Case Report

Ovarian Fibrosarcoma: Case Report of a Rare Lesion

Ebunoluwa S. Daramola, Olubanji A. Oguntunde[1], Lateef A. Odukoya[2], Kehinde S. Okunade[3], Adeyemi A. Okunowo[3], Adekunbiola A. F. Banjo[1], Rose I. Anorlu[3]

Departments of Obstetrics and Gynaecology and [2]Anatomic and Molecular Pathology, Lagos University Teaching Hospital,
Departments of [1]Anatomic and Molecular Pathology and [3]Obstetrics and Gynaecology, College of Medicine, University of Lagos, Lagos, Nigeria

Abstract

Ovarian fibrosarcoma is an extremely rare ovarian neoplasm and much rarer in the young. We present the case of a 35-year-old Nigerian woman who presented with ovarian mass with histologic and immunohistochemical profile in keeping with fibrosarcoma. She had an uneventful surgery and had postoperative chemotherapy with a combination of carboplatin and paclitaxel. She is currently on follow-up at the gynecology clinic.

Keywords: Fibrosarcoma, immunohistochemistry, ovary

Introduction

Fibrosarcomas are malignant neoplasms of fibroblasts. Fibrosarcomas can arise from superficial and deep connective tissues such as fascia, tendon, periosteum, and scar. They can grow slowly or rapidly and often appear well circumscribed.[1] Occurrence of this neoplasm in the ovary is rare and <100 cases have been reported since 1981 when a diagnostic criterion was established.[2] This is unlike the other tumors that arise from the ovarian stromal fibroblasts such as fibromas and thecomas, both of which account for about 4% of all ovarian neoplasms.[3] It is usually encountered in the peri/postmenopausal age group, but can also occur in other age groups. Clinically, it presents with an undefined symptomatology such as pelvic pain, abdominal mass, and heaviness in the lower abdomen.[4]

Ovarian fibrosarcomas are extraordinarily rare. The criteria for differential diagnosis are not too clearly defined, especially for differentiating between mitotically active fibromas and fibrosarcomas.[2] Immunohistochemistry (IHC) markers such as Ki-67 (MIB-1), vimentin expression, and nonexpression of CD 117 are important diagnostic markers.

These lesions are therefore prone to misdiagnosis especially in resource-poor settings where IHC may be unavailable or unaffordable.[2] When correctly diagnosed, it is not only difficult to treat patients with ovarian fibrosarcoma, but also most patients with this disease do not survive for >2 years due to early metastasis via the bloodstream or tumor recurrence.[5]

Access this article online	
Quick Response Code:	Website: www.atpjournal.org
	DOI: 10.4103/atp.atp_43_18

The rarity of this type of ovarian malignancy especially at the age of the presented patient, diagnostic difficulty in areas of practice where IHC may be unavailable, and attendant management dilemma prompted this case report.

Case Report

A 35-year-old-single nulliparous woman presented at the gynecological outpatient clinic of a tertiary hospital in Nigeria, following referral from a peripheral health center with 3-week history of left-sided lower abdominal pain and generalized abdominal swelling. There were associated significant weight loss, bloating, and lower back pain. There were no changes in urinary and bowel habits apart from early satiety. She had no family history of breast, ovarian, endometrial, or colonic cancer. She attained menarche at the age of 14 years and coitarche at the age of 23 years. There was no history of contraceptive use. Physical examination revealed a young and emaciated woman who was not in any distress. Pelvic organs were difficult to delineate on palpation. Abdomino-pelvic computed tomography scan done about a week prior to presentation revealed a left ovarian tumor (136 mm × 81 mm),

Address for correspondence: Dr. Olubanji A. Oguntunde,
Department of Anatomic and Molecular Pathology,
College of Medicine, University of Lagos, PMB 12003, Lagos, Nigeria.
E-mail: bjbanji@gmail.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Daramola ES, Oguntunde OA, Odukoya LA, Okunade KS, Okunowo AA, Banjo AA, *et al*. Ovarian fibrosarcoma: Case report of a rare lesion. Ann Trop Pathol 2019;10:90-2.

right cystic adnexal mass (34 mm × 26 mm) separate from the ovary, massive ascites, and tumor seedlings involving the omentum, small bowels, and peritoneum. Results of tumor markers showed elevated CA-125 (320 IU/mL) but normal carcinoembryonic antigen (1.2 μg/L), β-human chorionic gonadotropin (<5 mIU/mL), and lactate dehydrogenase (251 U/L) assays. A working diagnosis of bilateral ovarian tumor suspicious of malignancy was made.

The patient subsequently had staging laparotomy and primary debulking surgery (total abdominal hysterectomy, bilateral salpingo-oophorectomy, and infracolic omentectomy) following intraoperative frozen section analysis suggestive of fibrosarcoma. Intraoperative findings were as follows: 3 L of straw-colored fluid; bilateral complex ovarian masses (left 14 cm × 10 cm and right 8 cm × 6 cm); widespread tumor deposits on the uterus, bladder, peritoneal surfaces, small and large bowels, liver, subdiaphragmatic surfaces, and omentum. All specimens were sent for histopathological and cytological evaluation.

Grossly, the left and right ovaries measured 11.0 cm × 6.0 cm × 3.0 cm and 7.0 cm × 6.0 cm × 3.0 cm, respectively. Their capsule was breached and cut sections through the ovaries showed partly grayish white-to-necrotic solid surfaces and partly cystic surfaces. Histology of the bilateral ovarian masses was reported as showing a cellular neoplastic lesion composed of sheets and intersecting bundles of spindle-shaped cells having hyperchromatic-to-vesicular nuclei and eosinophilic cytoplasm with indistinct cell borders and characteristic herringbone growth pattern. The tumor cells showed abnormal mitoses of 4–5 per high-power field (HPF). There were also foci of necrosis. The omental and peritoneal seedlings showed sheets of malignant spindle cells similar to those in the ovaries. A histological diagnosis of malignant sex cord-stromal tumor was made to rule out fibrosarcoma. To further confirm the diagnosis, a tissue block was sent to a referral center in the United Kingdom where IHC was further done which showed strong uniform expression of CD34 and tumor cells completely negative with DOG1, desmin, S-100, CD10, broad-spectrum cytokeratins (AE1/3), epithelial membrane antigen (EMA), and neurofilament protein [Figure 1]. A conclusion of immuohistochemical features in keeping with fibrosarcoma was made, especially with the characteristic herringbone architectural pattern.

She had an essentially uneventful postoperative period and eventually had her first course of chemotherapy which comprised a combination of carboplatin (300 mg/m^2) and paclitaxel (175 mg/m^2). After about 5 weeks postoperatively, she was clinically stable and repeat CA-125 level was found to be 178.1 U/mL, 96 U/mL, and 47 U/mL after the first, second, and third courses of chemotherapy, respectively. This downward trend of CA-125 continued up until the 7th course of chemotherapy (31.6 U/mL). Repeat abdominal scan done at this point revealed minimal ascites but no peritoneal or visceral organ tumor deposits. She is currently being followed up at the gynecology outpatient clinic.

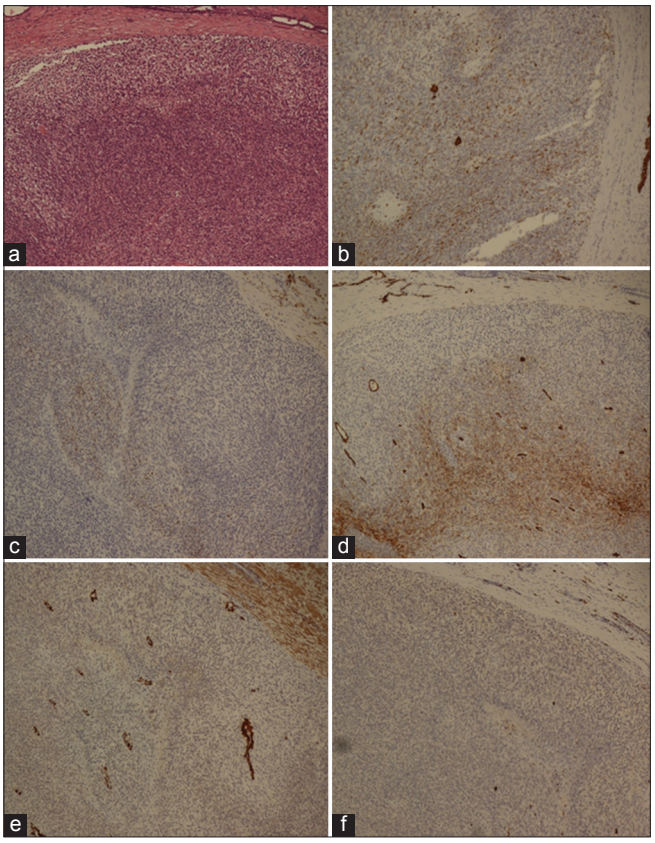

Figure 1: (a) Objective (H and E, ×10). (b) Objective (AE13, ×10). (c) Objective (desmin, ×10). (d) Objective (CD34, ×10). (e) Objective (SMA, ×10). (f) Objective (S100, ×10)

Discussion

Globally, epithelial ovarian cancers still contribute the majority of the disease burden (about 80%–90%), with germ cell tumors and sex cord stromal tumors being relatively uncommon in comparison. Rarer still is the histologic diagnosis of primary ovarian fibrosarcoma detected in the index patient, which is the first of such to be reported in our center. Less than 100 cases have been reported.[2] In the vicinity, Omotoso *et al*. in their study in Calabar in 2016 reported finding one case of ovarian fibrosarcoma and Buhari *et al*. recorded a case of an unclassified sarcoma in an 85-year-old woman in Ilorin.[6,7]

Ovarian fibrosarcoma can occur at any age, with results from a 15-year multicentric retrospective study from China giving a median age of 49 years (range: 20–73 years).[5] The index patient was 35 years old when she presented.

Criteria for the diagnosis of fibrosarcoma include mitotic count. This is considered the most important feature for distinguishing between benign and malignant lesions. Lesions with <4 mitotic counts per HPF were considered

cellular fibromatous lesions. These were shown to have better prognosis than those with ≥4/10 HPF.[8] Our patient had a histology that indicated tumor cells showing abnormal mitosis (4–5/HPF) with foci of necrosis and omental spread. Poor prognostic factors of fibrosarcoma highlighted include high grade, high cellularity with minimal collagen, mitotic rates >20/10 HPF, and necrosis.[9] Fibrosarcomas are typically large tumors that have often spread beyond the ovary at diagnosis as was the case in our patient. The differential diagnoses include leiomyosarcoma, a gastrointestinal stromal sarcoma, and various types of primary or metastatic soft-tissue sarcomas. Metastases were ruled out based on the clinical and radiologic findings. Immunophenotyping of fibrosarcomas is positive for vimentin and very focally for smooth muscle actin, representing myofibroplastic differentiation. Some cases arising from dermatofibrosarcoma or solitary fibrous tumor are CD34 positive.[9] In doubtful cases, high Ki67 (MIB-1) expression is consistent with a malignant diagnosis despite low visual mitotic rates.[5] The expression or nonexpression of other immunohistochemical markers such as vimentin, CD117, SMA, desmin, EMA, S-100, CD99, CD34, inhibin alpha, estrogen receptor, and progesterone receptor in addition to the characteristic herringbone morphologic pattern is contributory in establishing a diagnosis.[4]

This was one of the cases where the intraoperative frozen section was helpful, especially considering the age of the index patient. The frozen section machine is however frequently under repair which in itself poses a limiting factor to its use.

Treatment strategies to improve patient outcome for patients diagnosed with ovarian fibrosarcoma have not been clearly defined.[5] As a result, most patients experience fatal outcome due to early metastasis via the bloodstream and tumor recurrences that usually occur within 2 years of diagnosis.[5] Surgical resection for ovarian fibrosarcoma has included simple adnexectomy and total hysterectomy with bilateral adnexectomy and omentectomy. In many cases, postsurgical adjuvant chemotherapy or radiation is also required.[5] The index patient had primary debulking surgery with adjuvant chemotherapy with carboplatin and paclitaxel and is currently being followed up, with imaging and CA-125 as the tumor marker of choice.

Conclusion

Although primary ovarian fibrosarcomas are rare, they must be considered in the differential diagnosis, especially of a unilateral, heterogeneous, solid ovarian lesion. Despite being usually encountered in the peri/postmenopausal age group, they can occur at any age as seen in our case. They are often underdiagnosed preoperatively, resulting in diagnostic dilemmas. Accurate diagnosis should involve strict adherence to intraoperative surgical excision protocols, liberal use of immunohistochemical assessment of ovarian tumors, and effective postoperative management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Goldblum JR, Lamps LW, McKenney JK, Myers JL, editors. Soft tissues. Rosai and Ackerman's Surgical Pathology. 11th ed., Ch. 41. Philadelphia: Elsevier; 2018. p. 1810-915.
2. García Jiménez A, Castellví J, Pérez Benavente A, Díaz de Corcuera Frutos I, Ramón Y Cajal S. Ovarian fibrosarcoma: Clinicopathologic considerations about the intraoperative and post-surgical procedures. Case Rep Med 2009;2009:802817.
3. Kumar V, Abbas AK, Fausto N, editors. Robbins and Cotran Pathologic basis of disease. The female genital tract. 7th ed., Ch. 22. Philadelphia: Elsevier; 2005. p. 1092-99.
4. Grauso F, Messalli EM, Salzillo ME, Di Martino L, Falcone F, Orabona P, *et al.* Ovarian fibrosarcoma: Case report and latest trends in diagnostic and therapeutic management. Eur J Gynaecol Oncol 2015;36:742-5.
5. Huang L, Liao LM, Wang HY, Zheng M. Clinicopathologic characteristics and prognostic factors of ovarian fibrosarcoma: The results of a multi-center retrospective study. BMC Cancer 2010;10:585.
6. Omotoso AJ, Odusolu P, Ikpeme A, Irabor G, Nnoli M. Ovarian tumour in a tertiary institution in South Southern Nigeria. Int J Multidiscip Res Inf 2016;2:398-401.
7. Buhari MO, Ojo BA, Ijaiya MA, Aboyeji PA. Ovarian cancers in Ilorin, Nigeria-a review of over 80 cases. Nig Q J Hosp Med 2005;15:127-30.
8. Ray S, Biswas BK, Mukhopadhyay S. Giant primary ovarian fibrosarcoma: Case report and review of pitfalls. J Cytol 2012;29:255-7.
9. Mathur C, Agarwal M, Kothari DC, Tailor SB. Primary Ovarian Fibrosarcoma–A Rare Case Report. IOSR J Dent Med Sci (IOSR-JDMS) 2016;1:65-8.