

Giant Uterine Lipoleiomyoma Masquerading as Endometrial Carcinoma in a Young Nulliparous Lady - Clinical and Imaging Challenges

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Abstract

Uterine lipoleiomyoma is a benign variant of leiomyoma. It is rare and is composed of smooth muscles and mature adipocytes. It affects perimenopausal and menopausal women. It is important to differentiate it from neoplasms that demand surgical excision. We report a case of giant lipoleiomyoma in a 30 year old nulliparous lady. The objective of presenting this case is to review current concepts and present the imaging findings of this entity.

Keywords: Giant; Lipoleiomyoma; Imaging

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Introduction

Uterine Leiomyomas (UL) are benign mesenchymal tumors, which arise from smooth-muscle cells and extracellular matrix of the uterus and represent the most common tumor of the female genital tract [1]. They are described as giant when the tumour weighs more than 11.4 kg or have a diameter more than 17 cm or dimension of 33 × 28 × 22 cm². The overall incidence of UL is between 4% and 11%; this percentage rises to 35% during the reproductive age and to 40% in women over 50 years of age [1].

Lipomatous uterine tumors are uncommon benign neoplasms, with incidence ranging from 0.03% to 0.2% [3]. They can generally be subdivided into two types: pure or mixed lipomas. The latter consist of lipoleiomyoma, angiomyolipoma and fibrolipoma [2].

Mixed lipomas contain variable amounts of fat, fibrous tissue and smooth muscle while pure lipoma is composed of encapsulated adipose tissue with thin septa of fibrous tissue only [3]. Most of the reported cases are of mixed type and lipoleiomyoma is the most common. The diagnosis of lipoleiomyoma can be made with ultrasound, CT or MRI with good accuracy. However, in cases of giant lipoleiomyomas with cystic degeneration, the diagnosis is difficult using imaging and can be misdiagnosed as uterine malignancy.

Case Report

AD is a 30-year-old nulliparous lady who presented to the Nigeria

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Navy Reference Hospital Ojo, Lagos with progressive 12 months history of abdominal swelling, menorrhagia, abdominal and low back pain with urinary frequency all of 2 months duration. She also had inability to conceive in her 2 year old marriage despite regular, unprotected coitus. There was no history of nausea or vomiting but she admitted history of weight loss. She attained menarche at age 16. She had no known underlying medical condition. On physical examination, she was pale and not in any distress. She had bilateral pitting pedal edema, but had no inguinal lymphadenopathy.

Abdominal examination showed gross abdominal distension. There was a huge mass of 38 weeks size which was firm and non-tender. No vulva or cervical abnormalities. Other systemic examinations were grossly normal. Laboratory investigations which included Fasting blood sugar and two hours post prandial blood glucose, lipid profile, Liver function test, CA 125, serum electrolyte, and full blood counts were within normal range but for PCV which was 24%.

Ultrasonography revealed grossly enlarged mixed echogenic mass arising from the pelvis and extending to the epigastric region. It was difficult to delineate the mass from the uterus

and it demonstrated cystic changes at its central portion with poor vascularity evidenced by poor colour flow on Doppler interrogation. There was associated bilateral mild to moderate hydronephrosis. No ascites, lymphadenopathy or pleural effusion seen.

Abdominopelvic MRI of the (Figures 1 and 2) showed a large, well-defined T1 and T2 mixed intensity lesion with extensive areas appearing hyper intense on both sequences due to the fatty component of the mass. It measured 13.8 × 20.9 × 28.7 cm in AP, transverse and superior-inferior dimensions respectively. The uterine lesion extended to the epigastrium with distortion of the endometrial cavity. Above findings raised a high index of a suspected malignant uterine mass.

Tumor marker (Ca 125) was then requested and a normal value of 28 U/mL was obtained. She had total abdominal hysterectomy after optimization on admission and the resected specimen weighed 12.2 kg. The mass was soft to firm in consistency and the cut surface was yellowish grey with areas of altered blood. Postoperative period was uneventful. Histopathology of the lesion showed areas of smooth muscle cells admixed with nests of mature fat cells and fibrous tissue which were consistent with lipoleiomyoma.

Discussion

Giant uterine myomas are rare benign smooth muscle neoplasms with less than 100 cases documented worldwide. Although the exact etiology is still unknown, genetic predisposition and hormonal influence (Estradiol and progesterone) have been implicated [4]. Fatty metamorphogenesis of the smooth muscle cells are the most likely cause for the development of



Figure 1 Trans-abdominal ultrasonography demonstrating the uterine mass in part with mixed echogenic appearance; fatty components are seen as echogenic areas with central cystic hypoechoic degenerative change containing internal echoes. 'Finger like' inner marginal irregularity of central degenerative area noted.

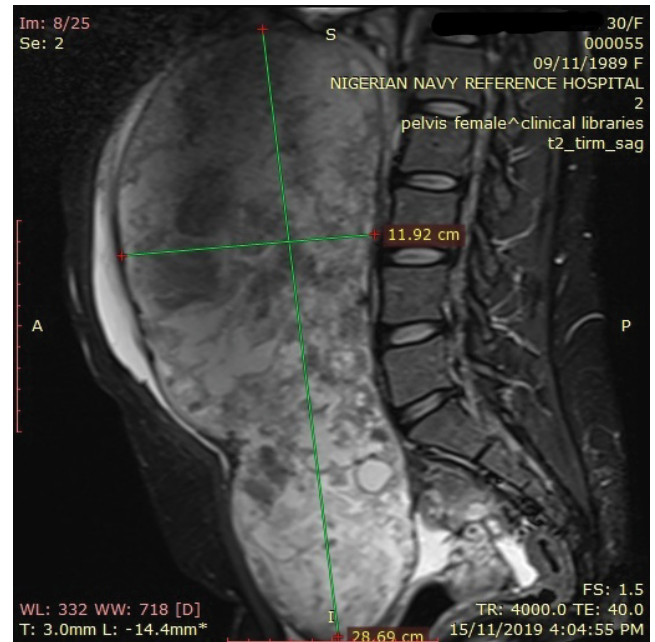


Figure 2 Sagittal T2 FLAIR Magnetic Resonance Image showing grossly enlarged uterus with mixed intensity appearance due to admixture of smooth muscles, fatty elements and areas of cystic degenerations in the mass.

lipoleiomyoma [5]. Besides genetic predisposition and ovarian hormones that play major roles in tumour expansion, a large number of growth factors have also been identified which favour expansion [6]. These are insulin-Like Growth Factor (IGF), Epidermal Growth Factor (EGF), Platelet-Derived Growth Factor (PDGF), Transforming Growth Factor beta (TGF beta), and Basic Fibroblast Growth Factor (BFGF) [6]. These may have a role to play in tumour expansion. The major differential dilemma remains the establishment of bizarre uterine myomas such as the index case versus Endometrial Stromal Sarcoma (ESS), benign cystic teratoma, fibromyolipoma and liposarcomas. The main characteristics of malignant differentials consist of infiltrative myometrial growth pattern and vascular invasion with presence of necrotic areas, and mitotic activity which imaging can depict with high degree of accuracy.

Lin and Hanai reported that uterine lipoleiomyoma may be associated with metabolic diseases including hyperlipidemia, hypothyroidism, and diabetes mellitus [7]. They also asserted that changes in lipid metabolism and other non-lipid mechanisms occurring during menopause might play important roles in the development of lipomatous changes in leiomyoma. Our patients' medical history and laboratory findings were negative for any of these metabolic abnormalities and the early age at presentation of our patient with the size of her tumour made her case unique.

Patients with lipomatous uterine tumours are often asymptomatic. However, they may present with spectrum of clinical manifestations that vary according to tumour size and location. Symptoms may include pelvic pain, increased or abnormal menstrual bleeding, infertility problem, or pressure

effects on surrounding organs like bladder or rectum [8]. Our patient presented with similar features.

Though various imaging features of the tumour in different modalities are important to guide the final diagnosis, pathological examination is required for confirmation.

Ultrasonography is the preferred initial imaging tool for evaluation of the tumour because of its wide availability, being inexpensive, and least invasive character [5]. On ultrasonography, the fat components of the mass demonstrate characteristic echogenic appearance with areas of cystic degeneration appearing echolucent as was seen in the index case. Colour Doppler interrogation can be used to assess vascularity of tumours, an important feature that helps differentiate malignant from benign lesions [8]. Colour Doppler interrogation of the mass in our patients revealed

poor vascularity evidenced by paucity of colour flow. This finding highly suggested benignity of the mass in question.

Computed tomography scan is more specific than ultrasonography as it clearly demonstrates the fatty component of the lesion which appears low in attenuation with Hounsfield units between -40 and -1009. In cases of malignant transformation, enhancement pattern with intravenous contrast administration and presence of regional lymphadenopathies are important findings.

With its multiplanar capabilities, MRI has better tissue characterization and the ability to demonstrate fat component by fat-saturated sequence is the best modality for diagnosis [9]. Lipoleiomyoma demonstrates heterogeneous signal intensity with fat and non-fat soft tissue content and shows decreased signal only in parts of the lesion on fat-saturated images as seen in the images of our patient (**Figures 2 and 3**). The fat component is usually easily differentiated from haemorrhage because of chemical shift, though this may sometime be difficult. In these instances, chemical shift imaging is helpful in making the distinction [9].

When asymptomatic, LL requires no treatment. However, numerous established management options are available for symptomatic variety of which one or a combination can be employed. They include surgery, medical or hormonal treatment and uterine artery embolization [8]. The treatment should be individualized according to many factors including age of patient, fertility status, the severity and type of symptoms, suspicion of malignancy, the location and size of myomas, and desire of patient [8]. Surgery is most frequently preferred for treatment of giant leiomyomas. In cases of patients of reproductive age with ultimate scope of fertility preservation as the index case, simple myomectomy as the gold standard remains a controversial issue. If fertility preservation is not required, the standard surgical intervention for bizarre leiomyoma that shows a benign clinical course is a simple hysterectomy [10].

Conclusion

Our patient had simple hysterectomy with no post-op complications. She was discharged home one week post-op and has been stable.

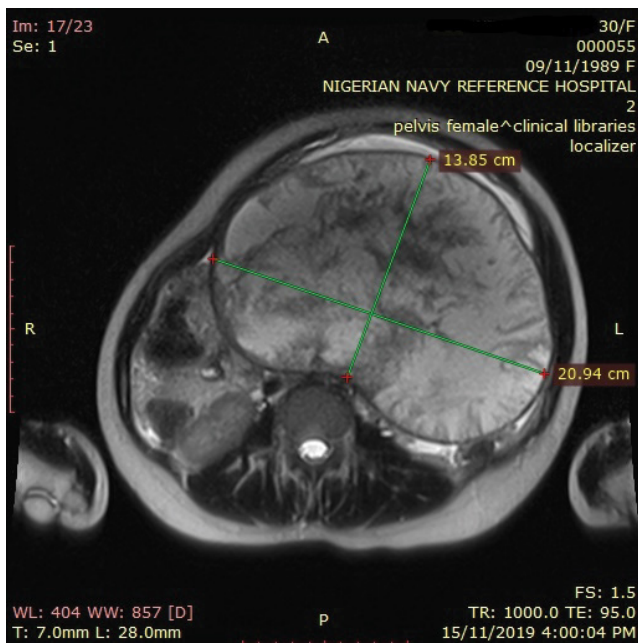


Figure 3 Axial Non-contrast T1W Magnetic Resonance Image showing grossly enlarged uterine mass with heterogeneously intense appearance.

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