A giant primary ovarian fibrosarcoma in a South Sudanese patient

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ABSTRACT

Ovarian fibrosarcomas account for less than 1% of all ovarian malignancies. Clinical diagnosis is extremely difficult. A 20-year-old, illiterate, married nulliparous woman presented to our Outpatient Department Clinic (OPD) with chronic abdominal distension. Ultrasound and CT scan revealed a very large intra-abdominal mass, 23.8x27.8x35.7cm. She underwent a laparotomy and left salpingo-oophorectomy with her uterus and right adenexum conserved. After surgery, the mass weighed 11.1kg and measured 43x38x35cm. Histological findings were in line with a giant primary ovarian fibrosarcoma. To date, as far as we are aware, this is the largest ever recorded primary ovarian fibrosarcoma.

Key words: giant primary ovarian, fibrosarcoma, exploratory laparotomy, salpingo-oophorectomy, South Sudan.

Introduction

Ovarian fibrosarcomas are exceptionally uncommon neoplasms contributing to less than 1% of all ovarian malignancies.¹ They are of fibroblast origin with slow or rapid growth and a well circumscribed appearance.² There are less than one hundred cases reported in the literature.³ They are seen more in the peri or postmenopausal age groups but can occur in other age groups. Furthermore, they present with nonspecific symptoms like abdominal mass, pelvic pain, and heaviness in the lower abdomen.⁴ This makes clinical diagnosis difficult. Diagnosis is mainly based on pathological and immunochemistry investigation. We present a case with a very large ovarian fibrosarcoma in a young married nulliparous woman.

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Case Report

A 20-year-old, nulliparous housewife was referred from the Gynaecological Department to the Surgical Department at Juba Teaching Hospital, Juba, which is one of the main referral hospitals in South Sudan.

She presented with a one-year history of abdominal swelling of a gradual onset. The swelling started from the lower abdomen and gradually reached the epigastric region with extension to both flanks. This was associated with nausea, loss of appetite, weight loss but no abdominal pain, vomiting, diarrhoea, and constipation. She did not notice any swelling of her lower limbs despite the size of the abdominal swelling.

There was no family history of breast, ovarian, uterine, endometrial, colonic or any other known cancers. Her menarche was at 14 years of age with a regular 28 days cycle. However, she started experiencing irregular menstrual cycles at about 3-4 months before presentation. There was no history of abortion or miscarriage and neither was she using any contraceptive pills.

Physical examination showed no cyanosis or jaundice. The abdomen was uniformly distended with prominent striae (Figure 1) and an everted umbilicus. The abdominal mass extending from the pelvic region to the epigastric region, firm and difficult to delineate the edges. There was no tenderness or shifting dullness on percussion. Normal bowel sounds were heard only at the flanks.

The differential diagnoses were mesenteric cyst, hydatid cyst, ovarian cyst and ovarian tumour. Other differential diagnoses include uterine fibroids, appendiceal mass, abdominal malignancy of uncertain location. Pregnancy test was negative.

An abdominal ultrasound showed a large solid abdominopelvic mass most likely of uterine origin, uterine malignancy being a possibility. CT scan with contrast showed a large, abdominopelvic, lobulated, heterogeneous, isohypodense soft tissue measuring 23.8x27.8x35.7 cm (Figure 2). Laboratory investigations showed normal electrolytes, renal function tests, liver function tests, full blood count, and normal blood glucose level. She tested for positive HIV. There were no facilities for tumour markers such CA125, Ca119, and CEA.

The patient was counselled and she consented to surgery the following day. An exploratory laparotomy with left salpino-oophorectomy was carried out with no intraoperative complications. A large, fleshy, smooth surface, vascularised, lobulated ovarian mass involving
left ovary was removed in one piece (Figures 3 and 4). It weighed 11.1kg and measured 43x38x35cm. This was sent for histological examination.

The non gravid uterus looked normal with right normal ovary and tube. There were no ascites, the omentum and the peritoneal surfaces looked normal.

The patient had an uneventful recovery from surgery and satisfactory post operative period. She was prescribed ceftriaxone 1g every 12 hours for 3 days and simple analgesia. On the third day she was discharged in good health.

Pathological section (Figure 5) showed a sectioned tumour, mostly grey-white, smooth, and lobulated, with focal haemorrhagic and necrotic areas. There was a separate fallopian tube tissue with fimbria measuring 57x5mm with tan grey colour.

Histopathological results revealed a neoplastic proliferation arranged in a herringbone pattern formed of highly cellular fibroblastic proliferation. The fibroblastic cells had elongated tapering dark staining nuclei with granular chromatin, scant pale eosinophilic cytoplasm and abundant mitotic figures. The stroma was formed of variable collagenous material. The fallopian tube consisted of mucosa thrown into plicae lined by stratified columnar epithelium with round to oval nuclei and moderate eosinophilic cytoplasm. Also seen were foci of hyaline degeneration and haemorrhages. There was no giant cell or significant cellular pleomorphism.

Discussion

Ovarian fibrosarcomas are exceptionally uncommon neoplasms contributing to less than 1% of all ovarian malignancies.[5] Ovarian fibrosarcoma is one of the members of ovarian fibroid tumours originating from sex-cord stromal tumours. Sex-cord stromal tumours contribute to 4.3% of ovarian tumours.[5]

Most of the ovarian fibrosarcomas are seen at peri and post-menopausal women with a mean age of 49 years.[5] In this case, we presented a 20-year-old woman with primary ovarian fibrosarcoma. This seems to be the youngest in comparison to other case reports[5] where the reported ages range from 41 to 76 years. Ray S et al reported[6] a case in a patient who was 23 years old.

It has been observed that patients normally present with non-specific symptoms such as pelvic mass or abdominal pain.[5] In this case, the patient presented with a painless, large abdominal mass causing abdominal distension. Eventually, as the mass grew into the epigastric region, she started to feel the discomfort and disfiguration which prompted her to seek medical attention. Our case is unique in terms of the young age of the patient, absence of ascites despite the huge ovarian mass, as well as the weight of the ovarian mass.

The sizes of ovarian fibrosarcomas vary from one case report to another. In the literature review conducted by Tingting et al.[5] ovarian fibrosarcoma sizes ranged from 5 to 23cm. However, Daramola et al[2] presented a case of size 13.6x8.1cm. There is one case report[6] of a giant fibrosarcoma measuring 25x17x12cm as the largest recorded. Our case measured 23.8x27.8x35.7cm. All these measurements were based on CT scan images. On a different note, gross measurement of our tumour showed a size of 43x38x35 cm. We believe our tumour to be the
largest so far reported and weighed 11.1kg. This may also be the heaviest registered tumour. The likely reason might be the delay in accessing the healthcare service because the patient lived in a distant rural area. In addition, being immunocompromised (confirmed as HIV positive) might have enhanced the rapid growth of the tumour to the current large size. Furthermore, being illiterate and uninformed, as well as the absence of abdominal pain, might have contributed to her delay in seeking medical attention.

In addition to ultrasound and CT scan imaging investigations, the diagnosis of fibrosarcoma is based on histopathological analysis. This is achieved by microscopic observation of growth patterns, cellular atypia, and mitotic counts. Classification and determination of the tumour is based on the mitotic counts in which counts <3/10 high power field (HPF) are considered as benign while counts >4/10HPF are fibrosarcomas. However, the size of the tumour, growth rate as well as the immunohistochemical markers like Ki-67 should be included in classification of ovarian fibrosarcomas. Ki-67 plays a key role as a proliferative activity index for these tumours. In South Sudan there is no histopathological laboratory that provides such services which include counts of mitoses.

The important laboratory test was the positive HIV test. A literature review conducted by Bhatia et al. showed that there is an association between immunosuppression by HIV and sarcoma subtypes such as leiomyosarcoma, angiosarcoma, and fibrohistiocytic tumours. This might be the reason why the tumour increased rapidly to a large size. A chest X-ray would have ruled out a pleural effusion which might suggest Meig’s syndrome or distant metastasis of ovarian cancer. CT scan of chest would have been an alternative.

The most desirable therapy for fibrosarcoma is surgical excision of the tumour. This can include removal of the ovarian fibrosarcoma with simple adnexectomy with or without total hysterectomy and total omentectomy. This depends on how much the surrounding tissues are involved. In our case, we did a total excision of the left ovary with salpingo-oophorectomy. The other nearby tissues were not involved. In view of the age and parity of the patient, fertility sparing surgical treatment is acceptable provided adequate follow up visits are arranged.

Usage of radiotherapy and chemotherapy for fibrosarcoma is very controversial. The response rate of fibrosarcoma is considered very low to radiotherapy and chemotherapy when used as a neo-adjuvant and/or adjuvant tumour treatment. In our case, the tumour was very large and there were no metastatic focal areas. Moreover, there is no Oncological centre in South Sudan. Most of the patients who need chemotherapy are managed locally by importing the required chemotherapeutic agents from neighbouring countries. Alternatively, patients are referred to the nearby centres in Uganda, Kenya, and Sudan. Patients with advanced ovarian fibrosarcoma usually benefit from chemotherapy. The commonly used drugs include anthracyclines as the first line treatment. This is combined with doxorubicin or actinomycin D with Ifosfamide. The risk of developing multidrug resistance (MDR) is high if vincristine, actinomycin D, vinblastine and etoposide are used with the first line chemotherapeutic agent, doxorubicin. Another report states carboplatin and paclitaxel after primary debulking surgery. Radiotherapy could be of benefit in some cases; however, these services are not available in the country.

We plan to follow-up this patient for early detection of any recurrence of the malignancy. She was advised to attend follow-up and treatment for her HIV.

Conclusion

Primary ovarian fibrosarcoma remains a rare and challenging condition. Management of ovarian fibrosarcoma poses a very serious challenge especially when patients present at late stages in a low resource setting such as South Sudan. Surgical removal of the tumour remains the accepted standard procedure. Presence of imaging facilities as well as pathological services has helped us in the management of this condition. More is required to include immunochemistry analysis as part of pathological diagnostic services. There is a need to establish an Oncology Centre with appropriate human resources as well as modern equipment for better care of cancer patients in South Sudan.

Finally, scaling up health education in different media outlets will increase community awareness of cancer and early presentation to healthcare services at an earlier stage to expedite interventions.

Declaration of patient consent: The authors declare that patient consent was obtained which included usage of the images and other relevant information to be used in any journal or academic activities for educational purposes. The patient understood that her name or initials should not be published.

Conflict of interests: None.
Case Report

Contributions: IR and LG were the operating surgeons assisted by SA as registrar for general surgery. SA did the clerking, examination, investigations and preparing the patient for surgery as well as postoperative follow up. KS, JT and JW prepared and proof read the manuscript. All authors read and approved the manuscript.

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References


