Case Report

Giant Primary Ovarian Leiomyoma with Cystic Degeneration and Calcification Complicated by Post-Surgical Acute Pulmonary Embolism in a 23-year-old Nulligravida: A Case Report

[1]Olalekan Abisola Ajayi, [2]Olufemi Oladipupo Adebawojo, *[1]Victor Chimezie Okebalama.
[1]Department of Histopathology, Babcock University Teaching Hospital, Ilisan-Remo, Ogun State, Nigeria.
[2]Department of Obstetrics and Gynaecology, Babcock University Teaching Hospital, Ilisan-Remo, Ogun State, Nigeria.

Abstract

Leiomyomas of primary ovarian origin are rare, benign, smooth muscle neoplasm constituting just 0.5-1% of the entire tumours of the ovary. They are often of small size, unilateral and occur predominantly in premenopausal women, usually diagnosed incidentally, aided by examination of the pelvis, post-surgery histopathological evaluation and confirmed by immunohistochemical staining. Additionally, primary ovarian leiomyoma of this size (with the greatest diameter of 29.5cm) has not been reported in the literature. It is not uncommon for large pelvic tumours to be associated with venous thromboembolic events (VTE). We hereby present a case of a 23-year-old nulliparous woman who presented with complaints of abdominal pain and swelling. Clinical examination revealed an abdominal mass of 34 weeks' size. Abdominal ultrasonography showed an intra-abdominal mass that extended to the pelvis and a misdiagnosis of giant uterine leiomyoma was made. On laparotomy, a huge, right, well-encapsulated ovarian mass with a smooth surface was seen. Histopathological and immunohistochemical staining with desmin and vimentin confirmed it to be a giant primary ovarian leiomyoma with cystic degeneration and calcification. Additionally, the patient developed sudden onset of chest pain and breathlessness a day post-surgery for which urgent computed tomography pulmonary angiogram (CTPA) confirmed pulmonary embolism (segmental and post segmental regions bilaterally) and cardiomegaly. She was placed on low molecular weight heparin (Clexane) and oral warfarin which she responded well to Indeed, primary ovarian leiomyoma should be considered an important differential in primary solid ovarian and pelvic tumours. Also, in large or giant pelvic neoplasms, anticoagulant prophylaxis should be considered to prevent untoward pulmonary embolism and its possible fatal outcomes.

Keywords: Primary Ovarian Leiomyoma; Cystic Degeneration; Calcification, Pulmonary Embolism.

Introduction

Primary ovarian leiomyoma is an extremely rare, benign smooth muscle solid tumour that is often of small size, unilateral and presents with no symptoms. It is responsible for 0.5-1% of the entire benign neoplasms of the ovary and it's commonly seen in women between the ages of 20-60 years [1]. The majority of these neoplasms are found out unintentionally [2]

The tumours can originate from smooth muscle cells of hilar blood vessels of the ovary, undifferentiated germ cells, ligament of the ovary, stromal smooth muscle cells of the ovary or metaplasia of cortical smooth muscles [3]. They are often misdiagnosed as germ cell tumours especially when of large size because of their rarity [4].

Corresponding Author: *Victor Chimezie Okebalama
Department of Histopathology, Babcock University Teaching Hospital, Ilishan-Remo, Ogun State, Nigeria.
okebalsvictor@gmail.com

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Case Report

A 23year old nulliparous woman presented to our gynaecology clinic with a complaint of progressive abdominal swelling of 7 years' duration. The abdominal swelling was initially felt around the suprapubic area but grew progressively over the years to reach the epigastrium. There was associated pain which was mild and intermittent. There was no history of swelling in any other part of the body. There was no family history of uterine leiomyoma nor ovarian, breast or any cancer in her first-degree relatives. Menarche was at 12 years, she experienced irregular menstrual flow since the onset of swelling.

There was neither previous history of deep venous thrombosis nor a family history suggestive of thrombophilia.

On clinical examination, she was not pale, and her vital signs were essentially normal. The calculated body mass index (BMI) was 22.2kg/m^2. The abdomen was distended and revealed a 34 weeks' size palpable abdominal mass. The intra-abdominal mass was firm with a smooth surface, well-defined edges, not tender. The examining hand could get above but not below it. Pelvic ultrasonography done showed a hypoechoic mass in the abdomen which extended from the xiphisternum to the pelvis and was difficult to measure. The uterus and right adnexa were difficult to image. The left ovary with the adnexa appeared normal. The impression was a giant uterine leiomyoma. Her pre-operative Packed Cell Volume was 31.8%, her platelet count was 360,000 while her International Normalizing Ratio was 1.5. She was adjudged to have a moderate risk of having a VTE using the Modified Caprini VTE Risk Assessment.

She subsequently had laparotomy with right ovariectomy and partial salpingectomy. The surgery lasted for 100 minutes. Findings at surgery were a huge, right, encapsulated ovarian mass with a smooth surface (Figs 1 & 2). The right fallopian tube was inseparably adhered (plastered) to it. The mass weighed 5.07kg while the left ovary had a simple left ovarian cyst which had dimensions of 2.0 x 2.0 x 2.0cm containing clear fluid. The left tube was normal. The uterus looked normal with no evidence of uterine leiomyoma. The excised tissue was immediately fixed in 10% neutral buffered formalin. However, a day post-op, she developed sudden onset of cough, chest pain and breathlessness. Significant findings on physical examination were tachycardia, tachypnea with basal crepitations. Her SP02 was 86% in room air. A diagnosis of acute pulmonary embolism was suspected. She had urgent CTPA which confirmed the diagnosis of pulmonary embolism (bilateral- segmental and post segmental regions) with cardiomegaly. Her Prothrombin time (PT) was 16 seconds and Partial thromboplastin time with kaolin (PTTK) was 40 seconds while the international normalized ratio (INR) was 1.2. She was commenced on intranasal oxygen therapy at 5L/min. The patient was commenced on the thrombolytic, subcutaneous low molecular weight heparin (Clexane) and oral warfarin. The above symptoms resolved, and she was later discharged on oral warfarin. Follow-up visits were uneventful.

Gross pathological examination showed a well encapsulated, ovoid-shaped, greyish-white mass attached to the fallopian tube and weighed 4.5kg (Fig 3). The mass measured 29.5 x 19.5cm and 16.5cm in the widest dimension. The fallopian tube measured 6.5cm x 2.5cm. Cut sections showed greyish-white tissue with a whorled pattern and areas of cystic degeneration and calcification were seen (Fig 4). Cut sections of the fallopian tube revealed lumen with greyish white tissue.

Microscopically, sections reveal proliferating and interlacing smooth muscle fibres, the cells have ample eosinophilic cytoplasm and bipolar nuclei (Fig 5). The fibro collagenous stroma is hyalinized with areas of cystic degeneration and calcification (Fig 6). No cellular atypia, necrosis nor mitotic figures were present. The fallopian tube showed mucosa thrown into labyrinthine folds and lined by tall columnar epithelial cells. The tubal wall was edematous and contained congested vascular channels (Fig 7).

Immunohistochemically, the mass showed strong positivity to desmin and vimentin. The final diagnosis of giant primary ovarian leiomyoma was made histopathologically and confirmed through immunohistochemistry.

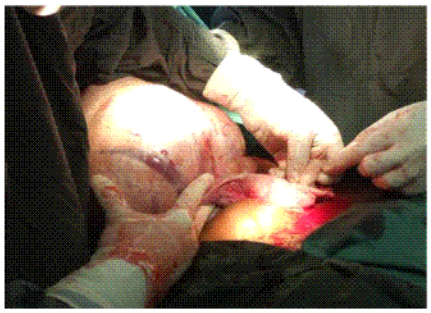

Figure 1: Intraoperative Finding: A huge, smooth, right-sided, well-encapsulated ovarian mass.

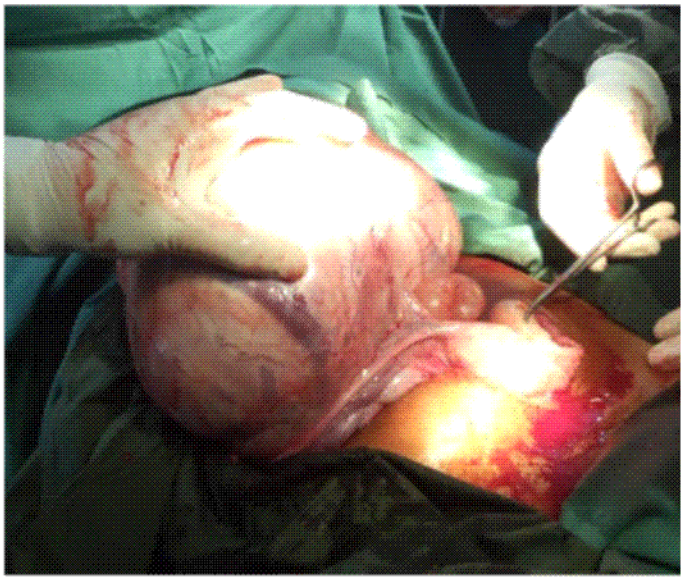

Figure 2: A huge, right-sided, smooth, well-encapsulated, solid ovarian mass found intraoperatively.

Figure 3: Macroscopic appearance of the ovarian mass with the adjoining fallopian tube

Figure 4: Macroscopic appearance: Cut sections showing greyish white soft tissue with a whorled pattern. Areas of cystic degeneration and calcifications are seen

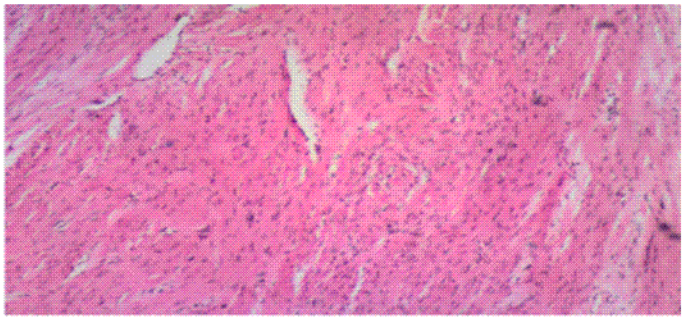

Figure 5:Microscopic appearance showing proliferating and interlacing bundles of smooth muscle fibres. (H&E x 40)

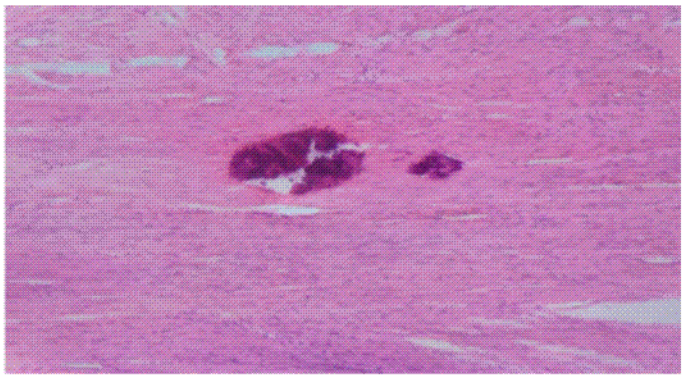

Figure 6: Microscopic appearance showing calcified areas

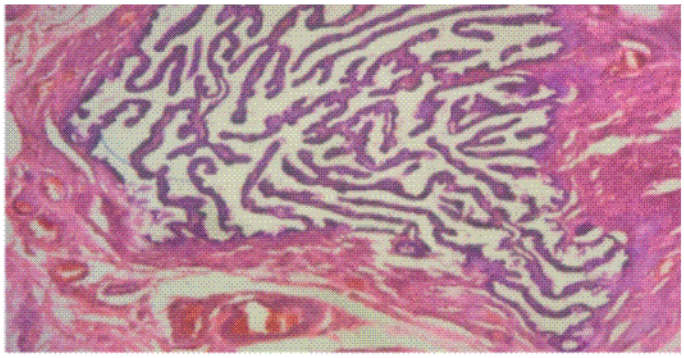

Figure 7:Microscopic appearance of the adjoining fallopian tube H&E x 100

Discussion

Primary ovarian leiomyoma is a very rare benign ovarian tumour. It accounts for 0.1-0.5% of the entire benign tumour of the ovary, with premenopausal women representing 80% of the patients while 16% of the cases are encountered in the post-menopausal age group [1, 2, and 5].

The majority of the cases are often unilateral with no preference for either of the ovaries, of small size <3cm but may be as big as 11kg [1, 5, 6]. Very few cases of huge primary ovarian leiomyoma have been reported in the literature. In most cases, primary ovarian leiomyoma (PLO) is asymptomatic, and detected, incidentally on

routine examination of the pelvis, surgery or at autopsy with diagnosis confirmed through histopathology and immunohistochemistry [7].

When present, clinical presentations which are usually variable and mostly seen in giant ovarian leiomyoma include abdominal distention, mass and pain, ascites, hydronephrosis, mild elevation of CA-125 and hydrothorax [1, 7, 8]. Radiologically, they are often not diagnosed preoperatively due to the paucity of radiological features that can be used to characterize them and their rarity [4]. Menstrual irregularity is rare in primary ovarian leiomyoma [4].

In line with the literature, the index patient was a premenopausal woman, had irregular menstrual flow since the onset of swelling. She presented with complaints of abdominal pain and swelling, and a definitive diagnosis could not be made before surgery even with pelvic ultrasonography but was only made post-surgically and confirmed with histopathological examination and immunohistochemical evaluation.

Doss et al [3] had reported the coexistence of the unilateral variety of ovarian leiomyoma with other lesions which could be contralateral or ipsilateral. In our patient, the contralateral ovary (left ovary) had a simple cyst. Uterine leiomyoma usually coexists with ovarian leiomyoma [1].In our case, the uterus was entirely normal and free of any leiomyomas as examined during surgery which suggests the lesion developed primarily from the ovary.

With uncertainty surrounding the histological origin of PLO, suggested sites of its origin include the smooth muscle in the walls of ovarian vascular supplies or that of ovarian ligament, cortical myofibroblast smooth muscles metaplasia, ovarian smooth muscle cells, myofibroblast, the stromal smooth muscle of the ovary, ovarian stromal undifferentiated germ cell and metaplasia of stroma of endometriosis [3, 8].

On macroscopy, PLO has smooth surfaces, a firm with oval or round shape. The cut section shows greyish surfaces with a whorled pattern. Microscopically, it reveals smooth muscle fibres. Present mainly in larger tumours are secondary changes which include hyalinization, cystic changes and calcification [5, 7] as was seen in our case.

The differential diagnosis of giant primary ovarian leiomyoma is several other spindle cell growths which include sclerosing stromal tumours, fibroma, thecoma, cellular fibroma, granulosa cell tumours, cellular thecoma, tumours of the Sertoli and legdig cells. Others are parasitic uterine leiomyoma, leiomyosarcoma, gastrointestinal stromal tumours with metastasis and carcinoma involving spindle cells [5].

With immunohistochemical evaluation, leiomyoma shows diffuse or strong positivity with desmin which differentiates it from other ovarian spindle cell tumours. Most leiomyomas also stain with vimentin. Fibromatous tumours stain negatively or show focal positive staining with desmin.

In differentiating PLO from leiomyosarcoma, criteria such as necrosis, cellular atypia and mitosis (mitotic count) are used [8 and 9]. None of the above criteria was seen in our case. The management approach is dependent on clinical presentations, patient's age, future pregnancy desire, closeness to menopause, tumour size, associated complications and malignancy risk [4].

Although ovarian preserving surgery can be considered in young patients, salpingo-oophorectomy with or without hysterectomy is the preferred surgical technique in premenopausal women with ovarian leiomyoma. Tumour size and malignancy risk are the factors considered in the choice between laparoscopy and laparotomy [8].

In our patient, because of factors such as age, desire for future conception, chronic abdominal pain and tumour size, we opted for laparotomy with right ovariectomy and partial salpingectomy.

Cases of VTE associated with pelvic tumours and gynaecological surgeries have been reported in the literature.[10] A clinical study of 498 cases undergoing gynaecological surgery revealed an incidence of 11.6% for deep venous thrombosis (DVT) while 10.3% of those that had DVT developed pulmonary embolism.[11] In addition, a case series have demonstrated the association between the presence of large uterine fibroids and VTE. Of note is that none of the patients reported had typical risk factors for a provoked VTE and they all had a negative hypercoagulable INR.[12] Our patient had a huge pelvic mass possibly compressing on the pelvic vessels predisposing her to a VTE. The other risk factor was that she had major gynaecological surgery. She was in a negative hypercoagulable state prior to surgery.

She developed pulmonary embolism a day after surgery before prophylaxis was commenced. Indeed, in an audit conducted to determine the risk profile and prophylaxis prescribed for venous thromboembolism in patients going for gynaecological surgeries, out of 109 patients audited, 45% had very high risk, 38% and 14% had high risk and moderate risk respectively while only 3% had no risk. Despite the risk profiling, 40% of the patient received no prophylaxis while 55% received inappropriate prophylaxis.[13] It is important for gynaecological units to have a formal risk assessment model to objectively categorize patients preoperatively and VTE prophylaxis guidelines readily available to ensure compliance.

Furthermore, our patient responded well to the thrombolytic, low molecular weight heparin (enoxaparin) and warfarin prescribed following the diagnosis of pulmonary embolism. She was discharged home on the 9th postoperative day to be followed up in the clinic. Our patient is doing well, 8 months following surgery.

Conclusion

Though extremely rare, primary ovarian leiomyoma should be considered a differential in pelvic and ovarian solid tumours.

However, immunohistochemical testing is essential for confirmatory diagnosis. PLO is usually small in size but can be large. In large PLOs, cystic changes and calcification can occur.

Additionally, anticoagulant prophylaxis should be considered in large, pelvic tumours, including giant PLO since such masses will likely compress the pelvic veins, thus increasing the risk of pulmonary embolism.

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