Case Report

GALLEY PROOF

www.ijrhs.com
ISSN (o):2321–7251

Ovarian fibroma with minor sex cord elements-A rare case report

Ezhil Arasi

Professor & HOD, Department of Pathology, Osmania Medical College, Hyderabad, Telangana.

Corresponding Address: Dr. Ezhil Arasi, Professor & HOD, Department of Pathology, Osmania Medical College, Hyderabad, Telangana. Email: sushanthisush@gmail.com

Abstract:

Sex cord-stromal tumors ovary constitute 8% of ovarian tumors, fibroma being the commonest which accounts for 4% of all ovarian tumors. Its uncommon benign tumor of ovary and the one with focal proliferation of sexcord elements <10% is designated as fibroma with minor sex cord elements. These tumors occur at all ages with a peak in the perimenopausal age. So clinical manifestation in a hyper oestrogenic state in a postmenopausal female should raise suspicion of this entity. Large sizes may be associated with Meig's syndrome. Here in we report this case of ovarian fibroma clinically suspected as malignancy with unusual histological features like presence of sex cord elements and focal increased cellularity.

Key words: Benign ovarian neoplasm, Cellularity, Fibroma, Ovary, Sex cord elements and Hyperoestrogenic state

Introduction:

Ovarian fibromas are almost always benign in nature always benign in nature, frequency seen in middle-age women and not hormonally active [1]. Most of them are asymptomatic until they enlarge or involve adjacent organs and structures [2]. Upon gross pathologic inspection, ovarian fibromas are firm and white or tan. They have a smooth lobulated surface [3].On microscopic examination, there are intersecting bundles of spindle cells producing collagen [3]. The cellular subtype, approximately 10% of ovarian fibromatous tumors, exhibit hypercellularity, increased mitotic activity, and mild-to-moderate nuclear atypia [4].The cellular fibroma is a tumor of uncertain malignantpotential that may recur or be associated with peritoneal implants. The degree of mitotic activity is the main parameter for differentiating cellular fibroma from fibrosarcoma [4]. Macroscopically, the cellular fibroma has a whitish appearance resembling uterine leiomyoma, a generally solid consistency and, sometimes, small areas of cystic degeneration and stromal edema [5]. Its behavior is usually benign, but the completeness of excision and presence or absence capsule rupture are important prognostic parameters [5]. The minor sex cord elements are seen as small nests or tubules of cells resembling granulosa cells, Sertoli cells, or indifferent cells of sex cord type [6]. The term "minor" component of sex cord elements is defined as sex cord elements occupying no more than 10% of the area of the tumor on any slide [6]. The individual aggregate of these minor sex cord elements should not be greater than 0.45 mm. Immunohistochemically, the minor sex cord elements are positive for inhibin, calretinin, CD99, CD56, antikeratin antibody KL1 and MIC. [7,8]. On *CT scan:* fibroma usually unilateral manifest as diffuse, slightly hypo attenuating mass with poor, very contrast enhancement.[9-11]. The main differential diagnosis is pedunculated subserosal uterine leiomyoma [1].

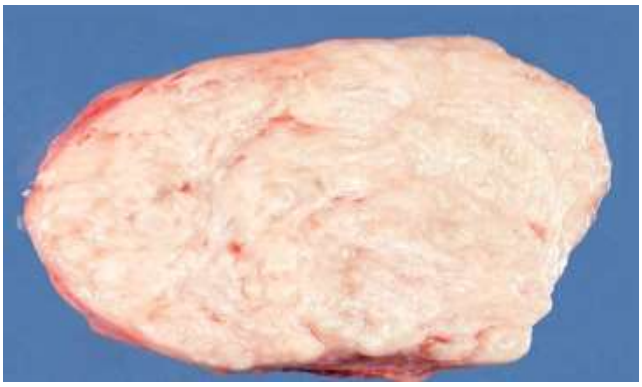

Figure 1: Gross: Cut surface shows a firm grey white solid mass

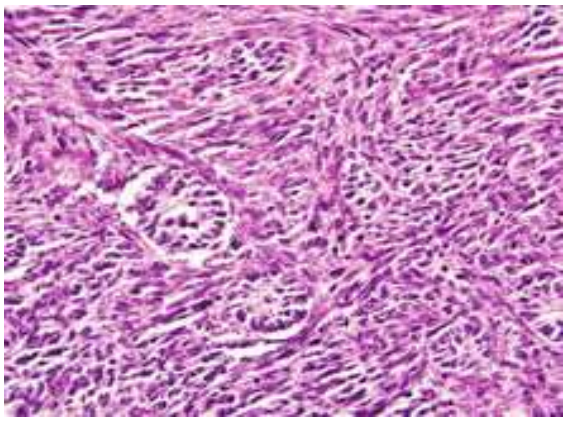

Figure 2: Microscopy: Highly cellular tumor composed of uniform spindle cells arranged in sheets and intersecting fascicles with scattered nests and cords of relatively uniform large cells with inconspicuous nucleoli

Discussion:

Ovarian fibroma is derived from the connective tissue of ovarian cortex. This group of ovarian neoplasm is rare. Based upon data from the Surveillance Epidemiology and End Results (SEER) United States national cancer database from 1992 to 1999, the overall incidence of this neoplasm was 0.2 per 100,000 women. Salmon in 1934 described the association of pleural effusion with benign pelvic tumors. In 1937, Meigs' and Cass described 7 cases of ovarian fibromas associated with ascites and pleural effusion [12]. This syndrome has been named after Meigs' and must fulfill the minimal criteria of pleural effusion, solid ovarian tumor and clearing of effusion after removal of the tumor [12]. The ascitic fluid collection, related to benign ovarian tumor is thought to be caused by excessive transudate from the tumor surface in a degree that the peritoneum cannot absorb [13]. Early ovarian cancer-associated symptoms constitute a constellation of mostly nongynecological complaints, suggesting a visceral disturbance, which do not point immediately to a pelvic origin. Abdominal bloating and pain predominate with recent onset and multiple symptomatic episodes [14]. Meigs' syndrome may be suspected when faced with an important pleural effusion, a very elevated CA-125 serum level, a negative cytologic examination of the ascitic effusion and no peritoneal implant on CT-scan" [15]. Cellular fibromas are predominantly solid. Cystic areas are usually small and without multiloculation. But "some fibromas undergo prominent cystic degeneration [16]. Irving et al. [13] studied 75 cases of cellular fibromas of the ovary and suggested the term "mitotic active cellular fibroma (MACF)" to cellular fibromatous neoplasm with bland cytology with ≥4 mitosis/10HPF. Carcino Embryonic Antigen (CEA) levels are often elevated in patients with ovarian cancer, but the test is too nonspecific and insensitive to have much use in the management of ovarian neoplasm [9]. The level of CA125, a surface glycoprotein associated mullerian epithelial tissue, is elevated in about 80% of patients epithelial ovarian cancers, particularly those with nonmucinous neoplasm [5,9]. A test called OVA-1is meant to be used as tumor marker in ovarian cancer [9]. The FDA (Food and Drug Administration) has approved this blood test called OVA1 [9]. The OVA1panel measures the level of five proteins in the blood. These biomarkers include: transthyretin, beta 2-microglobulin, apolipoprotein a1, CA125 cancer antigen and transferrin. The results of these lab tests converts an algorithm which helps indicate whether the ovarian mass is malignant [9]. HE-4 is another ovarian tumor marker can be used like CA125 to guide treatment. OVA-1 marker test should not be used for screening of ovarian cancer but may be used to determine the need for surgery in patient presenting with an ovarian mass [9].

Treatment:

Surgery is the recommended treatment for ovarian fibroma. Salpingo-oophorectomy can be considered in perimenopausal and post menopausal women and only cystectomy may be performed in younger women, preferably those who have not completed family. Some comparative analysis of the outcomes between the laparoscopy group and the laparotomy group showed that laproscopic surgery has the advantage. With the advances in operative instruments, and the techniques, laparoscopic surgery has become more popular, and it is being used frequently by many gynaecologists. The fibroma after morcellating can safely be removed through 12-mm trocar site [5,9].

Conclusion:

Ovarian fibroma a sex cord stromal tumor though uncommon can pose diagnostic difficulties mimicking malignancy. Our case report emphasizes the unique gross morphology with bare histological findings emphasizing the role of histopathologic diagnosis as gold standard. Surgical removal is recommended because of the low probability of malignant transformation.

Take home message:

Any enlargement of post menopausal ovary requires immediate investigation, with women at high risk for ovarian cancer need to undergo screening tests. A close follow up of the patients should be done as

hyperoestrogenimia may predispose to endrometrial carcinoma.

Acknowledgement

All individuals who participated in the study are gratefully acknowledged. The author is also grateful to authors/editors/publishers of all those articles, journals and books from where the literature for this article has been reviewed and discussed.

Source of Funding: Nil
Source of Conflict: Nil

References:

1. Philip J., Disaia, William T., Cancers of the female reproductive tract. In: Creasman, eds. Clinical Gynecologic Oncology. 7th ed. UK: British Library, Mosby Elsevier; 2007: 294-238.
2. Sheila Jorge Adad, Valeria Limb Laterza, Carlos David Teixeira dos Santos et al. "Cellular Fibroma of the Ovary with Multiloculated Macroscopic Characteristics: Case Report". Case Reports in Medicine. 2012;2012:5.
3. Foundations in Diagnostic Pathology. In: Nucci, Marisa R, Oliva, Ester, Goldblum, John R., eds. Gynecologic Pathology. 1st ed. New York: Elsevier Churchill Livingstone; 2009: 446-448.
4. J. Prat and R. E. Scully, "Cellular fibromas and fibrosarcomas of the ovary: a comparative clinicopathologic analysis of seventeen cases," *Cancer*, vol. 47, no. 11, pp. 2663–2670, 1981.
5. J. A. Irving, A. Alkushi, R. H. Young, and P. B. Clement, "Cellular fibromas of the ovary: a study of 75 cases including 40 mitotically active tumors emphasizing their distinction from fibrosarcoma," *American Journal of Surgical Pathology*, vol. 30,no. 8, pp. 929–938, 2006.
6. Young RH, Scully RE: Ovarian stromal tumors with minor sex cord elements: a report of seven cases. *Int J Gynecol Pathol* 1983, 2:227-34.
7. Zhang J, Young RH, Arseneau J, Scully RE: Ovarian stromal tumors containing lutein or Leydig cells (leuteinized thecomas and stromal Leydig cell tumors): a clinicopathological analysis of fifty cases. *Int J Gynecol Pathol* 1982, 1(3):270-85
8. Byrne P, Vella EJ, Rollason T, Frampton J: Ovarian fibromatosis with minor sex cord elements. Case report. *Br J Obstet Gynaecol* 1989, 96(2):245-248.
9. Berek & Novak's Gynecology. In: Berek Jonathan S., Berek Deborah L., eds. Berek & Novak's Gynecology. 15th ed. Philadelphia, PA: Lippincott Williams and Wilkins; 2012: 1264.
10. Mandal S, Mahajan D, Roy S, Singh M, Khurana N: Fibroma with minor sex cord elements – an incidental finding in a normal sized ovary A case report with literature review. *Diagn Pathol* 2007, 2:46.
11. Meigs JV, Cass JW, "Fibroma of the ovary with ascites and hydrothorax with report of seven cases".Am J Obstet Gynecol 1937, 33: 249-266.
12. Abad A, Cazorla E, Ruiz F, Aznar I, Asins E, Llixiona J. "Meigs' syndrome with elevated CA 125: case report and review of literature" Eur J Obstet & Gynecol reprod Biol" 1999, 82: 97-99.
13. Fuji M, Okino M, Fujioka K, Yamashita K, Hamano K. "Psuedo Meigs' syndrome caused by breast cancer metastasis to both ovaries" Breast cancer 2006, 13: 344-348.
14. Samanth KK, Black WC 3 "Benign ovarian stromal tumors associated with free peritoneal fluid" Am J Obstet Gynecol 1970; 107: 538-45.
15. Nemeth AJ, Patel SK. "Meigs syndrome revisited" J Thorac Imaging 2003; 18:100-103.
16. Frederick Rand Ueland, Andrew John Li. Serum biomarkers for evaluation of an adnexal mass for epithelial carcinoma of the ovary, fallopian tube, or peritoneum, 2013. Available at: URL: http//:www.Wolters Kluwer health/ up to date medicine. Accessed 17 April 2013.