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Disseminated Peritoneal Leiomyomatosis: A Case Report of an Incidental Finding During an Emergency Caesarean Section and a Review of the Literature

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Authors' contributions

This work was carried out in collaboration among all authors. Author ABL designed the case study, wrote the protocol and wrote the first draft of the manuscript. Author AOO revised the manuscript and managed the literature search while authors NID and OF contributed the clinical history of the case. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Aim: To present the case report of an incidental finding of disseminated peritoneal leiomyomatosis (DPL) found during an emergency caesarean section in a young Nigerian woman who presented with slow progress in labour and maternal exhaustion.
Presentation of Case: We present the case of a 35-year-old unbooked primigravida with previous myomectomy who presented with maternal exhaustion and slow progress in labour and subsequently had an emergency lower segment caesarean section. At surgery, multiple firm to hard nodules of varying sizes were seen scattered throughout the peritoneum. Histological examination and immunohistochemical analysis of the nodules showed features consistent with disseminated peritoneal leiomyomatosis. Clinical and radiological follow-up for 1 year was uneventful.

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Discussion: DPL is a rare benign smooth muscle tumour that clinically and macroscopically simulates disseminated intra-abdominal or pelvic malignancy. It occurs predominantly in women of child bearing age and is mostly discovered incidentally. The occurrence of DPL in association with pregnancy and uterine leiomyomas was also corroborated in this index case as pedunculated and intramural uterine nodules were also seen during surgery.
Conclusion: Disseminated peritoneal leiomyomatosis is a rare benign disease which may be misdiagnosed as disseminated or metastatic intra-abdominal malignancy. Therefore, a high index of suspicion is required for accurate diagnosis and proper management.

Keywords: Leiomyoma; disseminated peritoneal leiomyomatosis; emergency caesarean section.

ABBREVIATIONS

DPL : Disseminated Peritoneal Leiomyomatosis
LPD : Leiomyomatosis Peritoneal Disseminata

1. INTRODUCTION

Disseminated peritoneal leiomyomatosis, also known as leiomyomatosis peritonealis disseminata (LPD) is a rare but benign lesion [1]. It is characterised by multiple intra-abdominal nodules composed of proliferating smooth muscle cells [2]. LPD was first described by Willson and Peale in 1952 and eventually named by Taubert in 1965 [2,3].

Most LPD cases occur in reproductive age women, but a few cases occur in postmenopausal women and in men [2]. Although both the incidence and possible pathophysiology of this disease condition remains unclear, the possible causes have been divided into sub-categories including hormonal, genetic, metaplasia of sub-peritoneal mesenchymal stem cells, and iatrogenic [4]. The oestrogen and progesterone receptor positivity that is frequently seen in LPD supports the role of hormones in the aetio-pathogenesis, furthermore, it is also generally associated with high levels of exogenous and endogenous female gonadal steroids [4,5].

LPD usually causes widespread nodules in the abdominal cavity that frequently exhibits clinical and radiological symptoms mimicking some malignant neoplasms [3]. It is most often an incidental finding usually during laparotomy or pelvic surgeries for other disease conditions. Thus, the incidence quoted in literature may be far lower than the actual incidence because most cases may be asymptomatic and never diagnosed. While about 140 cases have been described in the English literature alone, only 3 cases have been reported so far from Nigeria to the best of our knowledge [2,6,7,8].

Here, we present a case of disseminated peritoneal leiomyomatosis in a young Nigerian woman discovered incidentally during an emergency lower segment Caesarean section which was performed due to maternal exhaustion and poor progress in labour.

2. CASE PRESENTATION

The patient described here is a 35-year-old unbooked primigravida who presented with maternal exhaustion and slow progress in labour at an estimated gestational age of 42 weeks. There is previous history of myomectomy 5 years prior to the index pregnancy. She had no history of any other chronic illness or co-morbidity. She also had no family history of malignancy or other chronic illnesses. There was also no history of chronic medication use. There was no history of use of oral contraceptive pills or hormonal supplements in the past.

On examination at presentation, she was in intermittent painful distress with poor uterine contractions. Foetal heart sounds were heard and regular. Perineal examination revealed a 6cm dilated cervix and an assessment of active phase labour was made. Augmentation of labour was commenced with 10 I.U (international units) of oxytocin because of the poor uterine contractions. Despite augmentation, labour progress was slow with complaints of physical exhaustion by the patient thus necessitating an emergency lower segment caesarean section.

At surgery, multiple firm to hard nodules of varying sizes were seen scattered throughout the omentum. There were also similar pedunculated uterine nodules. A portion of the omentum with some of these nodules was excised for histopathologic examination. She was delivered of a live male neonate with good APGAR scores.

Surgical procedure was complicated by intra-operative blood loss from the uterus (because

there was difficulty securing haemostasis due to uterine atony) and patient received 3 units of transfusion post operatively. Otherwise, recovery was relatively uneventful and she was discharged on the 4th day post op. The patient was followed up postoperatively in the surgical outpatient clinic. She was seen once every month for a year and had abdominal ultrasound done once in 3 months over the 1 year follow up period. The follow-up period was uneventful with no disease progression.

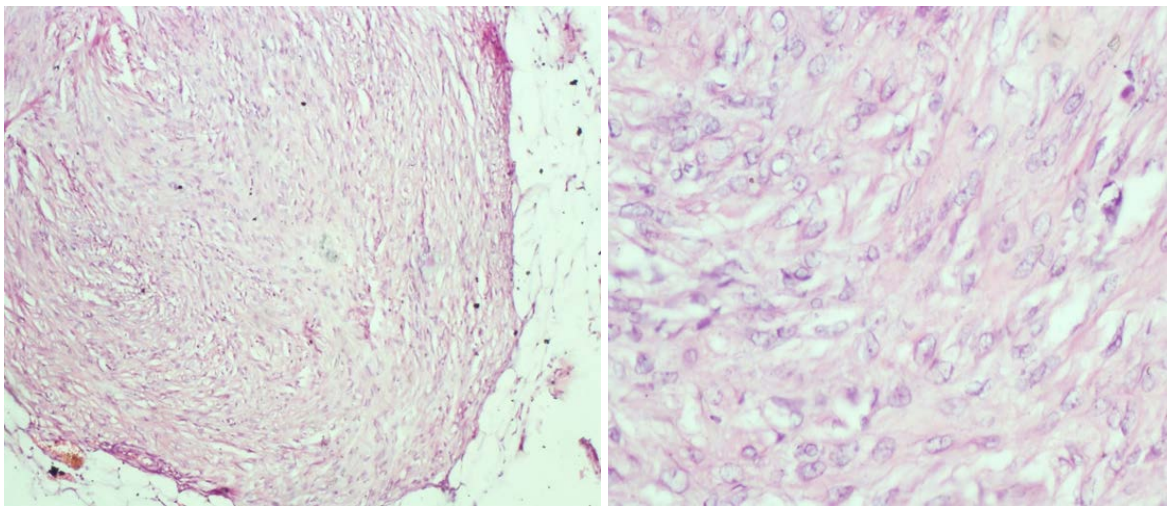
At grossing, a single fibrofatty tissue weighing less than 10g containing multiple firm greyish white nodules was received, the nodules ranged in size from 0.5 cm to 1 cm in diameter and had homogenous greyish-white whorled cut surfaces. Histological examination of theses nodules showed a benign mesenchymal neoplasm composed of plump monomorphic smooth muscle cells disposed in intersecting fascicles and whorled patterns with some focal areas of stromal hyalinization. No mitotic figure or focus of atypia was seen (Fig. 1a & b).

Subsequently, immunohistochemical staining was done for Desmin (Dako, Glostrup, Denmark; M0760; 20020138; 1:50 dilution), CD117 (BiocareMedicals, California, United States; CME296C;052617;1:50dilution), Estrogen Receptor (BiocareMedicals, California, United States; ACA054C; 071619-2; 1:50 dilution), and Progesterone Receptor (BiocareMedicals, California, United States; ACA424C;1:50 dilution) markers. The results showed diffuse and strong positivity of the tumour cells for Desmin (Fig. 2a) and Progesterone Receptor (Fig. 2b) stains. Estrogen Receptor stain was focally positive (Fig. 2c) while CD 117 stain was completely negative (Fig. 2d).

Based on the morphology of the lesion from H&E and the pattern of immunohistochemical staining, the case was conclusively diagnosed as disseminated peritoneal leiomyomatosis.

3. DISCUSSION

Leiomyomas are benign smooth muscle tumours that primarily affects the uterus, small bowel and the oesophagus [9]. Uterine leiomyomas are the most common gynaecological tumours occurring in 20% to 30% of women above 35 years of age [9]. Uncommon growth patterns of leiomyomas include disseminated peritoneal leiomyomatosis (DPL), intravenous leiomyoma, retroperitoneal leiomyoma, benign metastasizing leiomyoma and parasitic leiomyoma [9]. Fasih et al. [10] classified leiomyomas occurring outside the uterus according to the different sites and described them as a separate disease entity referred to as leiomyomas beyond the uterus (LBU) [10]. The atypical locations of these tumours usually present a diagnostic dilemma regarding their origin and biologic nature [11].



A **B**

Fig. 1. Photomicrographs show fascicles of monomorphic smooth muscle cells. Haematoxylin and Eosin stain. (A; x100magnification, B; x400magnification)

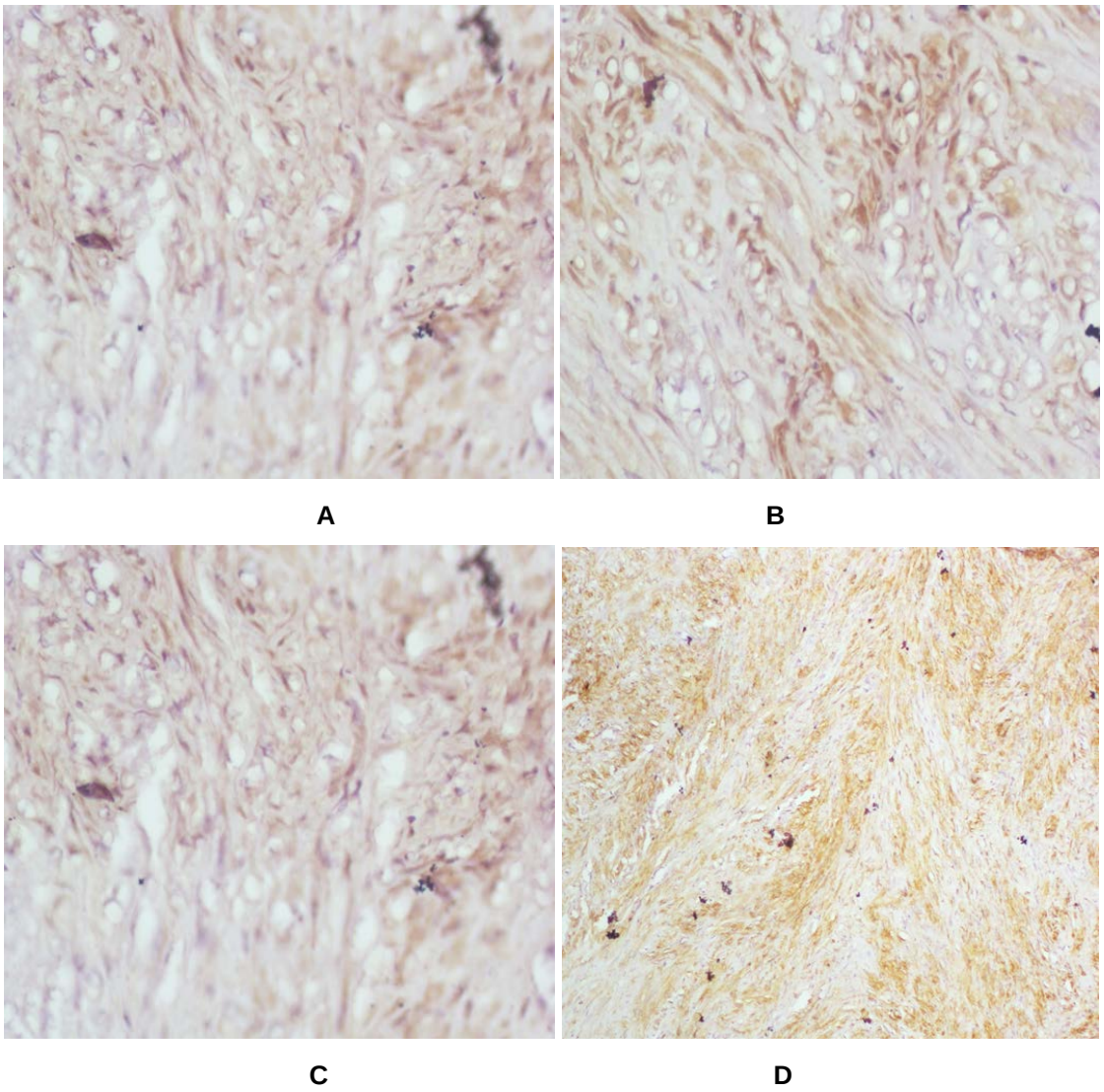


Fig. 2. Photomicrographs show positivity of tumour cells for ER (A; immunoperoxidase stain, x400magnification), PR (B; immunoperoxidase stain, x400magnification), and Desmin (C; immunoperoxidase stain, x100magnification). Tumour cells are completely negative for CD117. (D; immunoperoxidase stain, x400magnification)

DPL is a very rare benign entity characterised by the presence of multiple leiomyomatous nodules throughout the peritoneal cavity, often giving the appearance of metastatic ovarian or primary peritoneal carcinoma [11]. The estimated prevalence of DPL is less than 1/10,00,000 with fewer than 150 cases reported in literature [11].

Although the aetiopathogenesis is still not fully understood, underlying abnormal sub-mesothelial tissue sensitivity to ovarian hormones may be one of the major causes of DPL, this may explain why some DPL cases occur in pregnancy as seen in this index case [9]. The use of oral contraceptives has also been identified as a risk factor [9,12]. Assisted reproductive technology is also a risk factor because of the high serum oestrogen seen in these patients due to iatrogenic ovarian hyperstimulation [12]. Prior laparoscopic removal of uterine leiomyomas has also been identified as a possible aetiology in the development of DPL owing to the potential spread of tumour cells along the surgical tract [9]. Though this index patient has had a previous myomectomy, it was done though a laparotomy rather than laparoscopy. The occurrence of DPL in association with uterine leiomyomas was also corroborated in this index case as pedunculated and intramural uterine nodules were also seen during surgery [5,8,12].

The DPL in this index case was discovered incidentally during an emergency caesarean section (for poor progress in labour and maternal exhaustion), this is similar to what has been noted by other authors that most cases of DPL are asymptomatic and detected incidentally during abdominal surgery for other conditions [2,3,6-9] Some patients present with non-specific symptoms such as heavy uterine bleeding and lower abdominal pain or discomfort while less common clinical presentations include increased frequency of micturition, abdominal swelling and symptoms of obstructive uropathy [9].

Radiological investigations also help in diagnosis but may not be able to rule out other differentials such as peritoneal carcinomatosis, LPD with sarcomatous change, lymphadenopathy and multiple pedunculated leiomyomas [7]. Definitive diagnoses of LPD is made by histopathological examination of excised nodules after laparotomy or laparoscopy [9]. The peritoneal nodules seen in this index patient had the typical macroscopic and microscopic appearance of DPL with hard rubbery consistencies, homogenous grayish-white cut surfaces and bland monomorphic proliferation of smooth muscle cells disposed in interlacing fascicles and whorls with no mitotic activity, atypia or necrosis [12]. The absence of endometrial glands and stroma within these neoplastic nodules of smooth muscle cells rules out an endometriosis [13].

Immunohistochemical studies revealed strong and diffuse positivity for Vimentin, Desmin and Progesterone receptors (PR) with some focal positivity for estrogen receptor (ER) all of which is consistent with a diagnosis of disseminated leiomyomatosis peritonei [3]. The negative expression of CD117 also rules out an extra-intestinal gastrointestinal tumour which is one of the differential diagnosis considered based on the morphology of the tumour cells.

Although LPD is a benign disease, some rare cases of malignant transformation have been reported in the literature [14]. The risk factors for malignant transformation of LPD include a negative history of the following - oral contraceptive use, pregnancy and uterine leiomyoma. Other established risk factors for malignant transformation includes lack of expression of estrogen and progesterone receptors in leiomyoma nodules, and recurrence within 1 year of initial treatment [14].

No standard treatment guidelines exist for the management of DPL, it is individualised based on the patient's age, desire for conception and symptomatology [14]. Asymptomatic DPL requires no therapy because it is usually impossible to remove all nodules and the disease has an indolent course [9,14]. In the index case the diagnosis was incidentally made during an emergency lower segment caesarean section, and she has been stable and asymptomatic on both clinical and radiological follow-up. Treatment is necessary in patients with symptomatic disease and in those with growing or recurrent lesions [15]. Regression has been described with declining levels of estrogen, therefore bilateral salpingooophorectomy or Gonadotropin Releasing Hormone (GnRh) analogue therapy should be considered before contemplating surgical excision of the nodules [14]. This patient has not completed her family and thus desires further conception, hence the need for a conservative line of management. There might be a future possibility of doing a bilateral oophorectomy or gonadal suppression later in the future if the need arises.

Generally, DPL has good prognosis [4,15]. However, some patients may present with life threatening complications such as acute abdomen or peritonitis [8].

4. CONCLUSION

Disseminated peritoneal leiomyomatosis is a rare benign disease characterised by the presence of multiple leiomyomatous nodules throughout the peritoneal cavity which may be misdiagnosed as disseminated or metastatic intra-abdominal malignancy and thus over treated and on many occasions extensive dissections are performed which may lead to increased morbidity and mortality. Therefore, a high index of suspicion is required for accurate diagnosis and proper management of such cases.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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