patients is challenging, especially in cases with recurrence, regular follow-up of these patients is very important.*

Keywords: *mucinous borderline tumors, adolescent girls, ovarian cystectomy, recurrence*

I. Introduction:

Serous and mucinous neoplasms constitute the majority of borderline tumors and occur mostly in women of reproductive age. Howard C. Taylor, Jr., first used the term "semi malignant tumors" in 1929 for a subset of large ovarian tumors that had an indolent clinical course with relatively favorable patient outcome despite the presence of peritoneal disease. In 1967, FIGO established a separate borderline category of tumors. Since then, considerable controversy has surrounded the definition and management of borderline ovarian tumors because of their enigmatic pathogenesis and perplexing biologic behavior. Synonyms of borderline ovarian tumors include tumors of borderline malignancy, tumors of low malignant potential, and atypical proliferative tumors[1]. Borderline ovarian tumor comprise 10-15% of all ovarian epithelial neoplasms. They are infrequent in children and rare before puberty[2]. We report two such cases.

Case Report 1:

The first case was a 15 year old girl who with vague abdominal discomfort. On examination, a mass was palpable per abdomen. On ultrasound examination, a multiloculated large mass measuring 15x15 cm was seen arising from the left adnexa. A laproscopic cystectomy was performed and the specimen was sent to our department for histopathological examination. Grossly, it was a smooth walled cyst with thickness ranging from 0.4 to 0.7 cm sent in multiple bits, aggregate measuring 20 cm. the outer surface and cut surface were pale white in color. A part of the fallopian tube was noted stretched over the outer surface of one of the bits. Histopathology revealed cyst wall lined by tall columnar cell with apical mucin, thrown into filiform papillae with minimal stromal support, mild to moderate atypia, stratification and tufting of the lining epithelium. No evidence of stromal invasion was seen. It was reported as mucinous borderline tumor (Fig.1).

Case Report 2:

The second case was a 14 year old girl who with abdominal pain and constipation. On examination, a mass was palpable per abdomen. On ultrasound examination, a multiloculated large mass measuring 20cm in greatest dimension was seen arising from the left adnexa. The patient gave history of previous ovarian cystectomies for the same complaints on two separate occasions within a span of 2 years. The previous histopathogy reports were unavailable for review. A laproscopic cystectomy was performed and the specimen was sent to our department for histopathological examination. Grossly, it was a smooth walled cyst with thickness ranging from 0.7 to 1.5 cm sent in multiple bits, aggregate measuring 20 cm. the outer surface and cut surface were pale brown to dark brown in color. Histopathology revealed cyst wall lined by tall columnar cell with apical mucin, thrown into papillae and crowded glands arranged back to back, with mild atypia, stratification and tufting of the lining epithelium. No evidence of stromal invasion was seen. It was reported as mucinous borderline tumor (Fig.2).

II. Discussion:

A search of the published literature has revealed very scant information regarding mucinous ovarian tumors in premenarchal and perimenarchal girls. One author,Horiuchi et.al reported one case of huge mucinous borderline tumor in a 12 year old girl wherein he reviewed 16 case reports of mucinous ovarian tumors in premenarchal girl and concluded that in all cases age was more than 10 years and signs and symptoms were vague and non-specific. Abdominal pain presented in cases with torsion, but many of the remaining cases only displayed abdominal distension. Many patients presented with large tumors, with some as much as 6 - 7 kg. Histological findings included cystadenoma in 9 cases, cystadenocarcinoma in 2 cases, and borderline malignancy in 5 cases [2].

Mucinous borderline tumors (MBT), which comprises about 10% -15% of all epithelial ovarian malignancies, is characterized pathologically by features of malignant tumors, including cellular proliferation, stratification of the epithelial lining of the papillae, nuclear atypia, and mitotic activity, but without destructive stromal invasion[2]. Those findings were identified in both our cases, so MBT was diagnosed.

Mucinous borderline ovarian tumors consist of two distinct histologic subtypes: the intestinal (90%) and the müllerian (endocervical like 10%) histotypes. The intestinal subtype is usually unilateral and may coexist with pseudomyxoma peritonei in up to 17% of cases. The müllerian subtype is bilateral in up to 40% cases and coexists with ipsilateral ovarian or pelvic endometriosis in 20–30% of cases[1]. Both our cases were of intestinal subtype.

Mucinous borderline ovarian tumors are associated with KRAS mutations in more than 60% of cases. The increasing frequency of KRAS mutations (33–86%) has been described in mucinous cystadenomas, borderline ovarian tumors, and carcinomas[3].

Guidelines for surgical treatment of borderline ovarian tumors resemble those for ovarian cancer, and include total hysterectomy, bilateral salpingo-oophorectomy and staging procedures in women who do not wish to become pregnant in the future. However, fertility is an important issue for premenarchal children[4]. In young patients with ovarian cysts requiring removal, a conservative approach of ovarian cystectomy is advocated to enable retention of functioning ovarian tissue for endogenous hormone production and future conception. However, when a huge cyst is encountered, preserving the ovarian tissue may be difficult. Cystectomy may have a greater chance of preserving fertility, but is associated with higher recurrence rates than those seen after oophorectomy. Salpingo-oophorectomy has thus been recommended as fertility-sparing surgery[5,6] .

In the review of premenarchal mucinous ovarian tumors by Horiuchi et.al, all borderline mucinous tumor cases were stage I and all patients underwent salpingo-oopherectomy or oopherectomy and were alive without recurrence[2]. But in our case, one patient had three recurrences within a span of 2 years. Baksu et.al reported a case of recurrent mucinous cystadenoma, each time presenting of a huge size, unfortunately leading to hysterectomy and salphingo-oopherectomy at age 21 years[3].

Hence, it is important to keep in mind the possibility of recurrence, even in benign cysts. This is especially true for mucinous tumors as they are common and most of the time multilocular. Because the management of young patients is challenging, especially those with recurrence, the most important issue is probably the follow-up of these patients. Transvaginal ultrasound recommended every 3–6 months seems to currently be the most effective diagnostic tool for the follow-up of young patients treated with cystectomy for mucinous cystadenomas or borderline tumors[7].

III. Conclusion:

These cases have been presented for their rarity, the favorable prognosis associated with these tumors in the younger age group, the risks versus benefits of fertility sparing surgeries and the need for regular follow-up in these patients.

Acknowledgments:

I whole heartedly thank the authorities of MES medical college, my colleagues and H.O.D Dr. Asiq Sideeque for the moral support and encouragement rendered.

References:

[1]. Lalwani N, Shanbhogue AK, Vikram R, Nagar A, Jagirdar J, Prasad SR. Current update on borderline ovarian neoplasms. AJR Am J Roentgenol 2010;194(2):330–336.
[2]. A.Horiuchi, K.Kameoka, K.Sato Y Yamamoto, Y Watanabe. Huge mucinous borderline ovarian cystadenoma in a premenarchal girl. Open Journal of Pediatrics, Vol.2 No.1, 2012
[3]. Kurman RJ, Shih IeM. Pathogenesis of ovarian cancer: lessons from morphology and molecular biology and their clinical implications. Int J Gynecol Pathol 2008; 27:151–160
[4]. Baksu, B, Akyol, A, Davas, I, Yazgan, A, Ozgul, J, Tanik, C (2006) Recurrent mucinous cystadenoma in a twenty year old woman: was hysterectomy inevitable?. J Obstet Gynecol Res 32: pp. 61
[5]. Park, J.Y., Kim, D.Y., Kim, J.H., Kim, Y.M., Kim, Y.T. and Nam, J.H. (2009) Surgical management of borderline ovarian tumors: The role of fertility-sparin g surgery. Gynecologic Oncology, 113, 75-82

[6]. Tinelli, R., Tinelli, A., Tinelli, F.G., Cicinelli, E. and Malvasi, A. (2006) Conservative surgey for borderline ovarian tumors: A review. Gynecologic Oncology, 100, 185-191.

Figure legends:

Fig.1: Ovarian cyst lined by tall columnar cells with apical mucin, showing stratification,atypia and tufting. No definite stromal invasion was identified. 15/female. (10X,H&E)

Fig.2: Ovarian cyst wall lined by tall columnar cells arranged in papillary and cribriform patterns lined by mildly atypical cells. No definite stromal invasion identified. 14 year old girl with recurrent ovarian cysts. (10x, H&E)

FIGURE.1:

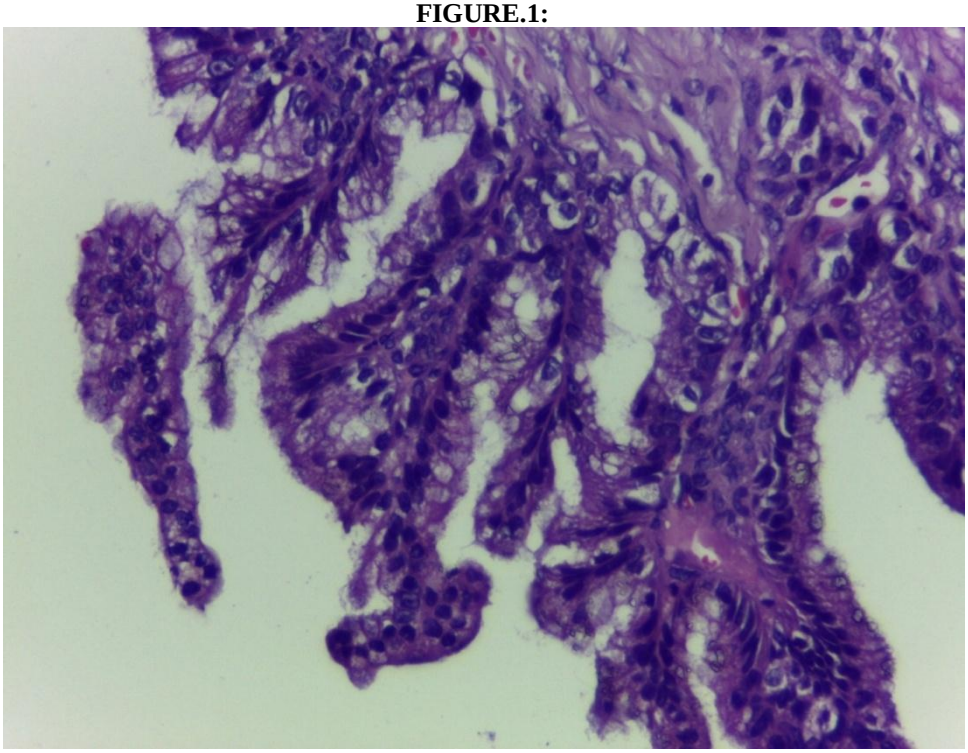

FIGURE.2: